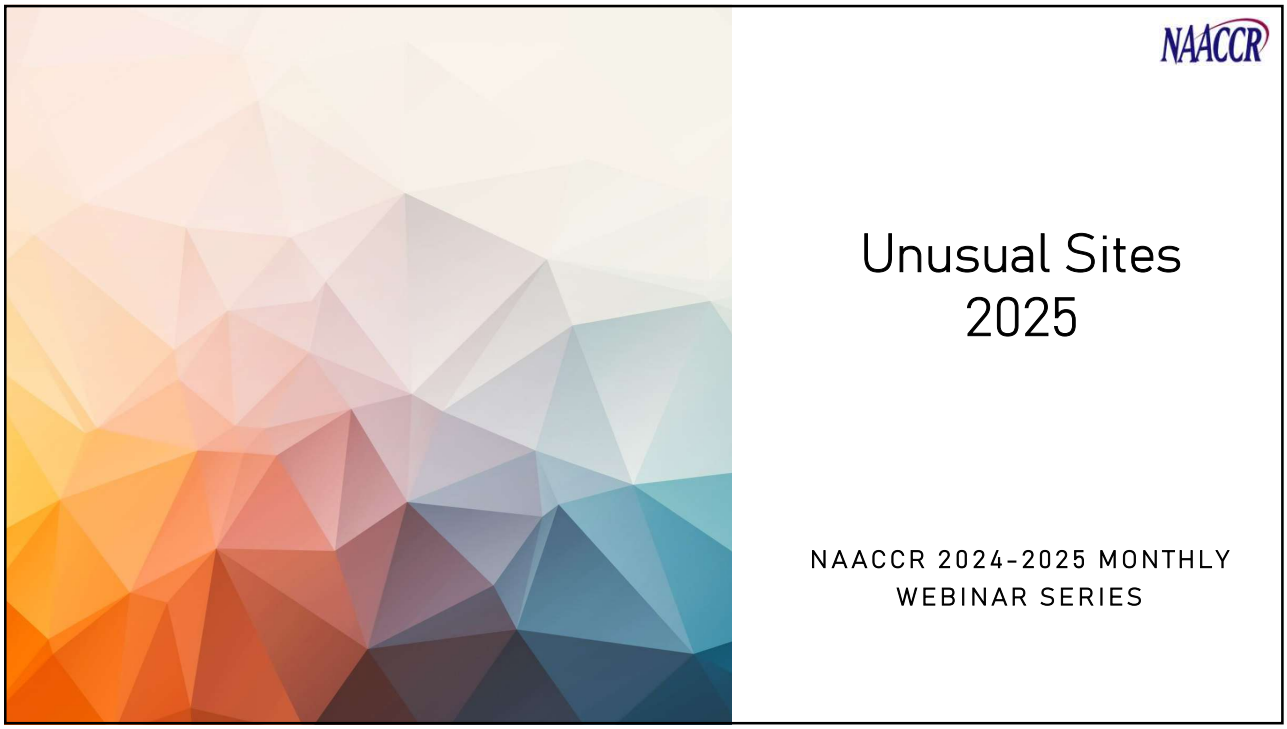
NAACCR
Unusual Sites
2025
NAACCR 2024-2025 MONTHLY
WEBINAR SERIES

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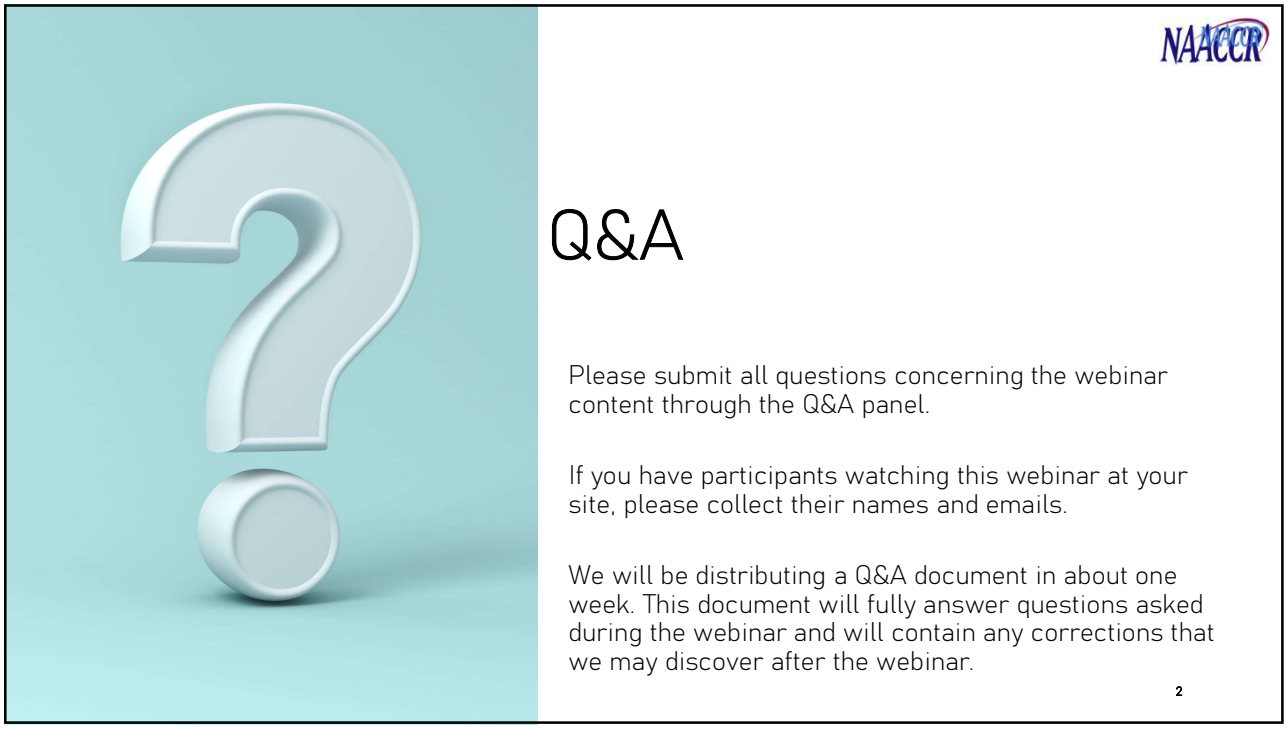
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Q&A
Please submit all questions concerning the webinar content through the Q&A panel.
If you have participants watching this webinar at your site, please collect their names and emails.
We will be distributing a Q&A document in about one week. This document will fully answer questions asked during the webinar and will contain any corrections that we may discover after the webinar.
2

2

Fabulous Prizes

3

GUEST PRESENTER

Vicki Hawhee, BSMT, MEd, ODS

4

Abstracting Unfamiliar Sites

5

Tips

- When abstracting an unfamiliar site, I always look at the AJCC Chapter (to identify specifically when I need for staging) and the specific SSDIs, etc.
- This way as I go through all of the potentially unfamiliar terminology (i.e. gonioscopy), I can concentrate on finding the specific information I need and not get caught up in all the words I don't know.

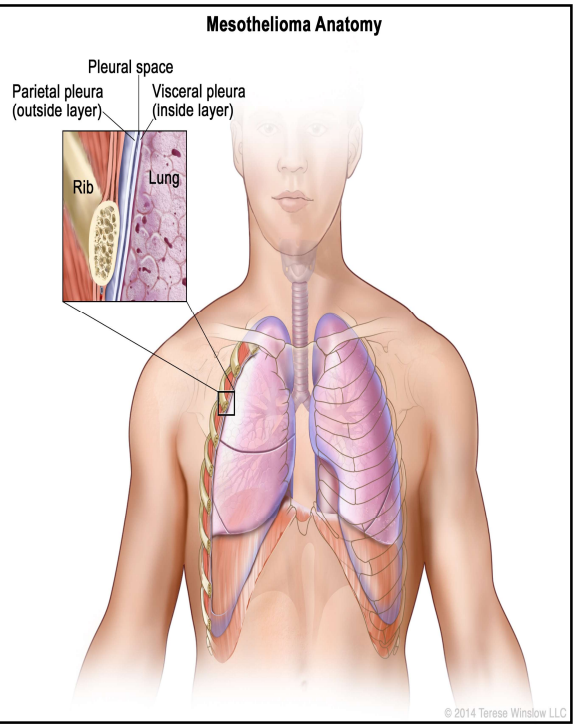
6

Pop Quiz 1

- What is a gonioscopy?
  - A. A scope of the lung pleura
  - B. Type of eye exam
  - C. A scope used to view the alveoli of the lung
  - D. A tool to distinguish melanocytes from goblet cells

7

Mesothelioma



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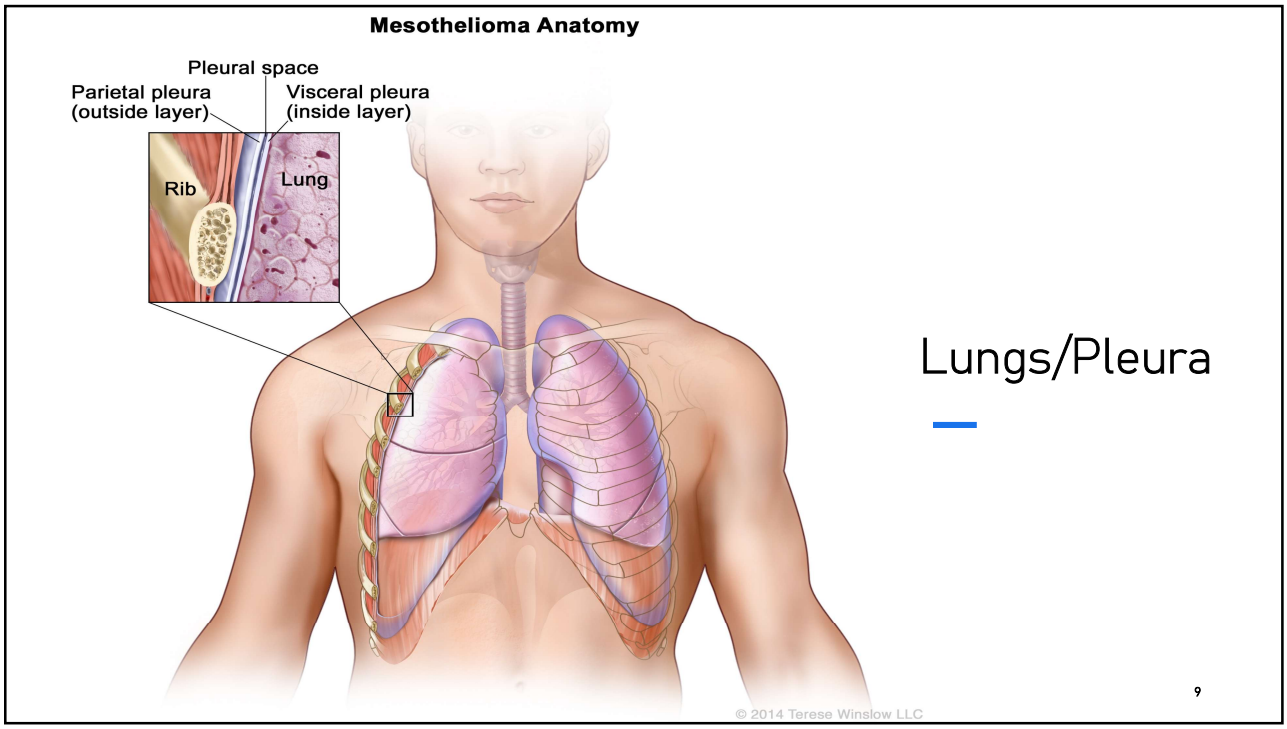

Lungs/Pleura

9

NAACCR

4 types of malignant mesothelioma

- 1. Pleural
- 2. Peritoneal
- 3. Pericardial
- 4. Testicular

10

What is asbestos?

