AMSER Rad Path Case of the Month:

Premature neonate with large sacral mass

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Patient Presentation

HPI: Female neonate born at 33 weeks to a 32-year-old G4P1021 woman by c-section delivery due to worsening maternal condition (suspected sepsis), preterm premature rupture of membranes, and management of sacral mass.

Pregnancy complications: 1st trimester teratogen exposure (topiramate), polyhydramnios, large for gestational age fetus, and large fetal sacral mass.

Maternal medical history: History of bipolar 1 disorder treated with topiramate. Pregnancy labs: O+ blood type, RPR neg., HIV neg., GBS +, gonorrhea/chlamydia neg., rubella immune, hepatitis B surface antigen neg.

Maternal social History: Mother reports a 1-year pack history and has not smoked in 15 years. Occasional marijuana use and denies illicit substance use.

Pertinent physical examination findings

APGARS: 1 and 8 at 1 and 10 minutes

Birth weight: 3560g Head circumference: 37.3 cm Birth length: 41.5 cm

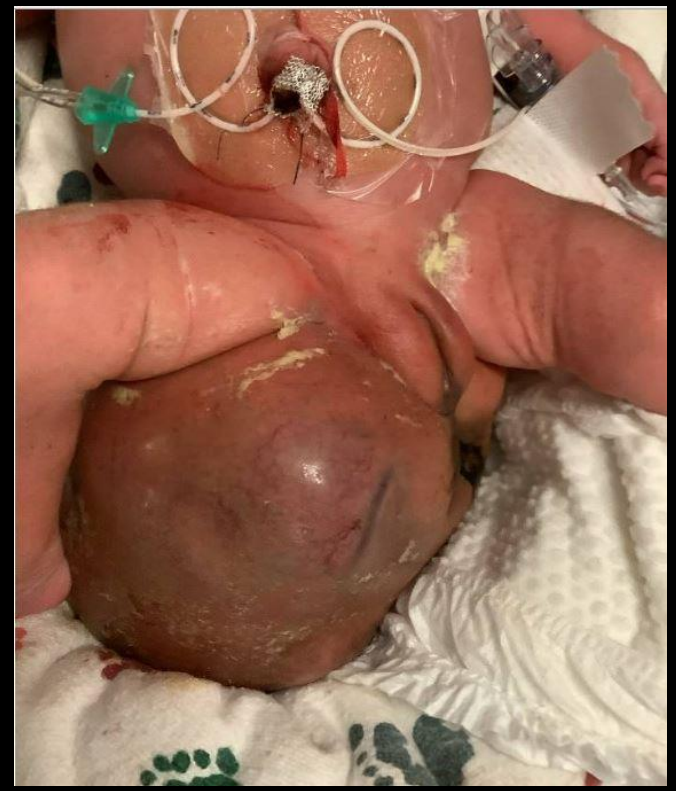
PEX:

General: In mild respiratory distress

Chest: Mild retractions present in the substernal and intercostal areas

Extremities: Large sacral mass with overlying skin intact. No deformities of extremities noted

Neurologic: Decreased tone and activity



What imaging should we order?

