Unusual Site2 2025 Case Scenarios

Case #1 Mesothelioma

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

7/7/24-PROCEDURE: 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 thorascopic camera. There were obvious metastatic nodules on the 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.

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

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.

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.

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.

CASE #2 Retinoblastoma

1/12/25

The patient presents now at 3 weeks of age with family history of retinoblastoma in his older brother. His parents are aware of the potential for familial transmission and the patient is brought to the ocular oncology clinic for comprehensive evaluation and during that evaluation a small peripheral lesion consistent with retinoblastoma was evaluated and documented. Because of the difficulty in evaluating the patient comprehensively he was scheduled for examination under anesthesia with potential tumor laser ablation. Risks, benefits and alternatives were discussed and informed consent was obtained.

PROCEDURE: Exam under anesthesia, and thermal laser ablation left eye

The patient underwent induction of general mask anesthesia without complication. Intraocular pressures at induction were 9 and 10. Lids and lashes were clear sclera and conjunctiva are intact. Corneas are clear. Anterior chamber is deep without cell or flare. Iris is flat without rubeosis or mass and lens reveals clarity OU 360 degree comprehensive dilated fundus examination was performed with scleral depression to both eyes the right eye showed small cup-to-disc Goodrum coloration and perfusion. Minimal vascular ectasia was noted. There was no evidence of retinal tumor or retinoblastoma vitreous subretinal seeding. There was no open hole to break epiretinal membrane or PVR.

Formation evaluation of the left eye showed small cup-to-disc good rim coloration and perfusion. An inferotemporal small white retinoblastoma lesion was present in the retina. Maximum dimension was 2mm. The lesion was located in excess of 4mm from the disc and fovea. There was no evidence of exudative retinal detachment, vitreous or subretinal seeding, open hole tear, or break.

I was able to clearly evaluate both eyes and with a unilateral unifocal lesion with known family history I proceeded to apply large spot size transpupillary thermal laser ablation confluently over the tumor surface. This was accomplished under direct visualization without complication. Excellent laser ablation was applied without difficulty.

Impression: The patient presents with unilateral familial unifocal retinoblastoma presenting within the first month of life. This early detection has given the patient an option to avoid chemotherapeutic requirements. I discussed this extensively with the parents and will reassess in 3 to 4 weeks with examination under anesthesia. There is significant potential for new tumor development in this or the second eye. We will coordinate ongoing follow-up and serial review. On follow-up will recommend MRI imaging in a nonemergent fashion

Patient has had an outstanding response to transpupillary thermal laser ablation at this point it seems likely that he will avoid the need for ongoing treatment progression to systemic or focal chemotherapy. I will continue to see this patient on a monthly basis for a minimum of 6 months. With early recognition and ablative therapy this patient has avoided the need for systemic chemotherapy or other alternative treatment approaches.

The patient presents with clinical RB1 unilateral unifocal retinoblastoma OS due to the early presentation recognitions under avoided systemic or focal chemotherapy will coordinate targeted follow-up and serial review all returns under to clinic and will continue to use the operative environment for comprehensive evaluation especially targeted to the far periphery

Case #3 Ocular Adnexa Lymphoma

4/24/24 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

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.

The patient has 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 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.

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

PET Scan: Intense FDG activity corresponding to an approximate 3 cm right supraorbital soft tissue mass compatible with the patient's history of orbital lymphoma. This is an isolated lesion in this patient. 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
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.

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

Case #4 Thymoma

1/7/2025

A 65 year old patient with a past medical history of significant coronary artery disease from hypercholesterolemia was undergoing a CT coronary screening and was incidentally discovered to have an anterior mediastinal mass. A subsequent PET scan showed the mass to be metabolically active with an SUV of 2.6. The patient was recommended to undergo a right robotic video-assisted complete thymectomy for diagnosis and curative intent.

- CT scan: Anterior mediastinal nodule measuring 2 x 1.1 cm possibly related to low-grade thymic neoplasm. This can be followed with CT chest or further evaluated with PET/CT.
- PET Scan: There are no suspicious FDG avid pulmonary nodules or masses. There is a 1.5 x 1.0 cm cm soft tissue nodule in the anterior mediastinum, anterior to the SVC, with mild FDG uptake, SUV max of 2.6. There is mild FDG uptake in right axillary lymph nodes, likely lymphatic uptake segment extravasation.

1/14/2025

Operative Procedure Performed:

1). Right robotic assisted thoracoscopy
2). Complete thymectomy

Operative Procedure Technique: We assessed the mass in the anterior mediastinum that was approximately 5 cm in diameter. We began by freeing up the thymus and fat along the inferior border of the pericardium. We then freed the thymus off the anterior border of the pericardium over to the left phrenic nerve. We then carefully dissected along the left phrenic nerve from inferior to superiorly visualizing it the whole time while freeing up the remaining thymus from the left phrenic nerve.

Pathology Report:

* SYNOPTIC REPORT:	Thymus
* Procedure:	Thymectomy
* Tumor size:	2.5 cm
* Type:	Type AB thymoma
* Extent:	Limited to thymus
* Margins:	Free
* Treatment effect, primary site:	Not applicable
* Lymphovascular invasion:	None
* Lymph nodes, # total:	0
* Lymph nodes, # involved:	Not applicable
* Distant metastases:	Not applicable
* Stage (AJCC 8):	p T1a N (not assigned) M (not applicable)
* Tumor block(s) for possible future studies:	Not applicable

Follow-up Appointment: The patient is doing well from a surgical standpoint. We discussed the results of her recent chest CT. We discussed that there is no evidence of recurrent or metastatic disease. If all continues to be stable, we will then begin to monitor on an annual basis.