11

Malignant Pleural Mesothelioma (2024)

1. Do not need size for the T category, you need the extension (what structures are involved) and – NEW – pleural thickness (2025).
2. Only one SSDI - Pleural effusion – malignant or not – microscopically confirmed or not

12

Case #1 Clinical Information

- 7/6/24- 72-year-old married female presents to the ER with chest pressure and difficulty catching her breath and back pain. History of smoking for 20 years (quit 30 years ago). Scans identified pleural effusions and patient scheduled for a thoracentesis. Suspected lung malignancy or mesothelioma.
- CT chest: Moderate right greater than left pleural effusions with confluent lower lobe atelectasis.
- Right ultrasound guided thoracentesis: Right pleural fluid: highly atypical mesothelial proliferation, strongly favor malignant mesothelioma
- Consultation following thoracentesis – physician states "patient with a malignant pleural effusion will be scheduled for a VATS procedure with biopsies."
- PET/CT: FDG avid adenopathy in the bilateral mediastinum and bilateral hila without clear primary lung lesion. findings concerning for metastatic disease. there is thickening of the esophagus with increased FDG activity in the distal esophagus and gastroesophageal junction and adjacent nodular areas of FDG accumulation concerning for adenopathy; bilateral pleural effusions

13

Pop Quiz 2

A minimally invasive surgical procedure that allows a doctor to examine the pleural cavity

A. Thoracentesis
B. Thoracoscopy
C. Pleurodesis
D. Thoracotomy

14

Procedure answer

- A minimally invasive surgical procedure that allows a doctor to examine the pleural cavity
  A. Thoracentesis – removes fluid or air from the pleural space
  B. Thoracoscopy
  C. Pleurodesis – procedure to prevent pleural effusion from returning
  D. Thoracotomy – Surgical procedure to gain access into the pleural space

15

Video Assisted Thoracoscopy

- 7/7/24-Right VATS (video assisted thoracoscopy) drainage of pleural effusion, pleural biopsies and talc pleurodesis. Flexible esophagogastroscopy.
- No abnormalities noted in stomach or esophagus. We then made a small incision and dissected through the rib cage and into the chest cavity. We inserted a pull sucker device and evacuated approximately a 2 liters of fluid. This was sent for cytology. We then inserted a 12 mm port and a thoracoscopic camera. There were obvious metastatic nodules on the right pleural surface. Biopsies were taken and sent to pathology for review. 5 g of aerosolized talc was aerosolized through the right chest cavity.
- **Right pleura biopsies: malignant mesothelioma**
- CK7 FOCALLY POSITIVE; CK20 NEGATIVE; TTF1 NEGATIVE; MOC31 NEGATIVE; WT1 POSITIVE; CALRETININ POSITIVE; D240 POSITIVE; GATA3 NEGATIVE; BAP1 NEGATIVE; KI-67 ELEVATED. CARIS: MSI STABLE; ALK-, BRAF-, EGFR-, KRAS-
- Right pleural fluid: clusters of atypical mesothelial cells, consistent with malignant mesothelioma.

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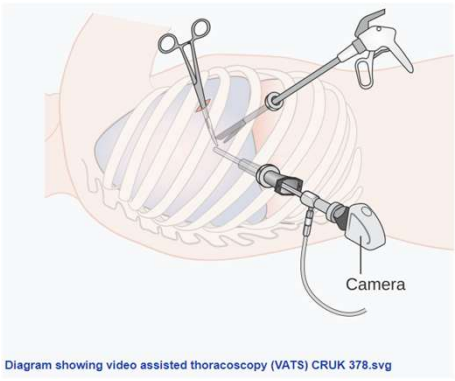
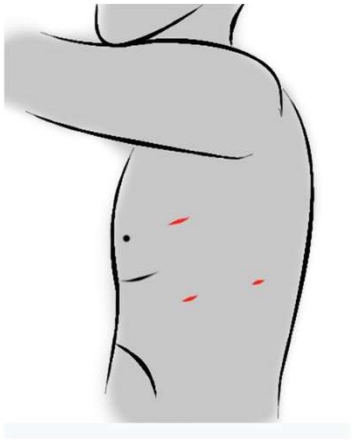
VATS





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Why do they do all the molecular testing?

- **Right pleura biopsies: malignant mesothelioma**
- CK7 FOCALLY POSITIVE; CK20 NEGATIVE; TTF1 NEGATIVE; MOC31 NEGATIVE; WT1 POSITIVE; CALRETININ POSITIVE; D240 POSITIVE; GATA3 NEGATIVE; BAP1 NEGATIVE; KI-67 ELEVATED. CARIS: MSI STABLE; ALK-, BRAF-, EGFR-, KRAS-
- Helps to identify that pleura is the primary site (not metastasis from another primary)
- Calretinin, D240 and WT-1 are markers of mesothelioma (and all positive in this specimen)
- May help identify specific drugs that will/will not work for this particular malignancy
- Help to identify an appropriate clinical trial

The Clinical Trials Connector lists agents that are matched to available clinical trials according to biomarker status. In some instances, older-generation agents may still be relevant in the context of new combination strategies and, therefore, will still appear on this report.

Visit www.CarisMolecularIntelligence.com to view all matched trials. Therapeutic agents listed below may or may not be currently FDA approved for the tumor type tested.

TARGETED THERAPY CLINICAL TRIALS (6)

Drug Class	Biomarker	Method	Analyte	Investigational Agent(s)
FAK inhibitors (2)	NF2	NGS	DNA-Tumor	defactinib
HDAC inhibitors (2)	BAP1	NGS	DNA-Tumor	abexinostat, entinostat
PARP inhibitors (2)	BAP1	NGS	DNA-Tumor	ZX-121, BGB-290

( ) = represents the total number of clinical trials identified by the Clinical Trials Connector for the provided drug class or table.

The Clinical Trials Connector may include trials that enroll patients with additional screening of molecular alterations. In some instances, only specific gene variants may be eligible.

18

Talc Pleurodesis (and VATS) – To Code or Not to Code

Talc - CAnswer Forum (from 2020)

- A Video-Assisted Thoroscopic Surgery (VATS) is done to diagnose lung cancer. If a biopsy was performed code 02 in Surgical and Diagnostic Staging Procedure. The pleurodesis is performed to prevent further build up in the lung in the space and the talc is the chemical used. Unless, this is stated in the medical record as a Palliative Procedure, code 1 Other Cancer treatment that cannot be appropriately assigned to specified treatment data items (surgery, radiation, systemic therapy).

19

Clinical Staging and Treatment Plan

- 7/20/24-Oncology consultation – patient with malignant mesothelioma, likely bilateral. Multiple thoracentesis done for recurring pleural effusions. Not a surgical candidate, needs to start first-line therapy with nivolumab and ipilimumab (mesothelioma protocol). Send tissue for next-generation sequencing. Will present case at tumor board. No prior history of asbestos exposure. Clinical stage IIIB
- Tumor Board: Reviewed pathology histology consistent with mesothelioma epithelioid type
  We all agreed that patient has bilateral disease making her unresectable
  Discussed options for systemic therapy including carboplatin pemetrexed, enrollment into a clinical trial combining chemotherapy with tumor treating fields and immunotherapy. We considered the patient is a candidate for all systemic treatment
- Med/Onc Note at the time the patient started treatment: Patient with Epithelioid mesothelioma, stage IV bilateral disease, started ipilimumab and nivolumab.

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Pop Quiz 3

Right ultrasound guided thoracentesis: Right pleural fluid: highly atypical mesothelial proliferation, strongly favor malignant mesothelioma

Code	Description
0	Pleural effusion not identified/not present
1	Pleural effusion present, non-malignant (negative)
2	Pleural effusion present, malignant (positive) Physician states pleural effusion is malignant in the absence of positive cytology
3	Pleural effusion, atypical/atypical mesothelial cells
4	Pleural effusion, NOS
8	Not applicable: Information not collected for this case (If this item is required by your standard setter, use of code 8 will result in an edit error.)
9	Not documented in medical record Pleural effusion not assessed or unknown if assessed

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Right ultrasound guided thoracentesis: Right pleural fluid: highly atypical mesothelial proliferation, strongly favor malignant mesothelioma
Consultation following thoracentesis – physician states "patient with a malignant pleural effusion will be scheduled for a VATS procedure with biopsies."

- **Code 2** when
- a. Pleural effusion microscopically confirmed to be malignant
- b. Pleural effusion is stated to be positive for malignant cells
- c. Pleural fluid cytology described as suspicious or suspicious for mesothelioma
- d. Physician states pleural effusion is positive in the absence of positive cytology
- **Code 3** when
- a. Pleural fluid cytology is described as atypical or atypical mesothelial cells but not
- specifically as non-malignant or malignant)

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Treatment

- 77 year female with a new diagnosis of mesothelioma , s/p C1D1 ipilumimab/nivolumab . She calls with complaints of new cough and chest tightness x2 days. She denies SOB, tachycardia,
- fevers/chills.
- Diagnosis: stage IV malignant pleural mesothelioma
- PleurX is changed every other day, 350-400 cc drained.
- CT Chest WO: Significant improvement/resolution of the right pleural effusion status post placement of a Pleurx catheter. Bilateral pleural thickening with associated changes related to talc pleurodesis. Peripheral pulmonary scarring, atelectasis. Persistent circumferential soft tissue thickening involving the esophagus with similar appearance of mediastinal adenopathy
- Scans continue to show improvement. Immunotherapy paused after 2+ months due to side effects including fatigue, epigastric pain and back pain. Will start back on immunotherapy after a 1 month break and palliative radiation to the chest wall.

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STORE Manual

- **Treatment Plan**
- A treatment plan describes the type(s) of therapies intended to modify, control, remove, or destroy
- proliferating cancer cells. The documentation confirming a treatment plan may be found in several
- different sources; for example, medical or clinic records, consultation reports, and outpatient records.

- The first course of treatment includes all methods of treatment recorded in the treatment plan and
- administered to the patient before disease progression or recurrence.

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PleurX Catheter

- Palliative Pleurx Catheter placement - CAnswer Forum
- If the patient is an analytic case for the reporting facility and a pleurX catheter was placed at the reporting facility to alleviate symptoms then the catheter placement would be captured under data item # 3270 and 3280, Please review all palliative codes, including the combo codes since the catheter and the chemo in your scenerio are stated to palliative. #
- If patient was already diagnosed at an outside facility, then came to your facility for placement of PleurX catheter with no cancer directed treatment administered at your facility then per STORE this case would not be reportable for your facility. #

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Radiation

- Patient has been recommended a palliative course of radiation therapy to the right post upper mid chest wall, site of excruciating pain, with minimal relieve while in pain medications.
- TREATMENT DELIVERED TO RIGHT CHEST WALL WITH PHOTONS AND CONFORMAL TECHNIQUE. TOTAL OF 40GY DELIVERED AT 4GY/FX FOR 10 FRACTIONS.

26

Progression?