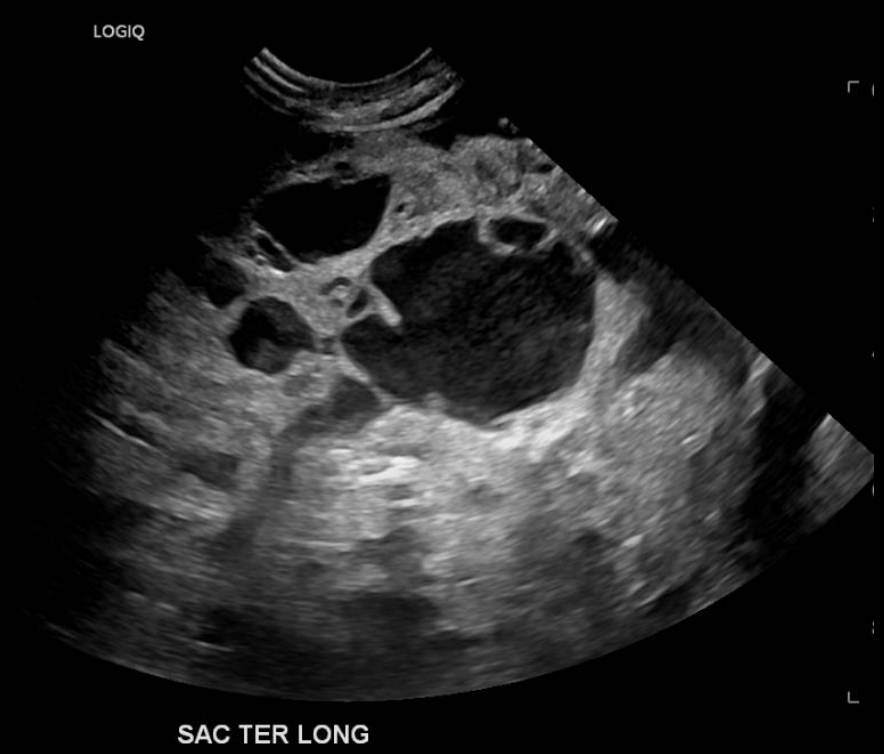
ACR Appropriateness Criteria

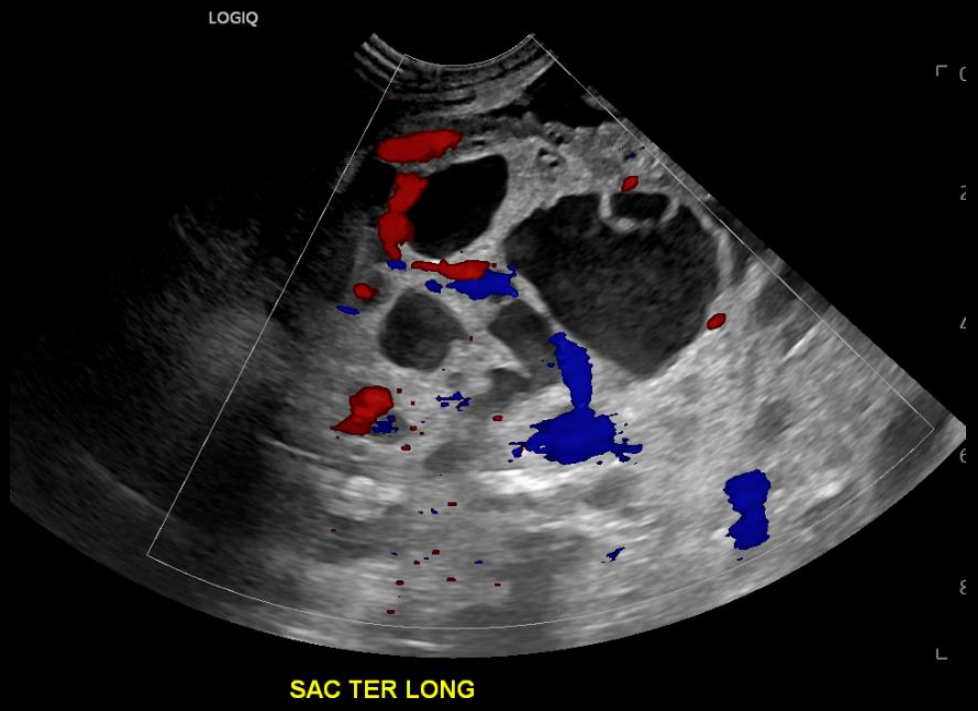
This imaging modality was ordered by the ordering physician

Variant 1: **Superficial soft tissue mass. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
US area of interest	Usually Appropriate	O
Radiography area of interest	Usually Appropriate	Varies
US area of interest with IV contrast	Usually Not Appropriate	O
Image-guided biopsy area of interest	Usually Not Appropriate	Varies
Image-guided fine needle aspiration area of interest	Usually Not Appropriate	Varies
MRI area of interest without and with IV contrast	Usually Not Appropriate	O
MRI area of interest without IV contrast	Usually Not Appropriate	O
FDG-PET/CT area of interest	Usually Not Appropriate	☢☢☢☢
CT area of interest with IV contrast	Usually Not Appropriate	Varies
CT area of interest without and with IV contrast	Usually Not Appropriate	Varies
CT area of interest without IV contrast	Usually Not Appropriate	Varies

