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Toddler with fever

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Case Presentation

HPI:

21 m.o. F with no prior medical history presents to the ED with 9 days of persistent fevers and reported T-max of 40.5 C. She has decreased energy, oral intake and urinary output, but normal bowel movements. She has not had any skin changes over her course. Parents do report a prior respiratory illness 3 weeks prior to presentation. Her family lives on a farm and interacts with a variety of animals.

OTHER RELEVANT HISTORY:

- No prior medical, surgical or family history
- No daily medications
- No allergies
- Up to date on vaccinations

Case Presentation (cont.)

VITALS:

BP 114/40, Pulse 98, Temp 36.4, RR 22, SpO2 98% RA

PHYSICAL EXAM:

- Constitutional: Active and not in acute distress.
- HEENT: No discharge or mucosal erythema. **Cervical lymphadenopathy** present.
- Cardiovascular: Regular rate and rhythm. **Tachycardic** with normal pulses.
- Pulmonary: Normal effort and breath sounds. No wheezing or rales.
- Abdominal: Normal bowel sounds. **Distension** but no tenderness.
- Skin: Warm and dry. No rashes or lesions.
- Neurological: No weakness. Normal reflexes.

Case Presentation (cont.)

PERTINENT LABS:

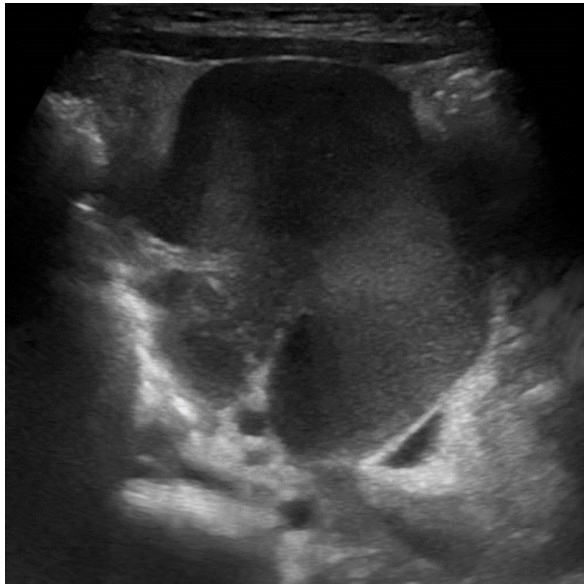
- CMP: Normal
- CBC: WBC 10.72, **Hgb 7.4**, **Plt 897**
- CRP: **176.9**
- ESR: **>130**

DIAGNOSTICS:

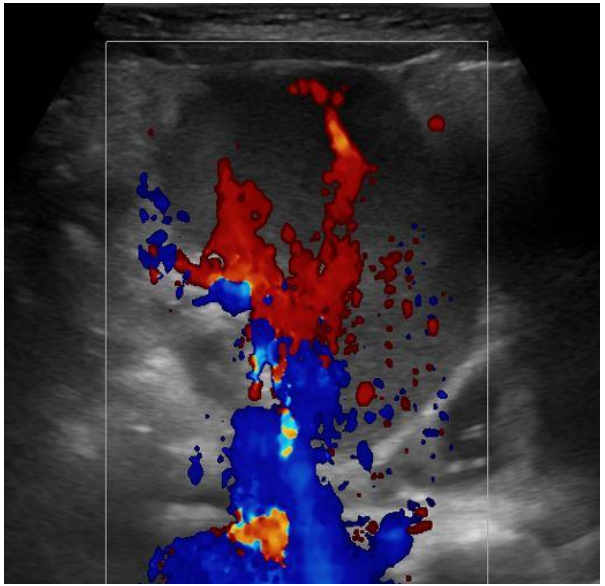
- ECHO: Normal
- Abdominal ultrasound because of abdominal distension and persistent fever

Ultrasound

- Well-defined minimally lobulated 4.2 x 4.4 x 4.2 cm mass
  - Uniformly hypoechoic
  - Hyper vascular on color Doppler