- CT Chest: Interval increase in mediastinal and hilar tissue, FDG avid on PET, consistent with active disease. Representative posterior mediastinal soft tissue encasing the distal esophagus measures 7.7 x 4.9 cm, previously 6.1 x 3.4 cm. Representative subcarinal node measures 2.1 cm short axis (1.4 cm). Interval increase in size of moderate loculated right pleural effusion, much of which is fissural. Interval increase in hyperdense right pleural thickening, most pronounced at the right apex.
- Oncology Note - Most recent CT chest was reviewed with Dr. X and Dr. Y. There is evidence of disease progression with enlarging mediastinal mass encasing the distal esophagus now measuring 7.7 x 4.9 (6.1 x 3.4 cm). Also worsening moderate right pleural effusion and pleural nodularity. We discussed the need to start second-line therapy with Carboplatin and Pemetrexed +/- Bevacizumab.
- Patient changed to chemotherapy, first course is over.

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AJCC

- New for 2025 is a version 9 for "Diffuse pleural mesotheliomas" This does not apply to localized pleural mesotheliomas.
- The T category now includes:
- Psum: Adds 3 pleural thickness measurements.
- Fmax: maximal thickness of pleural tumor along the fissures.
- **NOTE: This case was done prior to 2025 and the Psum and Fmax were not provided on any of the reports.

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Grade

2018-2024

Code	Grade Description
1	G1: Well differentiated
2	G2: Moderately differentiated
3	G3: Poorly differentiated
4	G4: Undifferentiated
9	Grade cannot be assessed (GX); Unknown

2025+

Code	Grade Description
L	Nuclear Grade I WITH or WITHOUT necrosis OR Nuclear Grade II WITHOUT necrosis
H	Nuclear Grade II WITH necrosis OR Nuclear Grade III WITH or WITHOUT necrosis
9	Grade cannot be assessed; Unknown

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Case Scenario 1

Data Item	Value
Primary Site	C38.4 (pleura)
Histology	9050/3
AJCC Clinical	cT0 cN2 cM1 Stage 4 (2024)
AJCC Pathological	Blank
Grade Clinical	9
Grade Pathological	9
SS2018	7 (Bilateral hilar nodes)
EOD Tumor	800 (No evid of primary)
EOD Nodes	700 (contralateral hilar)
EOD Mets	05 (malignant pleural eff)
Pleural Effusion	2

Data Item	Value
Diagnostic/Stg Procedure	01
Surgery of Primary Site	00
Chemotherapy	00
Immunotherapy	01
Palliative Care	02 (XRT for symptoms)
Primary Treatment Volume	42 (chest wall)
Draining Lymph nodes	00
Treatment Modality	02 (photons)
EB Planning Technique	04 (conformal)
Dose per Fraction	00400
Number of Fractions	010
Total Dose	004000

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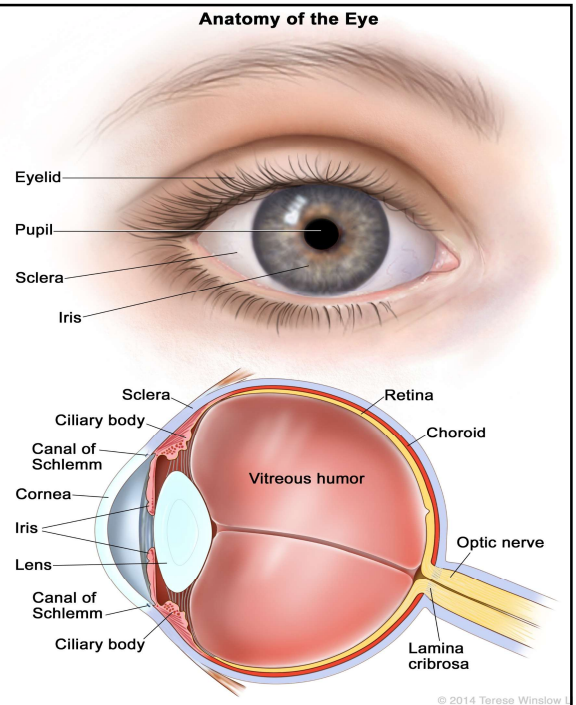
Thank you to Angela Costantini for your help with these slides!

Retinoblastoma



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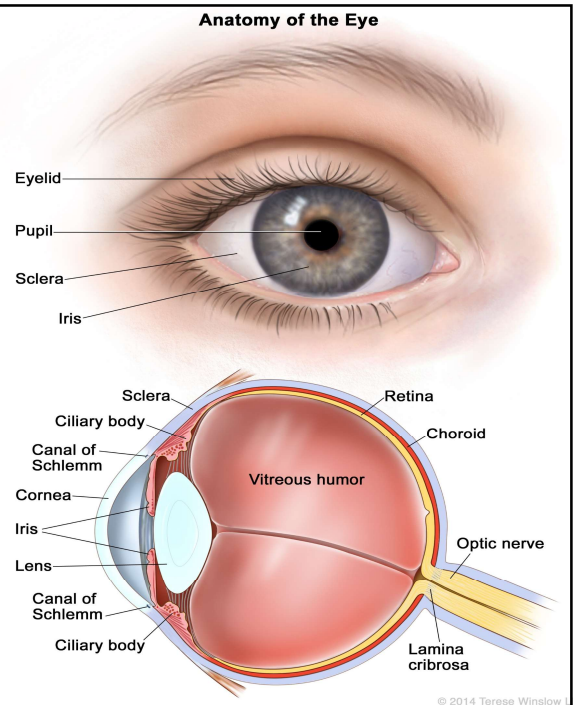
Retinoblastoma

- A rare cancer that develops in the retina
- Frequently occurs bilaterally. May be synchronous or metachronous
- Strongly associated with mutations to the RB1 gene



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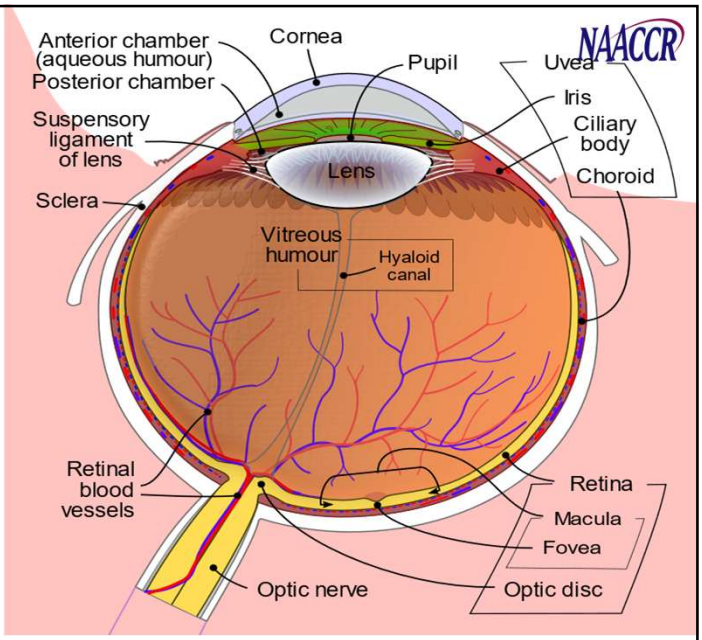
Anatomy of the Eye

- Retina
  - Fovea
  - Macula
  - Optic Disc
- Choroid
- Sclera
- Optic Nerve
- Vitreous humor



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RBL: The Heritable Form

- There are two main types of retinoblastoma, a heritable genetic form and a non-heritable form.
  - In children with the hereditary (or germinal) form of retinoblastoma, it is common for the cancer to affect both eyes, or multiple tumors in either eye.
  - Having the hereditary form of the disease also increases a child's risk of developing other cancers later in life (bladder cancer, osteosarcoma, melanoma, etc)

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RBL: Heritable Genes

- Having the RB1 gene mutation is not a guarantee of developing RBL or of passing it on to offspring.
  - Most children (85%) with the genetic form do not have a parent with retinoblastoma; however, the change in the gene occurred in either the egg or the sperm of one parent prior to conception.
    - If a child has the genetic form of RBL and neither parent has the tumor, the chance that RBL will occur in another child in the family is less than 5%. However, the risk of tumor in the offspring of a child with the genetic form of RBL is about 50%.

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35

RBL: Non-Heritable Form

- This form of the RB1 gene mutation cannot be passed to offspring.
  - All RBL patients should have genetic testing. They will be looking for the RB1 gene mutation, MYC-N and chromosome 13 deletions.
  - More than half of the children dxed with RBL have the non-heritable form of the cancer.
    - This means that the RB1 gene mutation is present only in the cells of the eye.

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Trilateral Retinoblastoma

- An intraocular RBL combined with a histologically similar brain tumor, commonly in the pineal gland.
  - About 5% of children with retinoblastoma from germline mutation of the RB1 gene are at risk of developing trilateral retinoblastoma
  - Often called a pineoblastoma
  - 5-year survival prior to 1995 was 6%.
  - From 1995 onwards, it increased to 57% due to high-dose chemo and stem-cell rescue
  - Non-pineal tumors are typically larger, but both have similar outcomes

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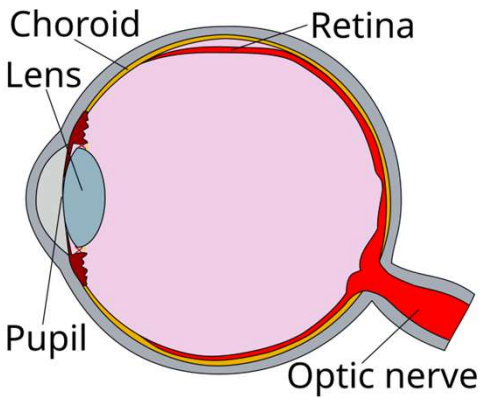
Vocabulary

- Retinal detachment
  - Condition where the retina pulls away from the tissue underneath it
- Vitreous seeding
  - Tumor cells within the vitreous humor
- Subretinal seeding
  - Tumor cells between the retina and the retinal pigment epithelium
- Phthisis bulbi
  - A shrunken eye with little to no vision



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Abstracting Issues

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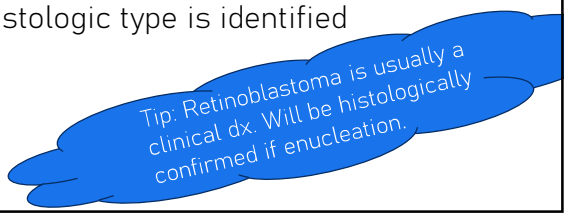
Solid Tumor Rules

- Site Group: Other
- Multiple Primary Rule:
  - M5 Retinoblastoma is always a single primary (unilateral or bilateral)
  - *Bilateral (synchronous or metachronous) are a single primary.*
- Histology Rules
  - H12 Code the histology when only one histologic type is identified
    - 9510/3 Retinoblastoma, NOS
    - 9511/3 Retinoblastoma, differentiated
    - 9512/3 Retinoblastoma, undifferentiated



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Grade

Code	Description
1	G1: Tumor with areas of retinoma [retinocytoma] (fleurettes or neuronal differentiation)
2	G2: Tumor with many rosettes (Flexner-Wintersteiner or Homer Wright)
3	G3: Tumor with occasional rosettes (Flexner-Wintersteiner or Homer Wright)
4	G4: Tumor with poorly differentiated cells without rosettes and/or with extensive areas (more than half of tumor) of anaplasia
A	Well differentiated
B	Moderately differentiated
C	Poorly differentiated
D	Undifferentiated, anaplastic
9	Grade cannot be assessed (GX); Unknown