ULTRASOUND SOFT TISSUE SACRAL REGION





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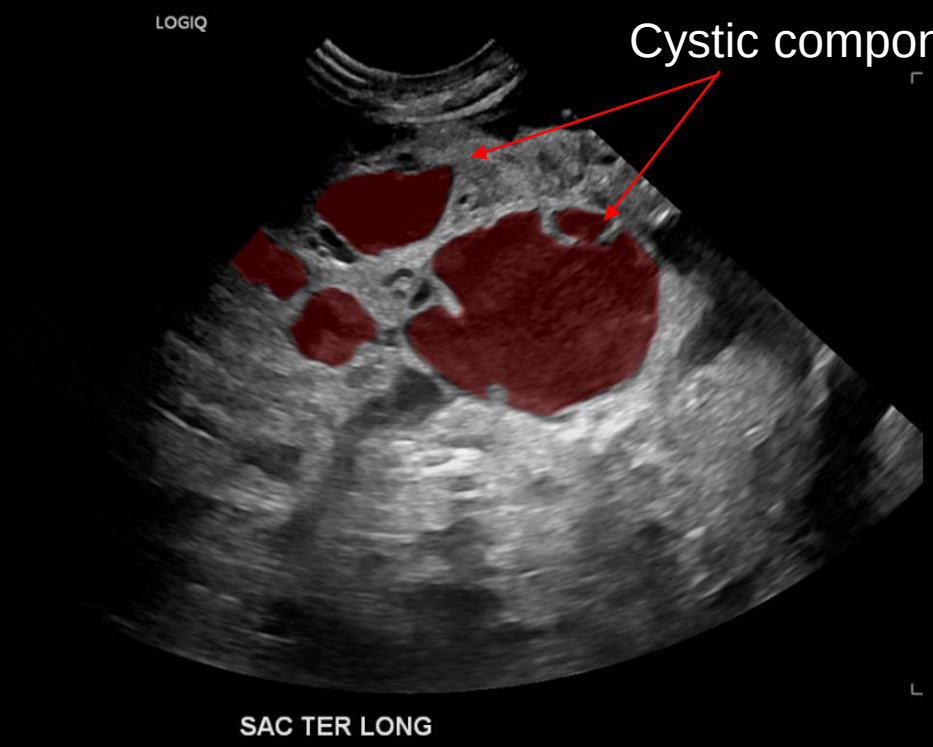
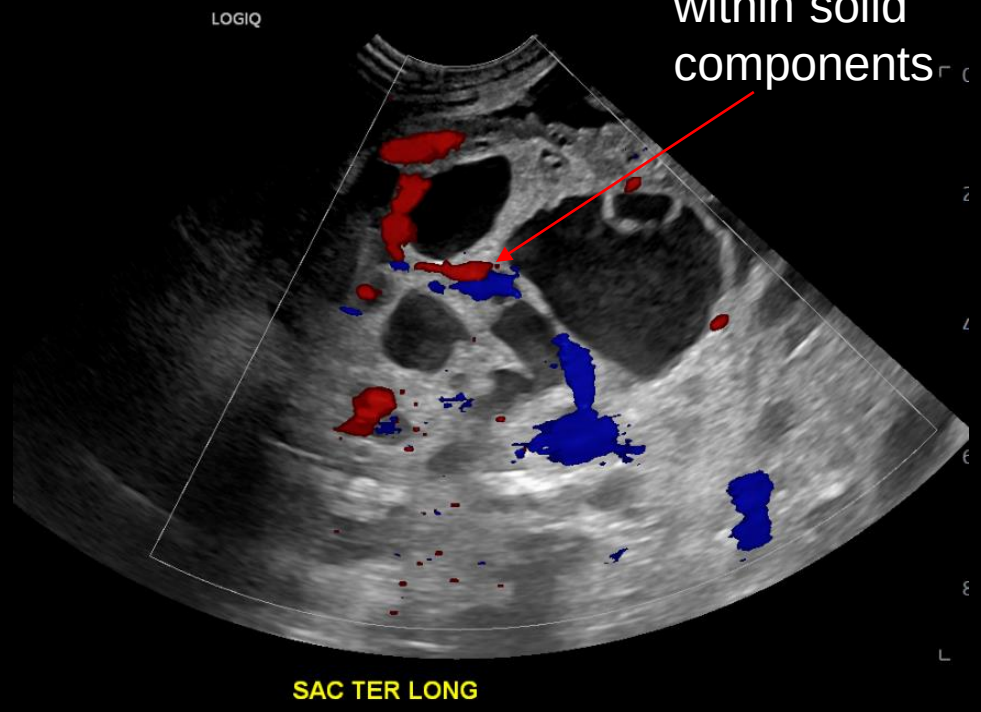
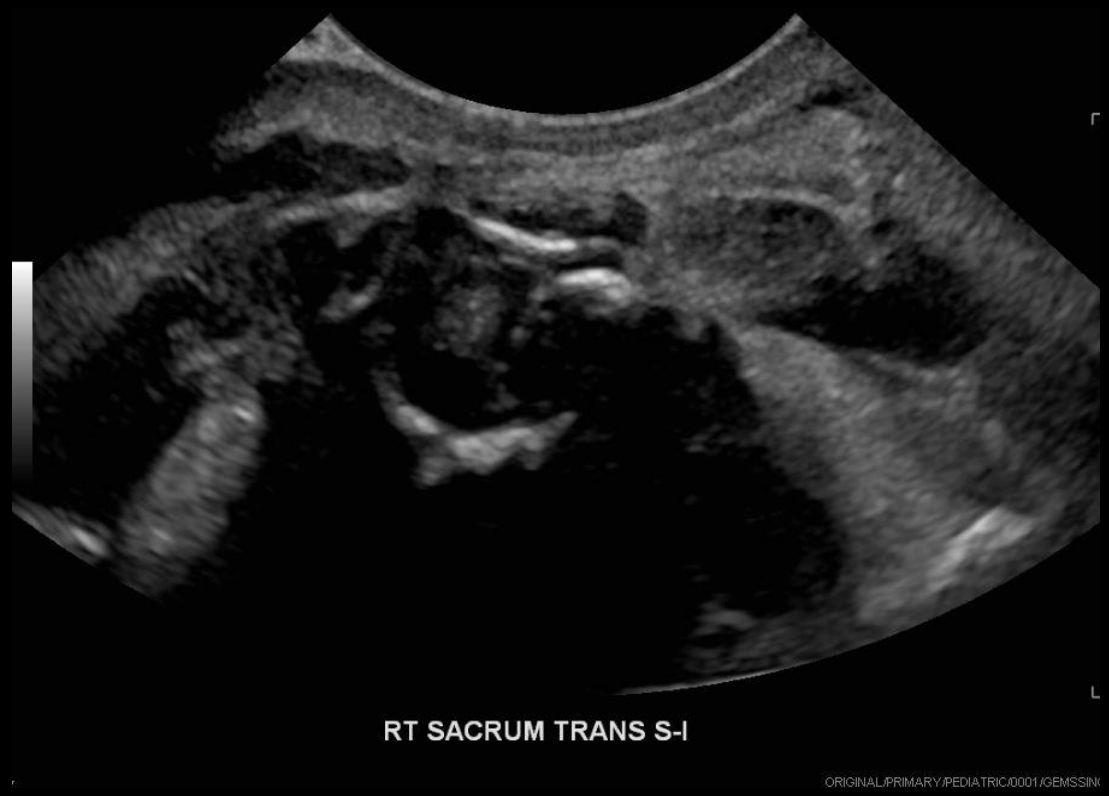