RLQ US



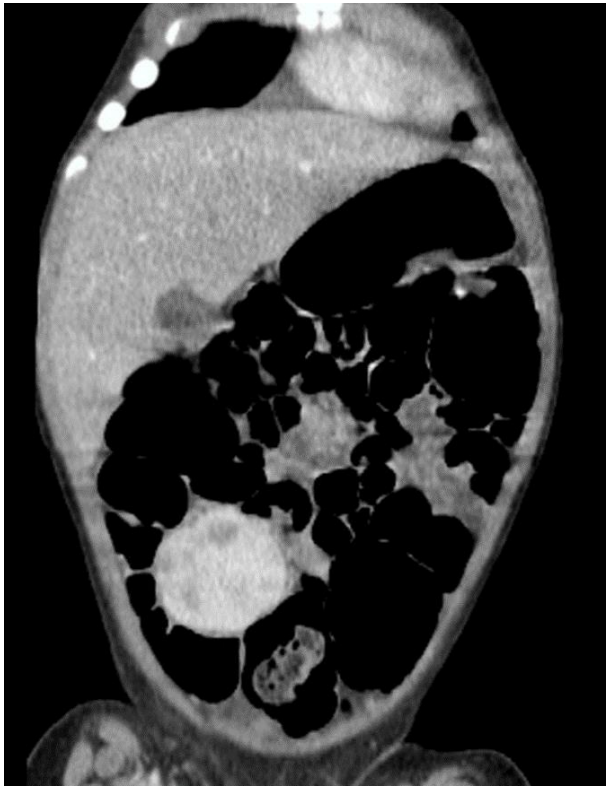
RLQ US with Doppler

Differential Diagnosis

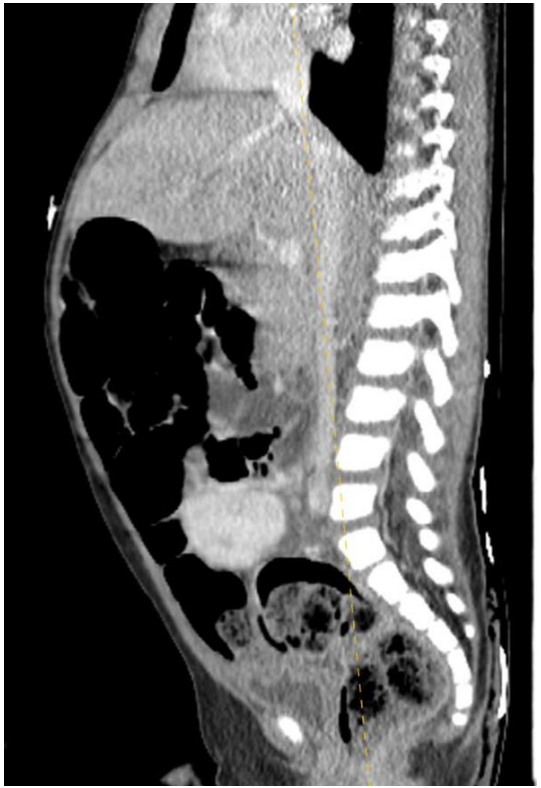
- Leiomyoma
- Gastrointestinal stromal tumor (GIST)
- Peripheral nerve sheath tumor
- Solitary fibrous tumor/fibroid
- Lymphoma/Burkitt lymphoma
- Carcinoma/Sarcoma
- Inflammatory myofibroblast tumor

CT

- Solitary right lower quadrant mesenteric mass
- Circumscribed
- Minimally heterogenous although hyperenhancing relative to liver
- Moderate diffuse distention of bowel but otherwise normal abdominal and pelvic viscera with no ascites.