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Stage

- AJCC Notes–See page 825
  - Clinical and Pathological values are significantly different!
  - Enucleation is required for pT
  - Heritable Trait
- EOD
  - Values for Clinical and Pathologic assessment (see note 3)
- Summary Stage 2018

Note 3: Pathological staging information from an enucleation always takes precedence over clinical staging, except in cases with neoadjuvant treatment where clinical disease is as extensive as or more extensive than disease at surgery

Code	Description
100	CLINICAL assessment only Intraretinal tumor(s) with subretinal fluid › Equal to but not greater than 3 millimeter (mm) › AND Location not closer than 1.5 mm to optic disk or fovea
125	CLINICAL assessment only Intraretinal tumor(s) with subretinal fluid › Greater than 3 millimeter (mm) › AND Location closer than 1.5 mm from disc or fovea
150	CLINICAL assessment only Intraretinal tumor(s) with subretinal fluid ≤5 mm from the base of any tumor Tumor confined to retina, NOS Localized, NOS
175	PATHOLOGICAL assessment only Intraocular tumor(s) WITHOUT any › Local invasion › Focal choroidal invasion › Pre- or intralaminar involvement of the optic nerve head Tumor confined to retina, NOS Localized, NOS

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International Intraocular Retinoblastoma Classification (IIRC) Staging

- Group A: very low risk; all tumors 3mm or smaller, confined to retina and at least 3mm from foveola and 1.5mm from optic nerve. No vitreous or subretinal seeding.
- Group B: low risk; no vitreous/subretinal seeding, discrete retinal tumor of any size or location not fitting a Group A tumor.
- Group C: moderate risk; focal vitreous/subretinal seeding and discrete retinal tumor of any size and location. Seeding must be local, fine and theoretically treatable with radioactive plaque.
- Group D: high risk; diffuse vitreous/subretinal seeding and/or non-discrete endo/exophytic disease. More extensive seeding than Group C. May consist of "greasy" vitreous seeding or avascular masses. Subretinal seeding may be plaque-like.
- Group E: very high risk; eyes that have been destroyed anatomically/functionally with one or more of the following: irreversible neovascular glaucoma, massive intraocular hemorrhage, aseptic orbital cellulitis, abutting lens, diffuse infiltrating RBL and phthisis or pre-phthisis

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International Retinoblastoma Staging System (IRSS) Staging

- 0: Patient treated conservatively
- I: eye enucleated, completely resected histologically
- II: eye enucleated, microscopic residual tumor
- IIIa: regional extension with overt orbital disease
- IIIb: regional extension with preauricular or cervical LN extension
- IVa: hematogenous metastasis (without CNS involvement)
  - 1: single lesion
  - 2: multiple lesions
- IVb: CNS extension (with or without any other site of regional or met dz
  - 1: prechiasmatic lesion
  - 2: CNS mass
  - 3. leptomeningeal and cerebrospinal fluid disease

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Treatment

- *The goal of first course treatment for retinoblastoma is remission.*
  - *For retinoblastoma cases, if laser therapy, cryotherapy and chemo are all utilized, but the eye is just not responding as hoped, enucleation will be done, and CAN be counted as first course treatment.*
  - *If a patient is given treatment and the eye is saved initially, a recurrence of the tumor that leads to an enucleation would be considered subsequent treatment.*

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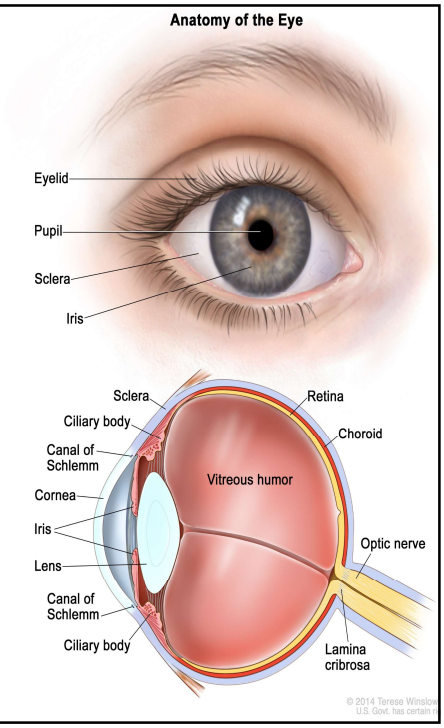
Treatment

- Intravitreal chemotherapy (IVitC)
  - Chemotherapy is injected directly into the eye
- Plaque therapy
  - HDR surgically placed directly onto the sclera over the tumor. The plaque is removed after a few days.
- Photocoagulation
  - Using laser light to destroy blood vessels that supply nutrients to the tumor
- Thermotherapy
  - Using heat to destroy cancer cells
- Cryotherapy
  - Using extreme cold to destroy cancer cells
- Enucleation
  - Surgical removal of the entire eye



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Treatment Tips

- Enucleation as a last resort is not automatically subsequent treatment.
- Surgical procedures will include enucleation (there's never a partial resection of an eye), cryosurgery (A130) or laser surgery (A140).
  - Cryo and laser surgeries often happen DURING an examination under anesthesia (EUA) and are usually done before taking the step towards enucleation.
- Radiation is no longer a common treatment method. If it's noted, it's likely for metastatic disease somewhere else in the body.

- A100 Local tumor destruction, NOS
  - A110 Photodynamic therapy (PDT)
  - A120 Electrocautery; fulguration (includes use of hot forceps for tumor destruction)
  - A130 Cryosurgery
  - A140 Laser
- [SEER Note: Assign code A140 for laser hyperthermia of eye for retinoblastoma.]
- *No specimen sent to pathology from surgical events A100–A140*

- A400 Total surgical removal of primary site; enucleation
  - A410 Total enucleation (for eye surgery only)

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Case Scenario 2

Retinoblastoma

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Case Scenario 2

EOD 100 CLINICAL assessment
Intraretinal tumor(s) with subretinal fluid
•Equal to but not greater than 3 millimeter (mm)
•AND Location not closer than 1.5 mm to optic disk or fovea

Data Item	Value
Primary Site	C69.2
Histology	9510/3
Grade Clin	9
Grade Path	9
Diagnostic Confirmation	6
Tumor Size Summary	002

Data Item	Value
AJCC Clin	cT1a cN0 cM0 Stage 1
AJCC Path	Blank
Summary Stage 2018	1 Localized
EOD Prim Tumor	100
EOD LN	000
EOD Mets	00

Data Item	Value
Diagnostic/Stage Proc	00-None
Surgery Prim Site 2023	A140 Laser Hyperthermia. No tissue sent to pathology.

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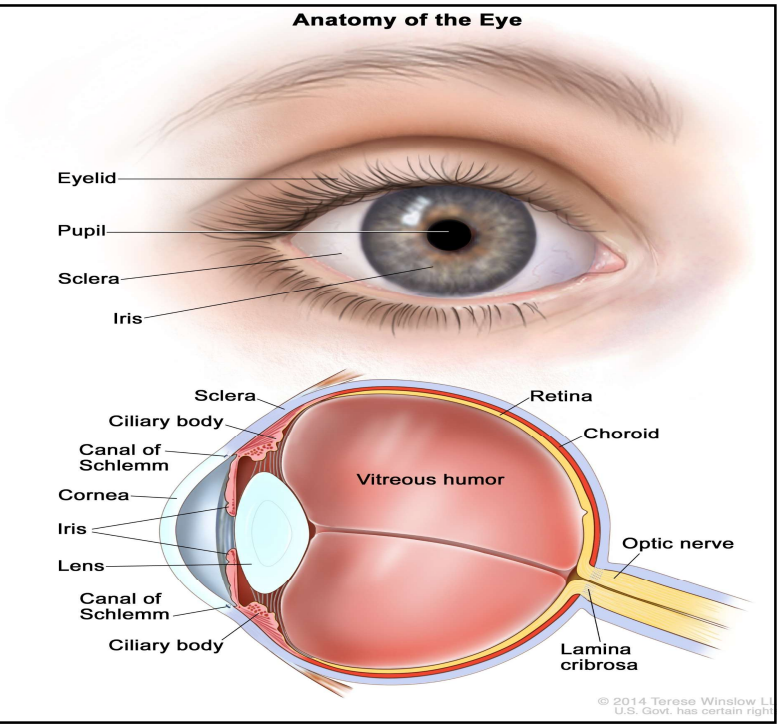
Ocular Adnexa Lymphoma



50

Case #3 Clinical History

- Patient went to see his ophthalmologist with progressive left eye swelling and associated double vision which began about 1 year prior and the ophthalmologist ordered and MRI/MRA. On PE the patient had left orbital proptosis.
- MRI: Bilateral extraconal solid hypercellular restricting orbital lesions. Primary differential considerations are orbital lymphoma . There is a secondary proptosis. No intracranial extension. Further evaluation with postcontrast study to better assess possible perineural spread. Tissue diagnosis is recommended. MRA normal.
- Consultation:
  - The patient is a 62-year-old male with a history of hypertension and hyperlipidemia who comes with bilateral extraconal solid hypercellular restricting orbital lesions. Consideration of orbital lymphoma. He also has secondary proptosis but no intracranial extensions. Tells me he feels relatively well with some degree of impairment of his vision.
  - His ophthalmologist has planned biopsy of the lesion noted on his most recent MRI. Currently denies any significant concerns at present. His left eye has been slightly protruding and reports that blood work revealed thyroid function was normal.
  - It appears he has these issues for about 1 year.
  - EYES: Left eye proptosis. Pupils equal and reactive. Extraocular movements intact. Conjunctive pink, moist. See me in about a month once we have the biopsy results. We briefly went over the treatment options about this disease including localized radiation therapy and in rare occasions systemic rituximab.