Case #5 Paraganglioma

9/13/24-22 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.

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.

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.

FINAL DIAGNOSIS: A. Right ear canal mass: Paraganglioma

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

Case #6 GIST

7/14/24

54-year-old married white Hispanic male with past medical history of hypertension. Patient presents to the ER with abdominal pain and undergoes CT scan showing mass in hemiabdomen. Patient reports fever and LLQ abdominal pain. Exam shows normal abdomen with left lower quadrant tenderness and no peritoneal signs. Patient underwent PET scan and met with surgery and oncology and was recommended to proceed with surgical resection and patient agreed.

CT abdomen/pelvis – IMPRESSION: 1. Almost 6 cm enhancing mass (5.1 x 4.3 x 5.9 cm) in the mid left hemiabdomen mesenteric contiguous with adjacent small bowel loops is concerning for neoplasm. *Differential includes* small bowel adenocarcinoma, lymphoma, or GIST. Surgical consultation recommended. 2. There is fat stranding around the mass suggesting inflammation. Mild upstream air and fluid filled dilation of small bowel loops raises possibility of partial obstruction secondary to mass.

PET Scan – Impression: 1. 5.8 cm FDG avid mass in the left midabdomen contiguous and involving the small bowel loops is suggestive of a malignant etiology, differential considerations include small bowel adenocarcinoma, lymphoma, or GIST. Recommend tissue sampling. 2. Borderline increased FDG uptake in the marrow may be reactive/physiologic.

Enterography MRI – Impression: 1. Enhancing soft tissue mass 6.4 x 5.7 x 7.1 with areas of peripheral necrosis located in the mesentery, arising from or encasing a segment of small bowel loop of jejunum leading to partial small bowel obstruction. (Enhancing component approximately 4.8 cm) The mass is most suspicious for neoplasm. Differential includes a gastrointestinal stromal tumor; other possibilities including small bowel lymphoma, neuroendocrine tumor. Tissue sampling recommended. 2. No evidence of mesenteric or retroperitoneal lymphadenopathy.

Operative Procedure Performed:

- Robotic small bowel resection
- Robotic resection of mesenteric mass
- Robotic drainage of intra-abdominal abscess

Operative Procedure Findings: On exploration of the bowel on the left side of the abdomen. Using suction and separating the bowel from the omentum there was spontaneous rupture and pus was encountered. It was suctioned. Cultures were obtained. There was friable bleeding of the mass. At this point the small bowel was identified in the mesenteric and small bowel mass was identified.. The small bowel was transected proximal and distal to the small bowel mass. The anastomosis was performed in a side-to-side functional end-to-end fashion.

DIAGNOSIS:

A. Anastomotic remnants
Benign small intestinal tissue.

B. Small bowel and mesenteric mass
Gastrointestinal stomal tumor, see table.
One benign lymph node.
Desmin and S100 are negative.

* SYNOPTIC REPORT:	GIST
* Procedure:	Resection
* Tumor location:	Jejunum
* Tumor size:	6.4 cm
* Tumor focality:	Single focus
* Multiple primary sites:	Not applicable
* Subtype:	Spindle cell
* Mitotic rate:	5 or less/ 5 mm squared
* Grade:	Low grade
* Risk assessment:	Moderate risk
* Margins:	Free
* Treatment Effect :	Not applicable
* Distant metastases:	Not applicable
* Stage (AJCC 8):	p T3 N (not assigned) M (not applicable)
* KIT(CD117) Immunohistochemistry:	Positive
* DOG1 Immunohistochemistry:	Positive
* SDHB Immunohistochemistry:	Not applicable
* SDHA Immunohistochemistry:	Not applicable
* Tumor block(s) for possible future studies:	B8

Gross Description

B. Specimen labeled small bowel and mesenteric mass is received in formalin, excision and formalin time 2012/0820, cold ischemic time 12-13 hours and consists of an unoriented segment of bowel, 8.2 cm in length, 3.2 cm in diameter with two stapled resection margins. The serosal surface is tan-pink, smooth with a bulging disrupted pink-tan hemorrhagic friable mass, 6.4 x 3.9 x 3.7 cm. The mass comes within 1.7 cm and 3.9 cm of the unoriented resection margins. The mass grossly appears to involve the serosa and attached mesentery. The mesenteric margin is inked black and the mass is inked blue. The mass comes within 1.4 cm of the mesenteric margin. The bowel is opened revealing tan-pink, and glistening mucosa with a 0.5 cm defect, which appears contiguous with the aforementioned mass. The mass is sectioned revealing tan-white, soft, focally hemorrhagic cut surfaces. The surrounding soft tissue is palpable for one lymph node, 0.4 x 0.4 x 0.3 cm. The specimen is submitted:

He is healing well postoperatively. ECOG of 0. We discussed starting adjuvant treatment with Gleevec. Reviewed side effects. However we are pending molecular analysis of tumor. Patient is at an intermediate risk given that it was a mesenteric mass with a mitotic rate less than 5 per high-power field and with a tumor size of 6.4 cm.

Following molecular analysis, patient was started on Gleevec to continue for 2 years.