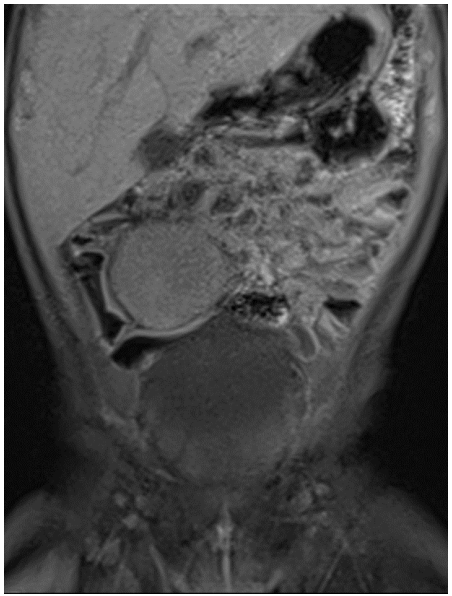


Coronal CT

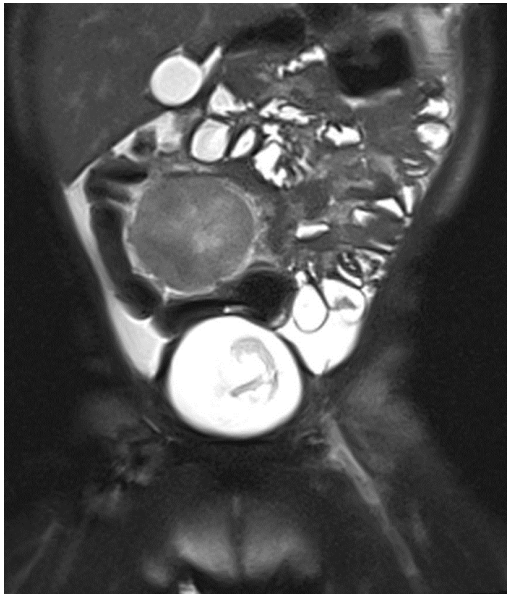


Sagittal CT

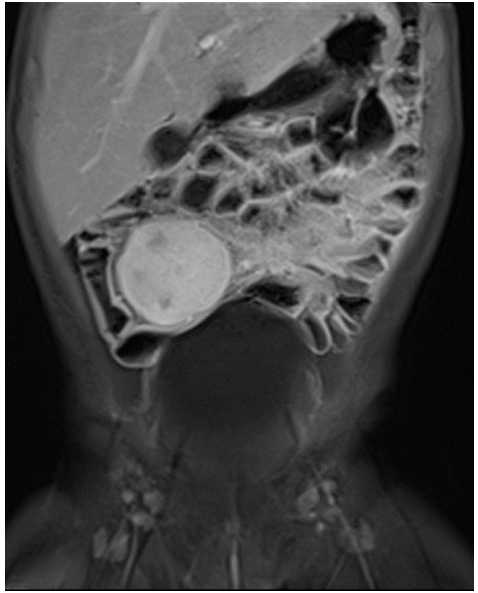
MRI/MRA



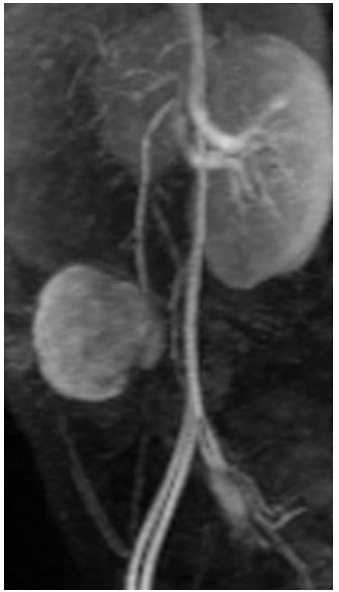
FS T1



T2 HASTE



Post Gadolinium T1



MRA

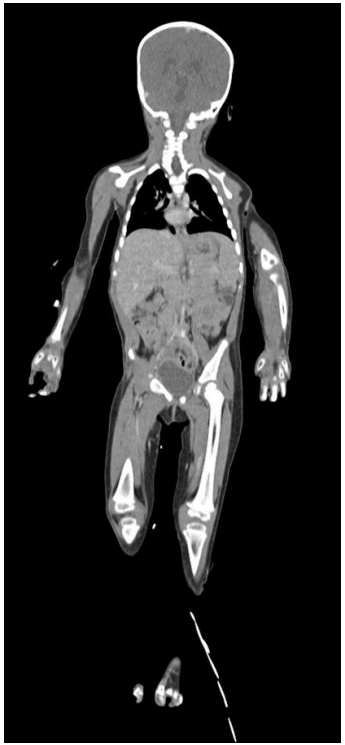
- MRI shows T1 isointense, peripherally T2 hypointense and enhancing mass in RLQ mesentery.
- MRA reveals arterial supply from the SMA.