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POP QUIZ 4
Bulging of the eyes

A. Conjunctivitis
B. Orbiculus ciliaris
C. Exophthalmos
D. Retinoschisis

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POP QUIZ 4 (answer)
Bulging of the eyes

A. Conjunctivitis – inflammation of the conjunctiva
B. Orbiculus ciliaris – Darkly pigmented posterior zone of the ciliary body
C. Exophthalmos (same as proptosis)
D. Retinoschisis – Degenerative splitting of the retina

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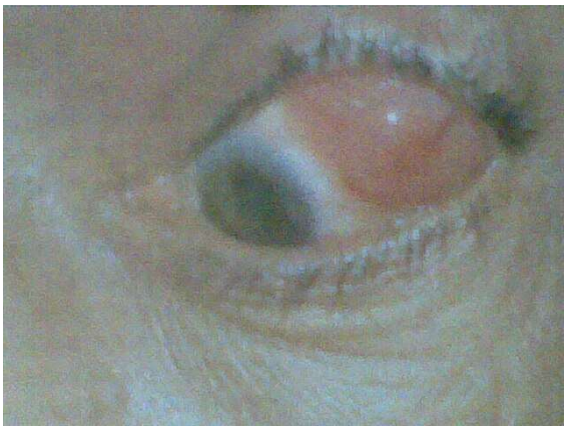
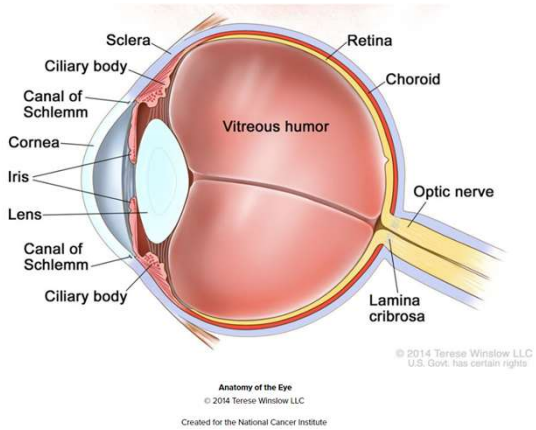
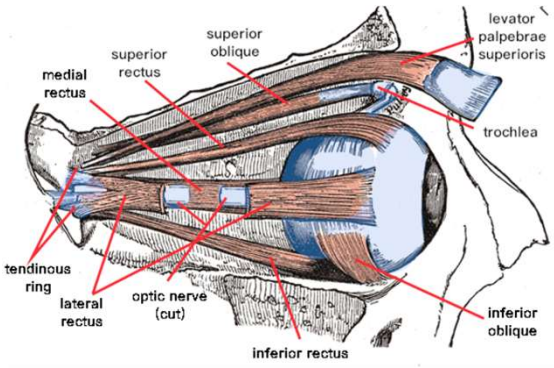


Orbital lymphoma with marked proptosis

54

Anatomy of the Eye





https://upload.wikimedia.org/wikipedia/commons/3/39/Extraocular_Eye_Muscles.png

55

What do I need to look for?

- AJCC Chapter 71 (Ocular Adnexal Lymphoma)
  - For the T category – is the conjunctiva involved or what other structures. (conjunctiva is a mucous membrane that covers the front of the eyes and lines the inside of the eyelids) Any head/neck nodes? Any other extranodal sites involved? Is the bone marrow involved?
  - No SSDIs

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Pathology

Procedure: Bilateral anterior orbitotomies with biopsy of orbital masses.

- Right orbit – fat pad was found to be infiltrated with a tan amorphous mass which was carefully dissected away and a large biopsy taken.
- Left eye did not reveal an easily accessible mass but a small amount of tissue was able to be obtained.
  - Orbit "mass", inferior, right, biopsy: Mature low grade B-cell lymphoma. See note.
  - Orbit "mass", superior, left, biopsy: Mature low grade B-cell lymphoma. See note.

NOTE:

- The lymphocytes are positive for PAX-5, CD20 and BCL-2 while negative for CD43, CD5, CD3, CD10, cyclin D1, BCL-6 and CD21.
- The immunophenotypic findings are consistent with a marginal zone B-cell lymphoma.
- Clinical correlation with the corresponding flow cytometry results is advised.
- The orbital location suggests an extranodal marginal zone B-cell lymphoma of mucosa associated lymphoid tissue (MALToma), however, clinical correlation is advised to exclude a systemic process.

Bone marrow biopsy: Negative

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Additional Clinical Information/Treatment plan

PET Scan: Intense FDG activity corresponding to an approximate 3 cm left supraorbital soft tissue mass and a smaller 1 cm right supraorbital mass compatible with the patient's history of orbital lymphoma. No other suspicious FDG avid lymphoma nodes or other lesions.

- **Diagnosis:** C88.4 Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue
- [MALTlymphoma] - Left and Right Eye Stage IIE

Radiation: Treatment was delivered to bilateral orbits, with photons and a(n) conformal technique on the TrueBeam(treatment unit). A total of 24 Gy was delivered at 2 Gy per fraction (treatment session). A total number of 12 fractions were delivered over 15 elapsed days.

MRI: He had a brain MRI 3 months later: When compared with the initial MRI, there has been complete resolution of previously noted intraorbital masses in keeping with excellent treatment response in a patient with lymphoma. Normal brain MRI

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Hematopoietic Database – 9699/3

Primary Site(s)

See Module 7

Note: Do not code primary site to spleen (C422). Most common sites of involvement: GI tract (stomach most common), salivary gland, lung, head and neck, ocular adnexa, skin, thyroid, and breast

Rule PH24 Code the **primary site** to the **organ** when neoplasm is present **only** in an **organ.**

Note 1: Includes lymphomas that are primary in the spleen. Splenic primaries are rare. Histologies that arise in the spleen include splenic marginal zone lymphoma (9689/3); hepatosplenic T-cell lymphoma (9716/3); splenic B-cell lymphoma/leukemia, unclassifiable (9591/3); splenic diffuse red pulp small B-cell lymphoma (9591/3); splenic marginal zone diffuse variant (9591/3); splenic EBV-associated B-cell lymphoproliferative disorder (9680/3). Follow-back for additional information when the histology is other than those listed **AND**

- The ***only information is a biopsy of the spleen OR***
- There is a physician statement that the spleen is the organ of origin

SINQ 20130213 confirms "orbits" is considered an organ
Per Rule PH24, code primary site to *orbit*. According to Rule PH24, one is to code the primary site to the organ when *lymphoma* is present only in an organ. Note 2 under this rule also instructs one to capture the secondary involvement of distant lymph nodes and/or bone marrow in CS extension fields.

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POP Quiz 5

What is the clinical grade for this case?

Orbit "mass", inferior, right, biopsy: Mature low grade B-cell lymphoma. Most compatible with a marginal zone B-cell lymphoma

A. 1 (G1 0-5 centroblasts per 10 HPF)
B. 9 (unknown)
C. 8 (not applicable)
D. L (low grade)

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Grade 9

Code	Grade Description
1	G1: 0–5 centroblasts per 10 HPF
2	G2: 6-15 centroblasts per 10 HPF
3	G3: More than 15 centroblasts per 10 HPF but with admixed centrocytes
4	G4: More than 15 centroblasts per 10 HPF but without centrocytes
9	Grade cannot be assessed (GX); Unknown Not a follicular histology (9690/3, 9691/3, 9695/3, 9698/3)

00700: Orbital Sarcoma	2018+	Grade 09
00710: Lymphoma Ocular Adnexa	2018+	Grade 23
00718: Eye Other	2018+	Grade 99

Note 1: Grade is applicable for the follicular lymphomas only (9690/3, 9691/3, 9695/3, 9698/3). For all other lymphoma histologies, code 9.

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Case Scenario 3

Data Item	Value
Primary Site	C69.6
Histology	9699/3
AJCC Clinical	cT2 cN0 cM0 Stg 88
AJCC Pathological	Blank
Grade Clinical	9
Grade Pathological	9
SS2018	1
EOD Tumor	300 (orbit w/o conj)
EOD Nodes	000
EOD Mets	00
No SSDIs	

Data Item	Value
Diagnostic/Stg Procedure	02
Surgery of Primary Site	00
Chemotherapy	0
Immunotherapy	0
Palliative Care	
Primary Treatment Volume	10 (eye)
Draining Lymph nodes	00
Treatment Modality	02
EB Planning Technique	04
Dose per Fraction	00200
Number of Fractions	012
Total Dose	002400

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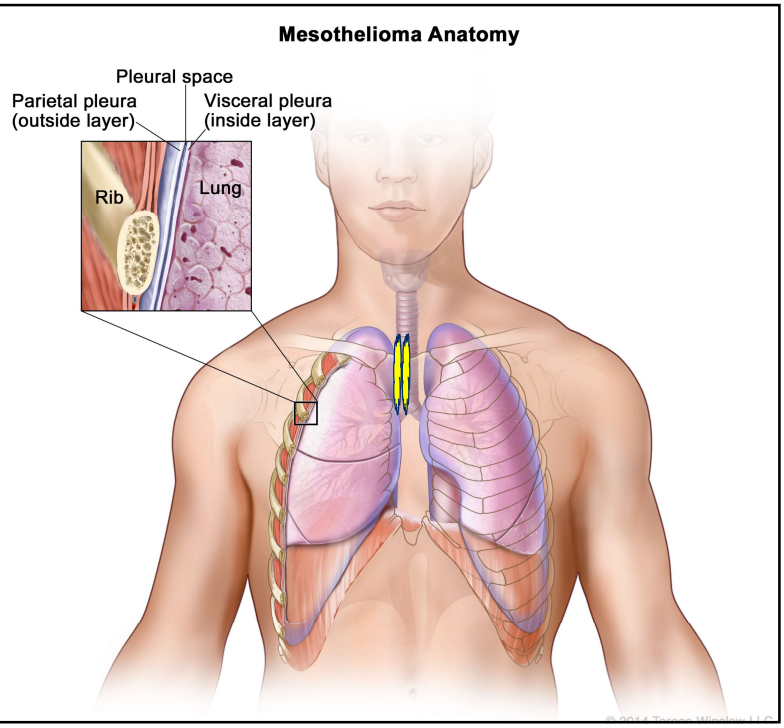
Thymoma



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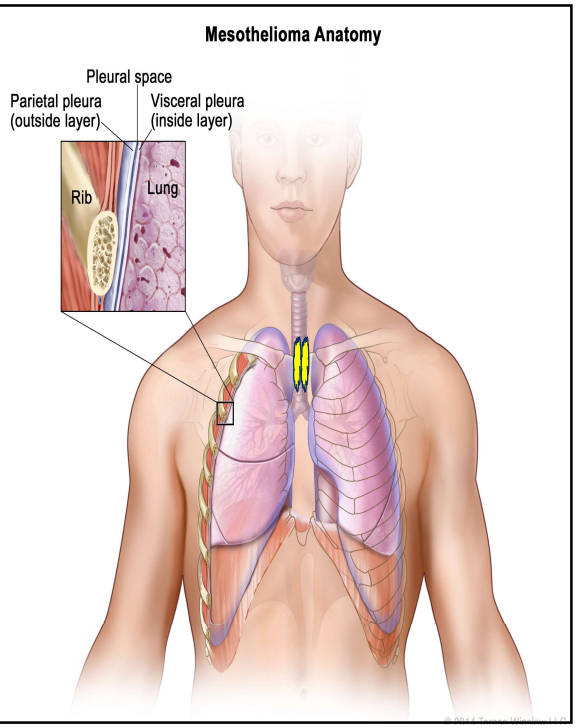
Anatomy

- A gland within the lymphatic system.
  - Develop (and educate) thymus cell lymphocytes (T-cells).
  - Consists of lymphoid and epithelial cells
    - Thymoma and thymic carcinoma develop from the epithelial cells.
- Thymus is active from birth to adolescence.
  - Atrophies as we get older
  - Most patients with thymoma are between 30 and 70
  - Childhood thymoma is very rare



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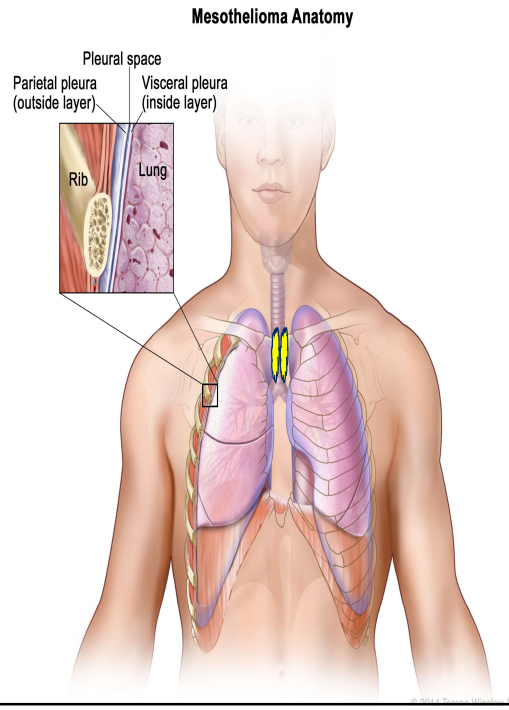
Anatomy

- Located behind the sternum, in front of the aortic arch between the lungs
  - Near, but not covered by, pleura.
- Surrounded by mediastinal fatty tissue.
- Extends from below the thyroid to the 4th rib
- Consists of two lobes
  - Cortex
  - Medulla
- Surrounded by a thin capsule



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Myasthenia gravis

- An autoimmune disease
- Less than half of patients with thymoma present with myasthenia gravis.