Ultrasound showed a very large sacral mass with both hyperechoic solid and anechoic cystic components and internal vascularity. There was no convincing presacral components by ultrasound

ULTRASOUND SOFT TISSUE SACRAL REGION



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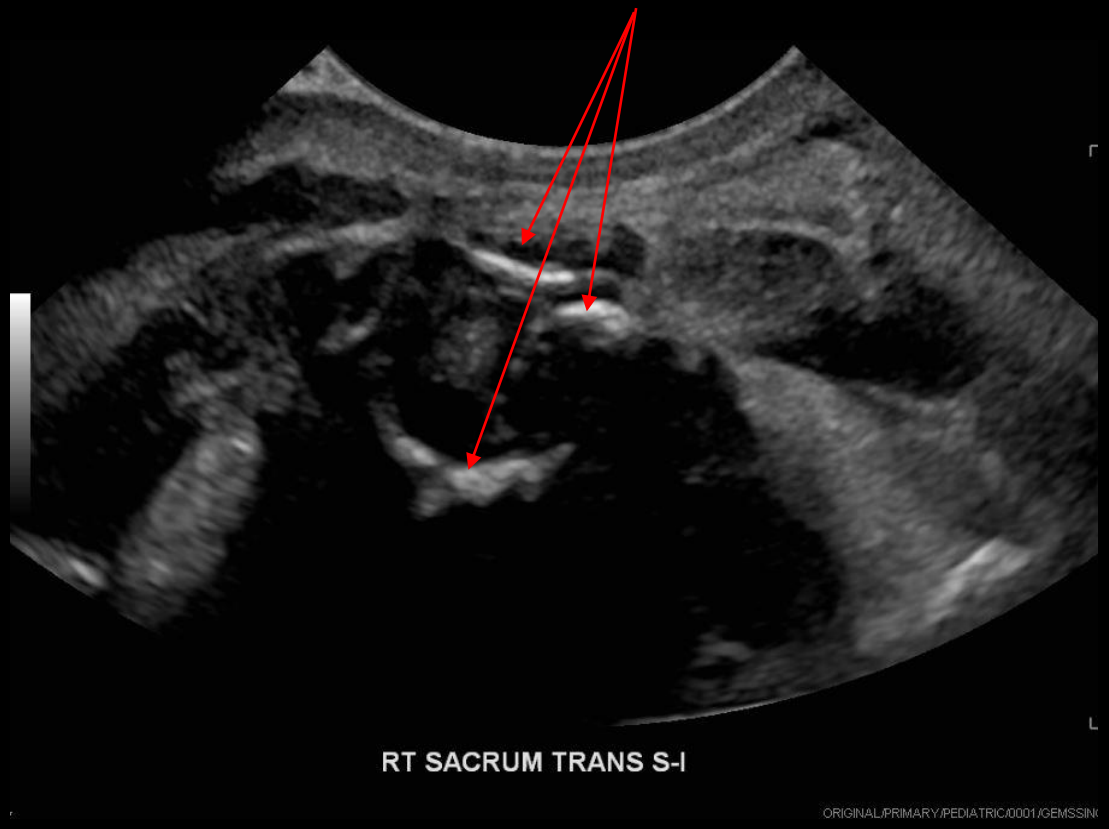


Probable calcifications (red arrows) within the sacral mass given associated posterior acoustic shadowing