PET-CT (Post Operative)

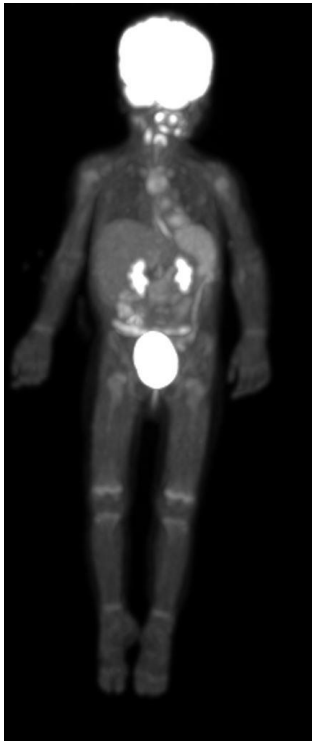
- F18-FDG PET-CT was obtained on POD 15
  - No anatomic residual neoplasm
  - No hypermetabolic residual neoplasm or evidence of synchronous or metastatic disease
- Normal avidity in the esophagus, spleen, ileocecal region and urinary tract.

DIAGNOSIS AND OUTCOME

- Post-op pathology report revealed **inflammatory myofibroblastic tumor**
- Patient has had no other issues and is feeling well at 3 months post-op



CT



FDG-PET

Fused PET-CT

Inflammatory Myofibroblastic Tumor (IMT)

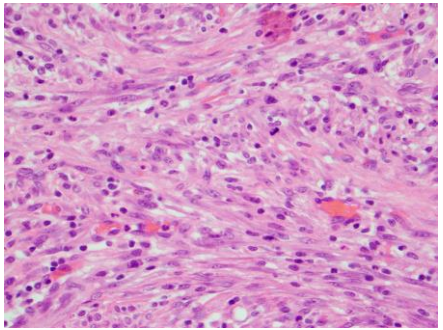
Teaching points

EPIDEMIOLOGY:

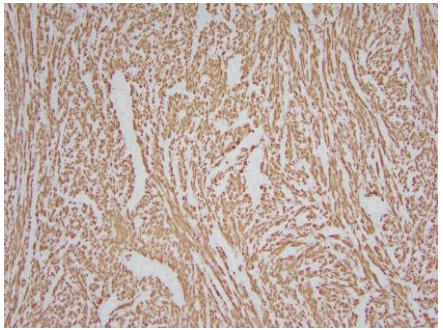
- Approximately 150-200 cases in the US annually[1]
- Typically present in the abdominal cavity in children, with lung and neck also being common[2,3]
- Clinical presentation usually related to mass-effect or non-specific constitutional symptoms
- Common sites: Lung, abdominal visceral, bowel, mesentery and urinary tract
  - Most common primary lung tumor in children

PATHOPHYSIOLOGY:

- Overgrowth of spindle fibroblast cells and inflammatory cells[2,3]
  - ALK gene mutation in 60%
- Unknown etiology but may be associated with chronic infection, autoinflammatory disease, and trauma[2]



Mass image showing spindle cells, mitotic figure and inflammatory cells



Mass image showing positive immunostaining for ALK 1

Inflammatory Myofibroblastic Tumor (IMT)

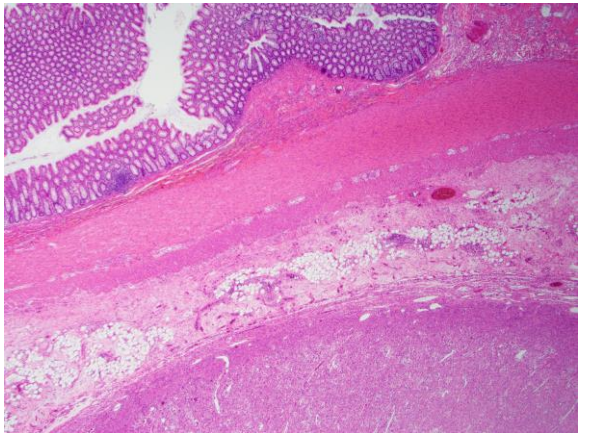
Teaching points (Cont.)