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Abstracting and Coding

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Reportability of Thymoma, NOS

- Benign thymoma has *never* been reportable.
- Malignant thymoma has *always* been reportable
  - Statements indicating a thymoma, nos is malignant.
    - Positive lymph nodes
    - Metastasis
    - Multiple foci
    - Invasion

- Thymoma, NOS
  - 8580/1
    - Cases diagnosed prior to 2021 (not reportable)
  - 8580/3
    - Cases diagnosed 2021 or later (reportable)

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Thymoma Histology

- Thymoma NOS 8580/3
  - Thymoma, type A/medullary/spindle cell 8581/3
  - Thymoma, type AB/mixed type 8582/3
  - Thymoma, type B1/lymphocytic/lymphocyte-rich/organoid/predominantly cortical 8583/3
  - Thymoma, type B2/cortical 8584/3
  - Thymoma, type B3/atypical/epithelial/well differentiated thymic carcinoma 8585/3
- Squamous 8070/3
- Adenocarcinoma 8140/3
- NUT carcinoma 8023/3
- Thymic carcinoma 8586/3
  - Thymoma, type C
- Neuroendocrine tumor
- Neuroendocrine carcinoma
  - Small cell
  - Large cell

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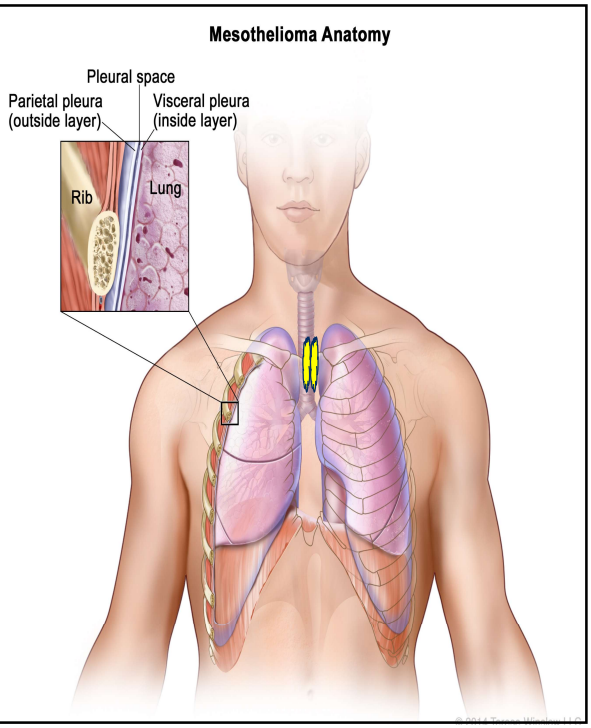
Staging

- AJCC V9 Protocol (for cases dx'd 2025+)
  - Is the tumor ≤ or > 5cm?
  - Is the tumor invading surrounding tissue?
- Summary Stage
- EOD



70

Treatment

- Thymectomy (A400)
- Systemic
  - Neoadjuvant
  - Primary
- Chemoradiation

Surgery Codes

All Other Sites
C142–C148, C170–C179, C239, C240–C249, C260–C269, C300–C301, C310–C319, C339, C379, C380–C388, C390–C399, C480–C488, C510–C5 C529, C570–C579, C589, C600–C609, C630–C639, C680–C689, C690–C6 C740–C749, C750–C759

Codes

A000 None; no surgery of primary site; autopsy ONLY

A100 Local tumor destruction, NOS
A110 Photodynamic therapy (PDT)
A120 Electrocautery; fulguration (includes use of hot forceps for tumor destruction)
A130 Cryosurgery
A140 Laser
[***SEER Note:*** Assign code A140 for laser hyperthermia of eye for retinoblastoma.]
No specimen sent to pathology from surgical events A100–A140

A200 Local tumor excision, NOS
A260 Polypectomy
A270 Excisional biopsy

Any combination of A200, A260, or A270 WITH
A210 Photodynamic therapy (PDT)
A220 Electrocautery
A230 Cryosurgery
A240 Laser ablation
A250 Laser excision

A300 Simple/partial surgical removal of primary site

A400 Total surgical removal of primary site; enucleation
A410 Total enucleation (for eye surgery only)

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NAACCR

Case Scenario 4

Thymoma

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Case Scenario 4

Data Item	Value
Primary Site	C37.9
Histology	8582/3
Grade Clin	9
Grade Path	9
Diagnostic Confirmation	1
Tumor Size Summary	025

Data Item	Value
AJCC Clin	cT1a cN0 cM0 Stage 1
AJCC Path	pT1a pNX cM0 Stage 99
Summary Stage 2018	1 Localized
EOD Prim Tumor	100 (Limited to thymus)
EOD LN	000
EOD Mets	00

Data Item	Value
Diagnostic/Stage Proc	00
Surgery Prim Site 2023	A400

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Paraganglioma

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Case #5

22 Year old female presented with some right ear pain for about 1 year with decreased hearing. Also reports intermittent episodes of bleeding from the ear as well as progressively worsening hearing. Because of insurance issues she has not seen a physician.

- EARS:
  - Right external ear without deformities, erythema or swelling. Right auditory canal with protruding soft tissue mass. Unable to visualize further into canal or tympanic membrane. No active bleeding noted. No mastoid erythema, swelling or tenderness on the right.
  - Left external ear without deformities, erythema or swelling. Left auditory canal is clear, no erythema, no swelling, no otorrhea. Left tympanic membrane is visualized and is intact without perforation or effusion. No mastoid erythema, swelling or tenderness on left.

75

Scans

- CT: New abnormality involving the right jugular foramen, right temporal bone with opacification of the right mastoid air cells and middle ear cavity. Recommend MRI.
  - Differential includes glomus tumor versus infection.
- MRI: Enhancing mass (3.1 x 2.0) of mixed-signal intensity centered in an expanded right jugular foramen with extension into the middle ear cavity and external auditory canal as well as intracranially dural extra-axial. In correlation to the CT there are associated permeative bony destructive changes present.
  - Imaging most consistent with a glomus jugulare paraganglioma.

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What do I need to look for?

- AJCC (Chap 77, Adrenal NET)– Size of tumor and extension
- Summary Stage/EOD (NET Adrenal)- Extension
- No SSDIs
- Specific Primary Site

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Diagnostic Information

Preoperative Diagnosis Right skull base lesion most consistent with jugulotympanic paraganglioma

Operation: Excision of right external auditory canal tissue lesion

- Examination revealed a fingerlike projection of pulsatile tissue filling the external auditory canal, with de-epithelialization of the lateralmost tip, patchy necrotic skin and exudate.
- The tumor was highly fibrous, not compressible, and as expected it was very vascular. The overlying epithelium was separated from the tumoral tissue during the dissection and this was kept separately.
- The first specimen was prepared for frozen section and I continued to remove tumor tissue medially using the scissors. The tumor was too fibrous to break it up with cup forceps.
- The second piece of tumor was removed retained as specimen. At this point, there was tumoral tissue filling the medial third of the external auditory canal, and the posterior tympanic membrane could be visualized by compressing the tumor anteroinferiorly. Keratin debris was cleaned from around the edges of the tumor. The epithelial separation of the anterior and inferior canal wall was partially effaced by infiltrative tumor and irregular bony contour.

PATHOLOGY FINAL DIAGNOSIS:

- Right ear canal mass: Paraganglioma

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Pop Quiz 6

- Paraganglioma is a tumor that forms near the:
  A. Central Nervous System
  B. Somatic Nervous System
  C. Autonomic Nervous System
  D. All of the above

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Pop Quiz 6 (answer)

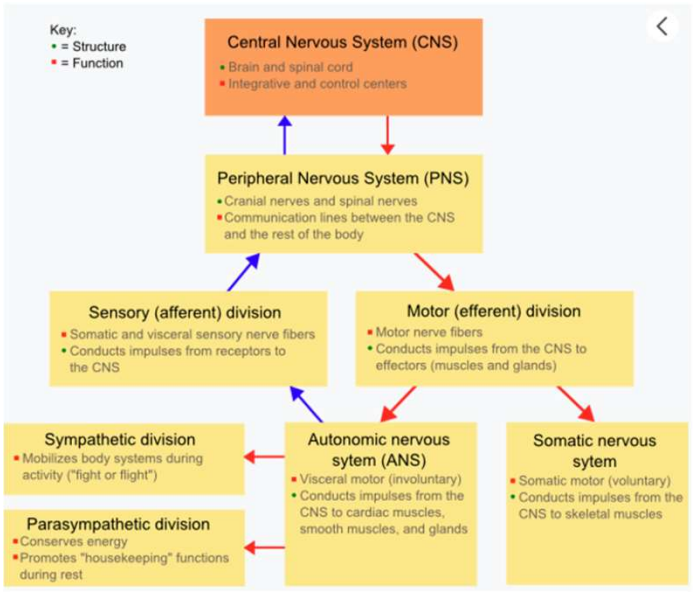
- Paraganglioma is a tumor that forms near the:
  A. Central Nervous System
  B. Somatic Nervous System
  C. Autonomic Nervous System
  D. All of the above

80

Paraganglioma

- Paraganglioma is a type of neuroendocrine tumor that forms near certain blood vessels and nerves outside of the adrenal glands. The adrenal glands are important for making hormones that control many functions in the body and are located on top of the kidneys. The nerve cells involved in paraganglioma are part of the peripheral nervous system, meaning the part of the nervous system outside of the brain and spinal cord. These tumors can also be called extra-adrenal pheochromocytomas.
- Approximately 35-50% of paragangliomas may spread to other parts of the body.
- Paraganglioma – NCI

81



Note:

- Paragangliomas can be either **sympathetic** (responsible for fight or flight) or **parasympathetic** (helps the body relax and digest food).
- Both are part of the autonomic nervous system (regulates involuntary processes like heart rate, BP, breathing, etc)

82

Paraganglioma

- Ganglia are nerve cells outside of the brain/spinal cord
- -oma is a tumor
- Para means close or next to

Tumor next to nerve cells (outside of the brain/spinal cord)