ACR Appropriateness Criteria

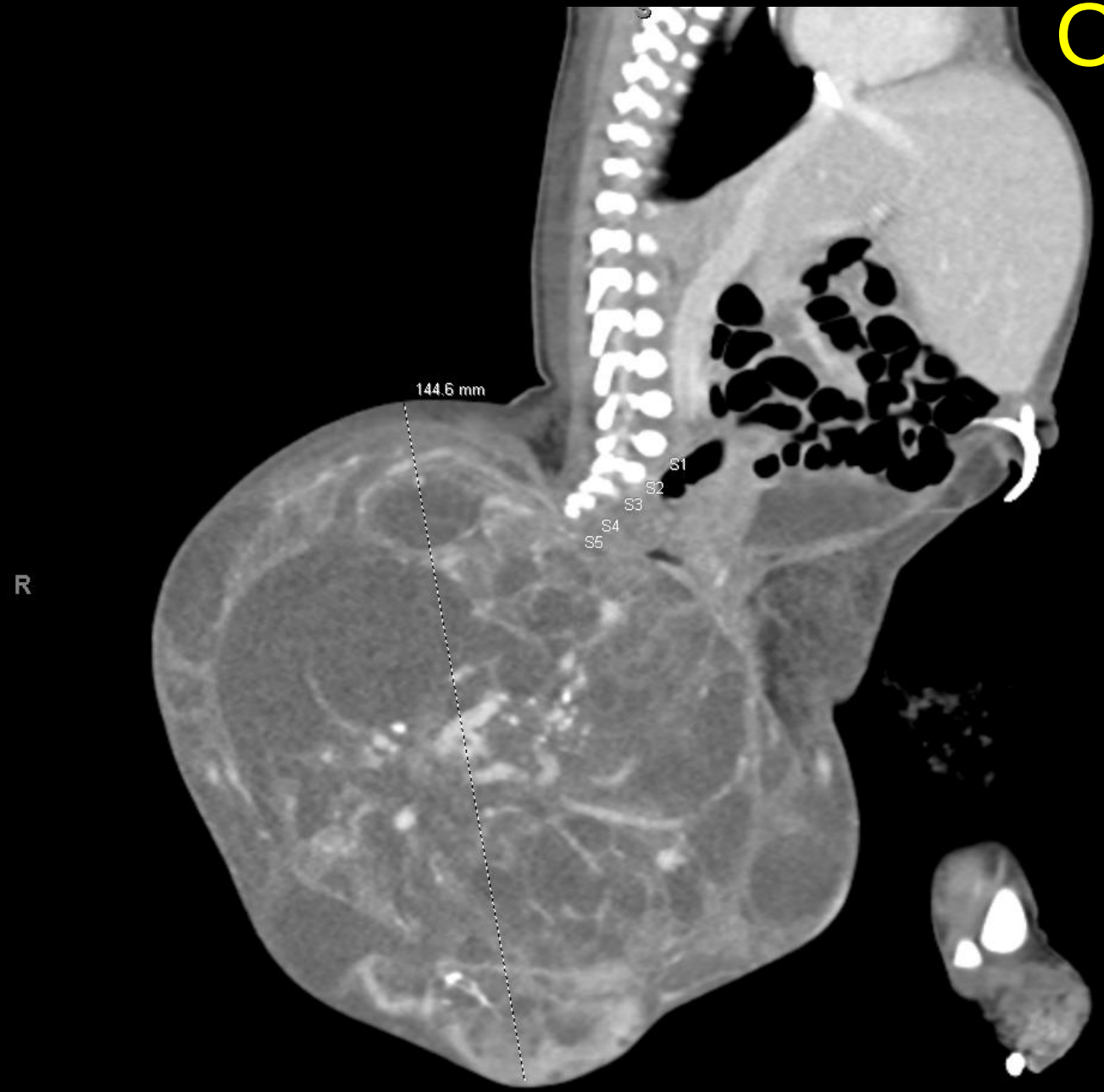
Additional imaging is needed as ultrasound could not rule out pre-sacral involvement