NATURAL HISTORY:

- Tumor will continue to grow
- Symptoms of systemic inflammation
- Untreated IMT will continue to infiltrate and worsen mass effect

TREATMENT:

- Resection is the most common treatment although recurrence is common[2,3]
  - Low risk of distant metastasis or synchronous lesions (~5%)
  - Locoregional recurrence more likely
- Chemotherapy if resection is not achievable
  - Steroids
  - ALK or COX inhibitors



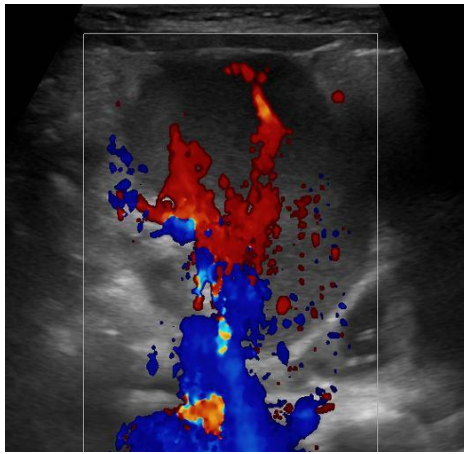
Full section of bowel showing wide margins of excision. Mucosa is seen at the top while the mass is seen within the mesentery below.

Inflammatory Myofibroblastic Tumor (IMT)

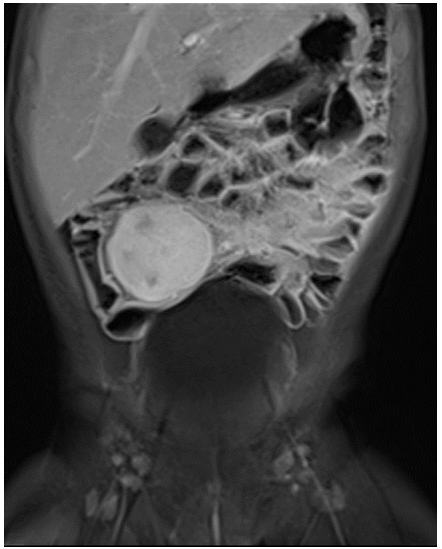
Teaching points (Cont.)

IMAGING:

- General features
  - Typically well-defined and round
    - May infiltrate (especially in the bowel and mesentery)
  - 7-8 cm mean size at time of resection
- US
  - Hypoechoic solid mass
  - Hypervascular on Doppler
- CT/MRI
  - Central necrosis and calcification common
  - T2 hypointensity reflects fibrous stroma
  - Hyperenhancing
- PET
  - Increased glucose uptake



Abdominal US



Post gadolinium T1

Conclusion

- IMTs are known as the great mimickers of neoplasm
- Imaging findings are similar among many abdominal masses in children
  – Thus, IMTs are under reported on radiological differentials
- Clinical correlation with elevated inflammatory markers can be a hint
- IMTs can only be differentiated by pathology
- Treatment options and outcomes are very different between IMTs and most abdominal neoplasms in kids
- It is critical for radiologists to be aware of these mimickers to guide proper disease management

References

1. National Institutes for Health, *Inflammatory Myofibroblastic Tumor (IMT),* National Cancer Institute https://www.cancer.gov/pediatric-adult-rare-tumor/rare-tumors/rare-soft-tissue-tumors/inflammatory-myofibroblastic-tumor February 27th, 2019 (Accessed April 22nd, 2024)

2. Krzysztof Siemion, Joanna Reszec-Gielazyn, Joanna Kisluk, Lukasz Roszkowiak, Jakub Zak, Anna Korzynska, *What do we know about inflammatory myofibroblastic tumors? – A systematic review,* Advances in Medical Sciences, Volume 67, Issue 1, 2022, Pages 129-138, ISSN 1896-1126, https://doi.org/10.1016/j.advms.2022.02.002. (https://www.sciencedirect.com/science/article/pii/S1896112622000062)

3. Cantera, et al. *Inflammatory myofibroblastic tumours: a pictorial review,* Insights into Imaging, 85:96:6:1, https://doi.org/10.1007/s13244-014-0370-0DO, February 2nd, 2015

Thank you

Contact

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