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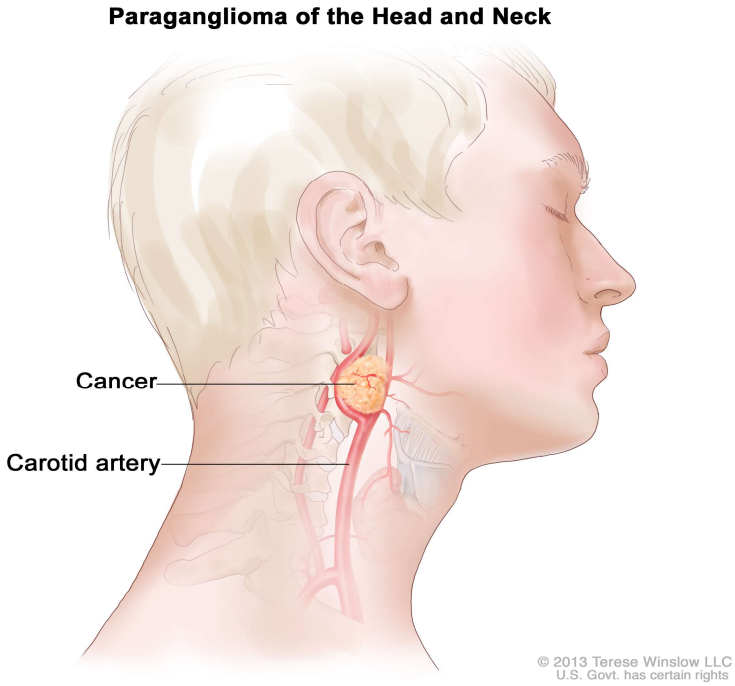
Jugulotympanic PG



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STM – Head/Neck (12/09/2024)

Table 9: Paraganglioma of Carotid Body, Extra-adrenal, Larynx, Middle Ear, Vagal Nerve

Specific or NOS Term and Code	ICD-O Code DX prior to 1/1/2021 *Must be stated to be malignant*	ICD-O Code DX 1/1/2021 forward *"Malignant" no longer required to assign /3*	Synonyms (Per ICD-O-3.2)
Aortic body paraganglioma (C75.5)	8691/3	8691/3	Aortic body tumor Aorticopulmonary paraganglioma
Carotid body paraganglioma (C75.4)	8692/3	8692/3	Carotid body tumor
Extra-Adrenal paraganglioma, NOS	8693/3	8693/3	Nonchromaffin paraganglioma, NOS Chemodectoma Composite paraganglioma
Laryngeal paraganglioma	8690/3	8693/3	
Middle ear paraganglioma (C75.5)	8690/3	8690/3	Glomus jugulare tumor Jugular Jugulotympanic paraganglioma
Paraganglioma, NOS	8680/3	8680/3	
Parasympathetic paraganglioma	8682/3	8682/3	
Sympathetic paraganglioma	8681/3	8681/3	
Vagal paraganglioma *Note:* Vagal paraganglioma has the same histology code as laryngeal paraganglioma. Extra-adrenal, laryngeal and vagal are in separate rows to emphasize primary site.	8690/3	8693/3	

Cases diagnosed prior to 1/1/2021:
Only report these neoplasms when the pathology/tissue specifies malignant (/3) behavior. Change the behavior using ICD-O-3 Rule F Matrix Concept.

Cases diagnosed 1/1/2021 forward:
The term "malignant" is no longer required to assign malignant (/3) behavior. Paragangliomas diagnosed 1/1/2021 or after are malignant unless otherwise stated by the pathologist.

Coding Primary Site:
Paragangliomas have a separate chapter in the WHO Classification of Head and Neck Tumors which is why they are included in the Head and Neck Solid Tumor Rules. Some variants of paraganglioma are specific to certain sites but may occur in sites other than the nervous system. Variants that have specific sites are noted with the appropriate C-code in Table 9. Always code the site noted by the physician. If site is not stated or unclear and histology term does not have a specific site noted in Table 9, code to autonomic nervous system **C479**.

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SINQ discussion about primary site (20210003)

- The instructions for Table 9 of the Head and Neck Solid Tumor Rules instruct registrars to code the primary site to C479 (Autonomic nervous system) for *paragangliomas* that arise in the head and neck region, but the ICD-O-3.2 provides a site-associated code for most of these tumors (C754, Carotid body and C755, Paraganglion). Which primary site is correct? See Discussion.

- While we recognize that *paragangliomas* originate in the parasympathetic or sympathetic nervous system, these are endocrine tumors and endocrine glands/structures are not included in ICD-O site code C479 (Autonomic nervous system). Endocrine tumors of the paraganglia have their own site codes (C75_) per the ICD-O. Additionally, the ICD-O-3.2 provides specific sites for most of the *paragangliomas* included in Table 9. Per the ICD-O-3.2, carotid body *paraganglioma* is C754, and middle ear *paraganglioma*, glomus jugulare tumor, jugulotympanic *paraganglioma*, and *paraganglioma* (NOS) are C755.

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ICD-O-3.0 (purple book)

C75.4 Carotid body

C75.5 Aortic body and other paraganglia

Coccygeal body
Coccygeal glomus
Glomus jugulare
Para-aortic body
    Organ of Zuckerkandl
Paraganglion

CT: Right jugular foramen

MRI: Right jugular foramen c/w glomus jugular paraganglioma

Surgery: Jugulotympanic

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Surgery versus biopsy:

- Operation Excision of right external auditory canal tissue lesion –
- First piece of tumor for frozen section, second piece retained as a specimen, debris removed from around the edges of the tumor.
- Radiation oncology consultation – patient with a biopsy of the lesion confirming the diagnosis of paraganglioma.

- Specimen: A. Right ear mass; B. Right ear canal skin; C. Additional right ear mass
- GROSS:
- A. Specimen A labeled right ear mass is received fresh and consists of a 1 cm tan-brown segment of soft tissue, which is submitted in toto for frozen section purposes (FS1).
- B. Specimen B labeled right ear canal skin is received in formalin and consists of 0.9 x 0.4 x 0.2 cm tan-gray irregular to elliptical segment of skin. The margins are inked blue. The apical ends are submitted (1) with the cut surface en face and the center is submitted (2).
- C. Specimen C labeled additional right ear mass is received in formalin and consists of four tan segments of soft tissue aggregating 1
- x 1 x 0.3 cm. The specimen is submitted in toto (1).

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Purpose of the procedure

Surgical Diagnostic and Staging Procedure

- Record the type of procedure performed as part of the initial diagnosis and workup, whether this is done at your institution or another facility.
- If there are macroscopic positive margins (visible with the naked eye) then capture the procedure as a Surgical Diagnostic and Staging Procedure

Indications for surgery (on the operative report)

- Recommendation was made for treatment, with options of radiation to arrest the growth of the tumor as well as surgery to remove the tumor. Importantly, curative surgical excision has a moderate risk of bleeding, voice and swallowing problems. Subtotal tumor excision, although an available option to reduce the risk of morbidity, necessitates residual tumor surveillance and likely additional treatment;
- furthermore it was felt by radiation oncology that the jugular foramen disease alone would be too large for single-fraction SRS, and other radiotherapy techniques would have undesirable radiation exposure.
- Her candidacy for proton therapy as a definitive treatment is attractive due to the high radiosensitivity of these tumors and her high functional status.
- Consequently, recommended excision of external auditory canal portion of the lesion, possibly tympanotomy if necessary, to obtain a histologic diagnosis and proceed with treatment planning. The risks of bleeding, infection, and additional procedures were discussed. She expressed an understanding of her options and the recommendations and agreed to proceed with the procedure.

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Treatment

- The patient is a 25 y.o. female with a Paraganglioma of the Right Face and Neck. Biopsy of the lesion in the Right External Ear confirmed the diagnosis. The patient was referred for consultation regarding radiation therapy treatment options. After discussion, she consented to definitive proton therapy.

  The patient was treated with proton therapy to a total dose of 54 CGE in 30 fractions. She tolerated treatment well without any Grade 2 or higher toxicities

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Pop Quiz 7

What Does CGE Stand For:

A. Cobalt Gray Equivalents
B. CentiGray Equivalents
C. Computable Gray Equivalents
D. Czech Gray Equivalents

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Pop Quiz 7 (answer)

What Does CGE Stand For:

A. Cobalt Gray Equivalents – Units used for proton therapy
B. CentiGray Equivalents
C. Computable Gray Equivalents
D. Czech Gray Equivalents

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Clinical Staging – Initial thoughts

- Radiation Oncologist: Patient with a paraganglioma of the right carotid sheath with extension into the brain/right CPA (cerebellopontine angle), s/p 54 CGE in 30 fractions
- Enhancing mass (3.1 x 2.0) of mixed-signal intensity centered in an expanded right jugular foramen with extension into the middle ear cavity and external auditory canal as well as intracranially dural extra-axial. In correlation to the CT there are associated permeative bony destructive changes present.
- Tumor is <5 cm but invades surrounding tissue (cT3)
- No regional nodal involvement. (cN0)
- No distant mets (cM0)
- Stage 3
- SS2018:
- Per AJCC, pathologic classification requires total tumor removal.

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Always read additional notes under the staging tables

- Parasympathetic paragangliomas are normally in the head/neck region and are nonfunctional
- These are not staged as they are largely benign.
- **A clue to look for additional information is if your registry software does not allow staging.

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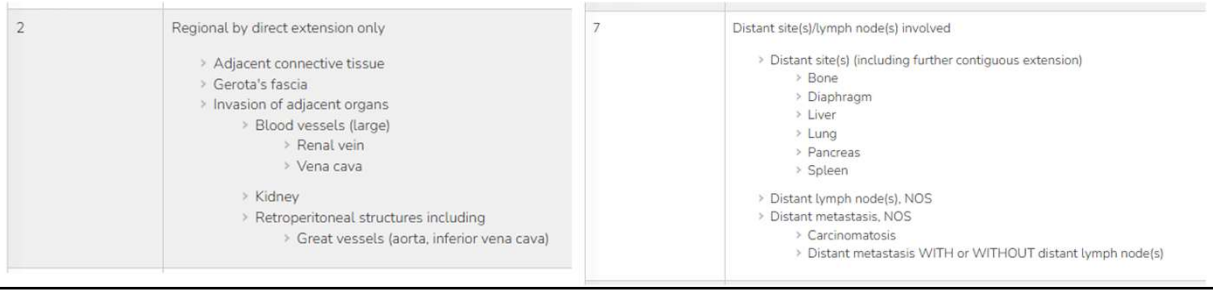
SS2018

Radiation Oncologist: Patient with a paraganglioma of the right carotid sheath with extension into the brain/right CPA (cerebellopontine angle)

Enhancing mass (3.1 x 2.0) of mixed-signal intensity centered in an expanded right jugular foramen with extension into the middle ear cavity and external auditory canal as well as intracranially dural extra-axial. In correlation to the CT there are associated permeative bony destructive changes present.



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Case Scenario 5

Data Item	Value
Primary Site	C75.5
Histology	8690/3
AJCC Clinical	88
AJCC Pathological	88
Grade Clinical	9
Grade Pathological	9
SS2018	2
EOD Tumor	300 (invasion of adjacent)
EOD Nodes	000
EOD Mets	00
No SSDIs	

Data Item	Value
Diagnostic/Stg Procedure	02
Surgery of Primary Site	0
Chemotherapy	0
Immunotherapy	0
Palliative Care	
Primary Treatment Volume	29
Draining Lymph nodes	00
Treatment Modality	03
EB Planning Technique	04
Dose per Fraction	00180
Number of Fractions	030
Total Dose	005400

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Gastrointestinal Stromal Tumor (GIST)

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What is GIST?