This imaging modality was ordered by the ordering physician

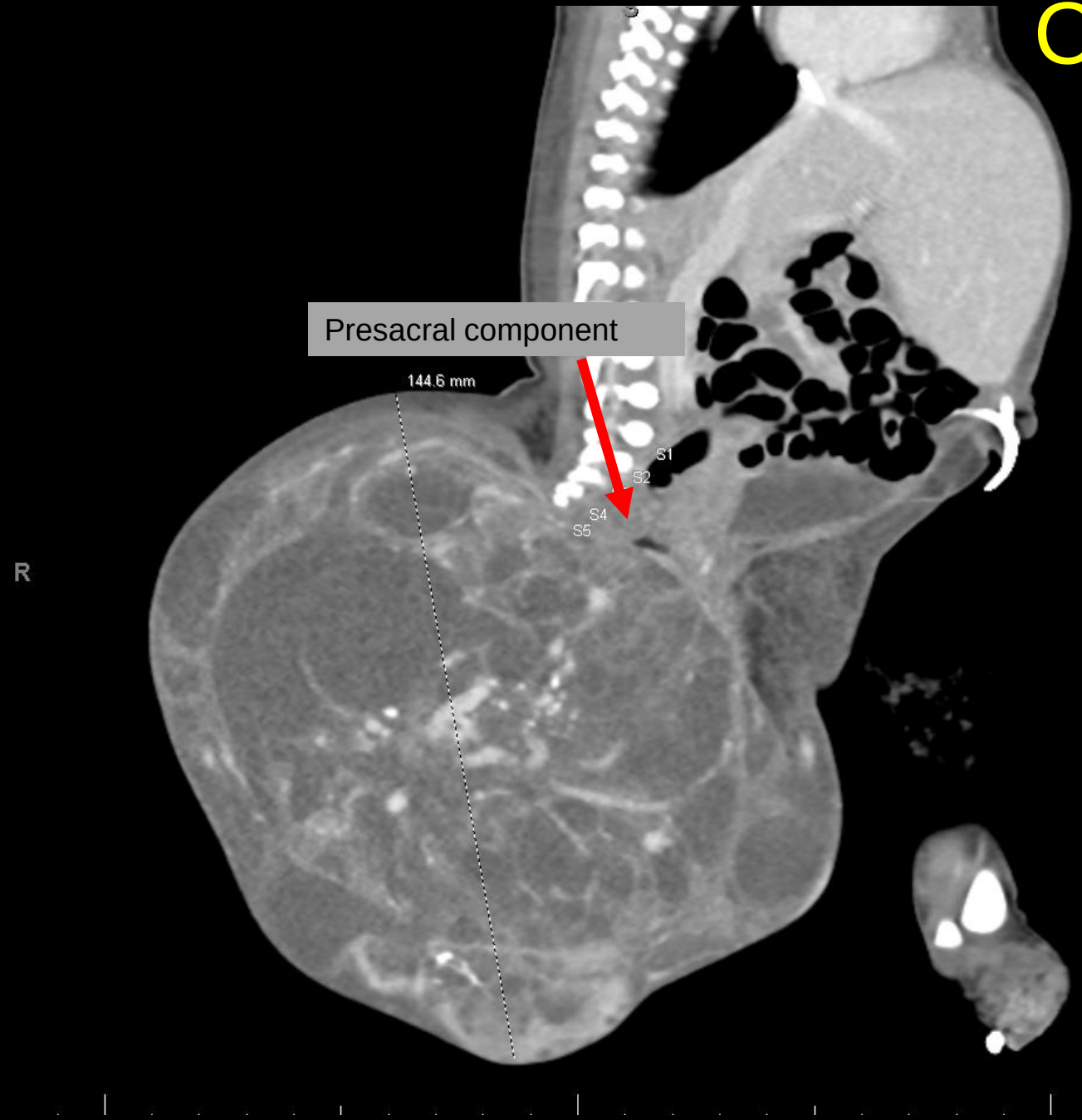
Variant 3: **Soft tissue mass. Nondiagnostic radiograph and noncontrast-enhanced ultrasound. Next imaging study.**

Procedure	Appropriateness Category	Relative Radiation Level
MRI area of interest without and with IV contrast	Usually Appropriate	O
MRI area of interest without IV contrast	May Be Appropriate	O
CT area of interest with IV contrast	May Be Appropriate	Varies
CT area of interest without and with IV contrast	May Be Appropriate	Varies
US area of interest with IV contrast	Usually Not Appropriate	O
Image-guided biopsy area of interest	Usually Not Appropriate	Varies
Image-guided fine needle aspiration area of interest	Usually Not Appropriate	Varies
FDG-PET/CT area of interest	Usually Not Appropriate	☢☢☢☢
CT area of interest without IV contrast	Usually Not Appropriate	Varies

CT ABDOMEN AND PELVIS



CT ABDOMEN AND PELVIS



Heterogenous mass measuring 12.3 x 14.2 x 14.5 cm. The mass was predominantly exophytic and had calcified, cystic, fatty, and soft tissue components. A small presacral component was apparent

Hospital course

Female neonate born at 33 weeks to a 32-year-old G4P1021 woman by c-section delivery due to worsening maternal condition (suspected sepsis), preterm premature rupture of membranes, and management of sacrococcygeal teratoma.

- Intubated for respiratory failure after delivery. Admitted to NICU for cardiac and respiratory monitoring.
- Surgical resection of type 1 sacrococcygeal teratoma. Pediatric oncology consulted.

Pertinent lab findings

Pre-operative:

- HCG: 71
- AFP: 294,515 ng/ml

Post-operative day #7:

- AFP: 33,992 ng/ml

Differential diagnosis

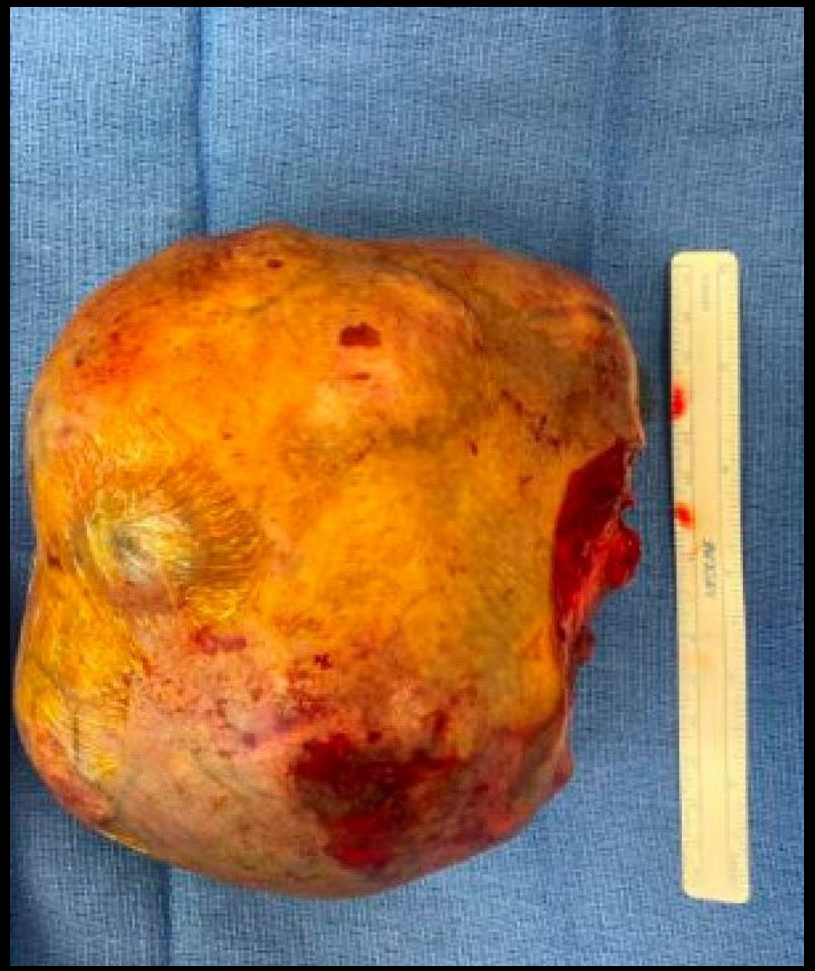
- Sacrococcygeal teratoma
- Sacral chordoma
- Sacral myelomeningocele
- Sacral meningocele

Surgical pathology findings

Surgical pathology: sampling of the mass reveals a teratoma containing various mature epithelial, neuroectodermal and mesenchymal tissue types.

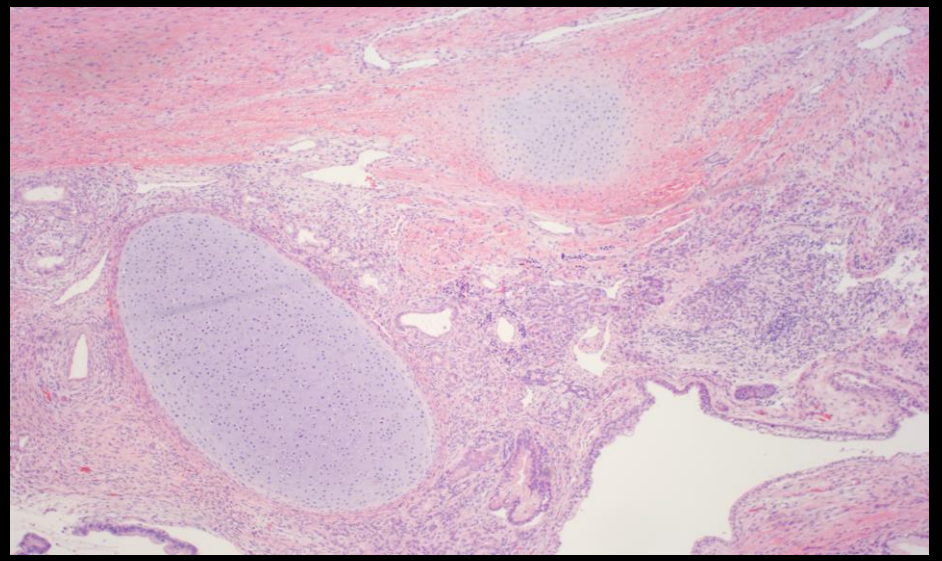
- Also present is a component of immature neuroectoderm with prominent proliferative activity focally forming rosette-like structures. The tumor grossly appeared completely excised with a pseudocapsule of compressed connective tissue at the margin.
- Tumor weighed 1163g and was sent to pathology