- Rare type of soft tissue sarcoma
  - Develop in muscle layer of gut rather than mucosa
  - Grow outward (exophytic)
- Median age: 65
- Male to female ratio: 1:1
- Often incidental finding

- Gastrointestinal stromal tumor 8936
  - GANT
  - GIST, spindle cell type
  - Gastrointestinal pacemaker cell tumor
  - Gastrointestinal stromal tumor
  - GIST NOS
  - GIST, malignant
  - Gastrointestinal stromal sarcoma
  - Succinate dehydrogenase-deficient
  - gastrointestinal stromal tumor

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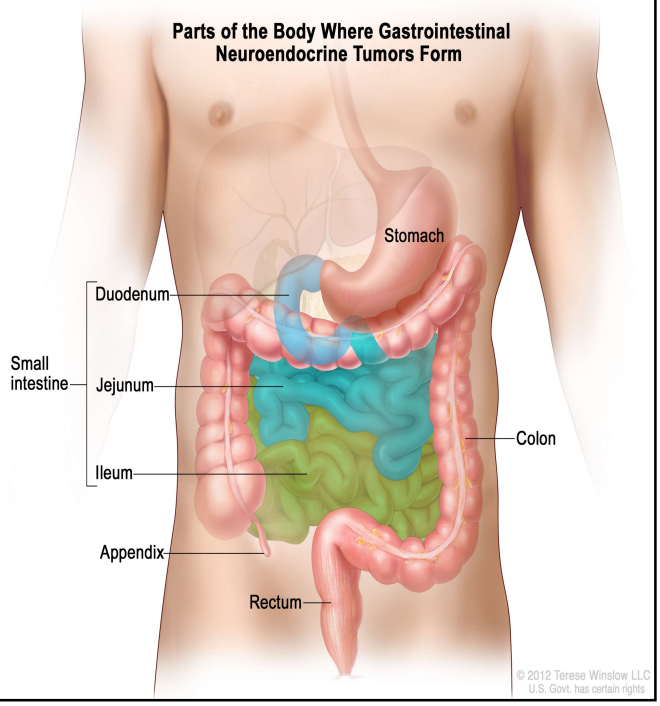
Tumor location

Stomach	56%
Small Intestine	32%
Colon/Rectum	6%
Esophagus	0.7%
Other	5.5%

- GISTs occurring outside of the stomach are associated with higher malignant potential
- Exophytic growth noted in 79% of GIST
- Intraluminal or mixed growth is less frequent

https://pmc.ncbi.nlm.nih.gov/articles/PMC6351301/



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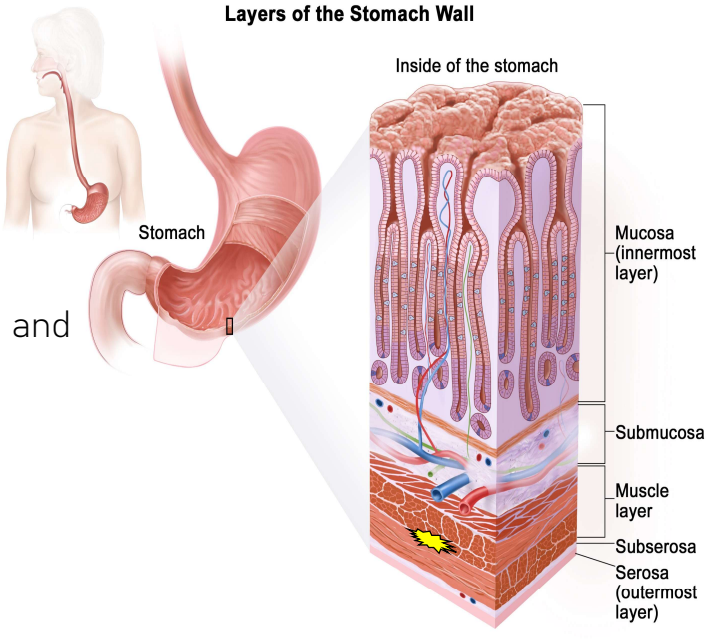
GIST

- Interstitial cells of Cajal
  - "Pacemaker cells"
  - Sends signals to move food and liquid through system (peristalsis)



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Oncogenic mutations

- ~85% of GIST contain oncogenic mutations in one of two receptor tyrosine kinases
  - KIT-Mutant GIST (80%)
    - Mutations of exon 9, 13, and 14 – sunitnib
    - Mutations of exon 18-avapritinib
  - PDGFRA (Platelet-derived Growth Factor Receptor Alpha) (8-10%)
- Wild Type GIST (KIT/PDGFRA WT) 8-10%
  - GIST contain no genetic mutation of KIT or PDGFRA

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Reportability of GIST, NOS

- Benign GIST has ***never*** been reportable.
- Malignant GIST has ***always*** been reportable
  - Statements indicating a GIST, nos is malignant.
    - Positive lymph nodes
    - Metastasis
    - Multiple foci
    - Invasion

- Gastrointestinal stromal tumor, NOS
  - 8936/1
    - Cases diagnosed prior to 2021 (not reportable)
  - 8936/3
    - Cases diagnosed 2021 or later (reportable)

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Grade

- Mitotic Rate
  - Mitotic rate should be based on mitosis per 5 square mm
  - Is a factor when assigning AJCC stage

Code	Description
L	Low: 5 or fewer mitoses per 5 square mm
H	High: Over 5 mitoses per 5 square mm
A	Well differentiated
B	Moderately differentiated
C	Poorly differentiated
D	Undifferentiated, anaplastic
9	Grade cannot be assessed; Unknown

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Stage

- AJCC
  - Size divisions-2cm, 5cm, 10cm
  - cN may be used when assigning pStage
  - Mitotic rate is a factor when assigning stage.
  - Different stage tables based on location of primary tumor.
- Summary Stage
- EOD

SS2018	Description
0	**In situ, intraepithelial, noninvasive**
1	**Localized only (localized, NOS)** › Confined to site of origin
2	**Regional by direct extension only** › Adjacent (connective) tissue, NOS
3	**Regional lymph node(s) involved only** › Regional lymph node(s), NOS › Lymph node(s), NOS
4	**Regional by BOTH direct extension AND regional lymph node(s) involved** › Codes (2) + (3)
7	**Distant site(s)/lymph node(s) involved** › Distant site(s) (including further contiguous extension) › Adherent to organs/structure, NOS › Extension to organs/structures, NOS › Liver parenchymal nodules › Peritoneal nodules

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Treatment

- Surgery
  - Tumors greater than 2cm-Resection of primary tumor without rupturing
  - Tumors less than 2cm without high-risk features-surveillance
- Systemic-TKI inhibitors
  - Imatinib
  - Sunitinib
  - Avapritinib

Codes

A000 None; no surgery of primary site; autopsy ONLY

A100 Local tumor destruction, NOS
- A110 Photodynamic therapy (PDT)
- A120 Electrocautery; fulguration (includes use of hot forceps for
- A130 Cryosurgery
- A140 Laser

No specimen sent to pathology from surgical events A100–A140

A200 Local tumor excision, NOS
- A260 Polypectomy
- A270 Excisional biopsy

Any combination of A200, A260, or A270 WITH
- A210 Photodynamic therapy (PDT)
- A220 Electrocautery
- A230 Cryosurgery
- A240 Laser ablation
- A250 Laser excision

A300 Gastrectomy, NOS (partial, subtotal, hemi-)
- A310 Antrectomy, lower (distal-less than 40% of stomach)***
- A320 Lower (distal) gastrectomy (partial, subtotal, hemi-)
- A330 Upper (proximal) gastrectomy (partial, subtotal, hemi-)

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Case Scenario 6

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Case Scenario 6

Data Item	Value
Primary Site	C17.1 Jejunum
Histology	8936/3
Grade Clin	9
Grade Path	L- 5 or less/5mm^2
Diagnostic Confirmation	1
Tumor Size Summary	064

Data Item	Value
AJCC Clin	cT3 cN0 cM0 MR 9 Stage 99
AJCC Path	pT3 pN0 cM0 MR L Stage 2
Summary Stage 2018	2 Regional
EOD Prim Tumor	400
EOD LN	000
EOD Mets	00
KIT Gene IHC	7

Data Item	Value
Diagnostic/Stage Proc	00
Surgery Prim Site 2023	A300 Partial Resection of primary site
Chemotherapy	02

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Coming Up!

- Boot Camp 2025
  - *Nancy Etzold, ODS*
  - Sarah Burton, ODS
  - *3/12/25 and 3/13/25*

Date change!
Second week of March!

- Treatment 2025
  - Wilson Apollo, ODS
  - 4/3/25

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Fabulous Prizes

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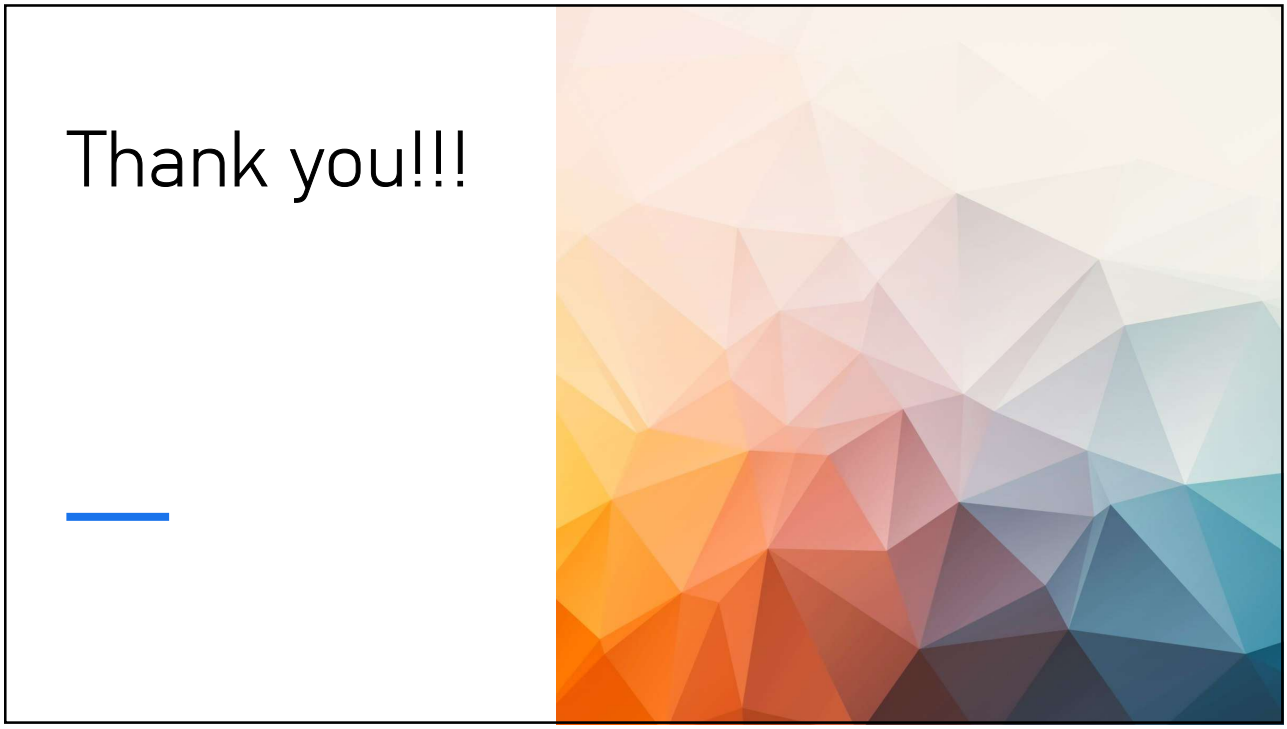

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