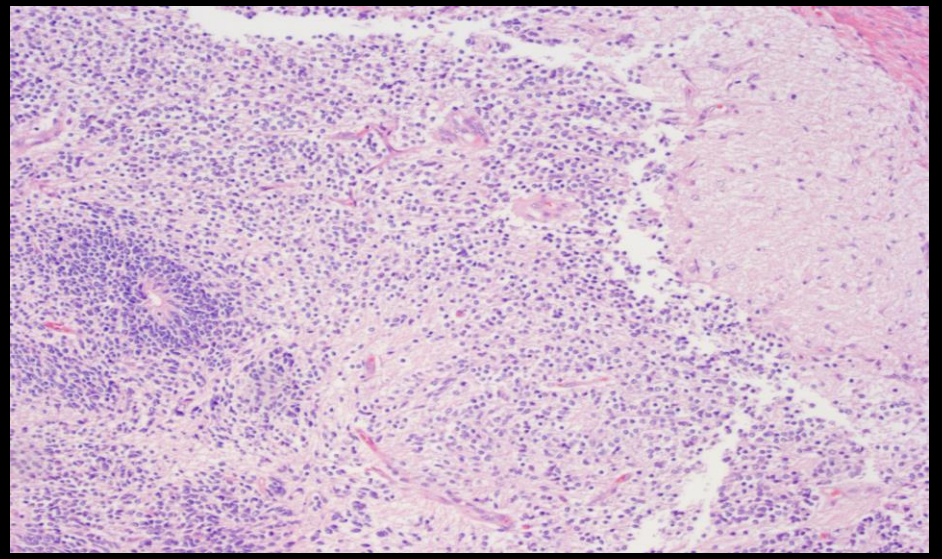
Gross appearance



Large resected sacrococcygeal teratoma measuring 14.5 cm weighing 1163 g

Micro Path





Sampling of the mass revealed a teratoma containing various mature epithelial, neuroectodermal, and mesenchymal tissue types. Also present was a component of immature neuroectoderm with prominent proliferative activity focally forming rosette-like structures. Microscopically, tumor is focally found within 1 mm of the inked margin.

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Final Dx:

Type 1 sacrococcygeal teratoma
with presacral component

Discussion

Sacrococcygeal teratomas (SCT) are extragonadal germ cell tumors that develop in fetal and neonatal periods. The American Academy of Pediatric Surgery Section Survey classifies sacrococcygeal teratomas based on their location and degree of invasion.

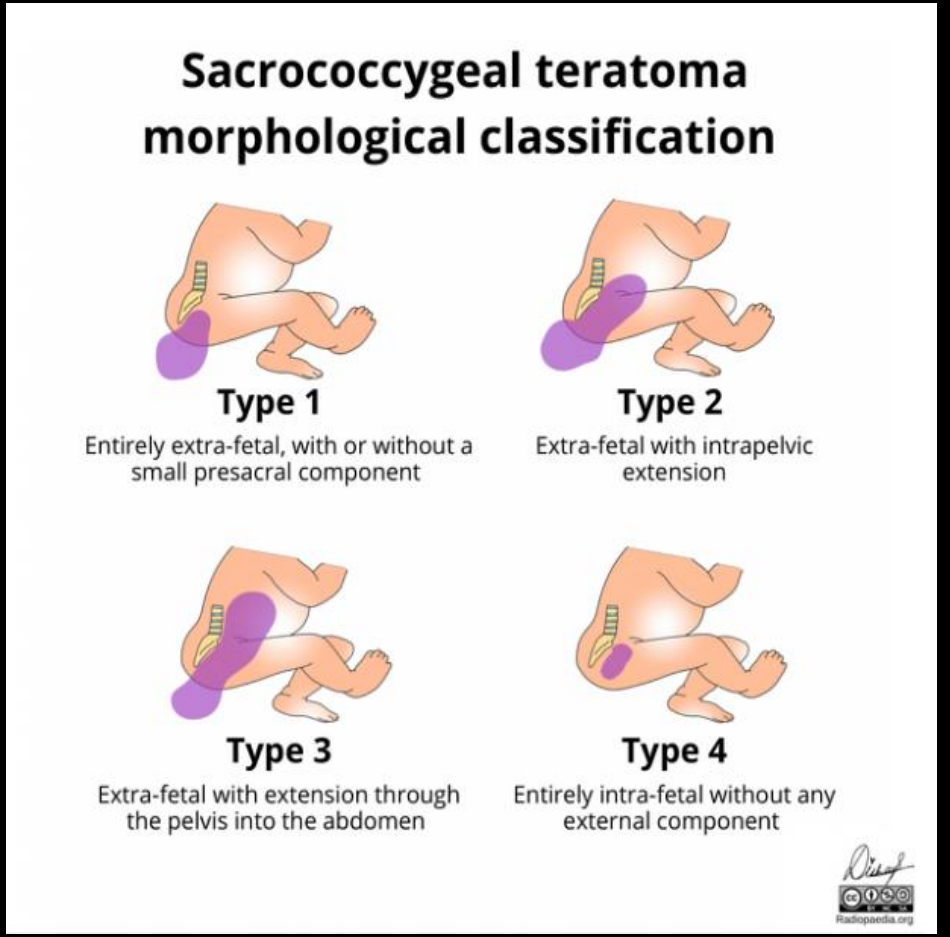
Type 1 teratomas develop exclusively on the outside of the fetus and may or may not have a pre-sacral component.

Type 2 teratomas are extra-fetal with intrapelvic extension.

Type 3 teratomas are extra-fetal with extension through the pelvis into the abdomen.

Type 4 teratomas develop entirely within the fetal pelvis.

The SCT found in this patient was diagnosed *in utero* and was morphologically classified as Type 1 with presacral component as no significant intrapelvic involvement was noted.



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Discussion (cont.)

Rarely, mature teratomas have the potential for malignancy. Factors that may confer greater risk of malignancy include presentation beyond the newborn period, greater degree of intra fetal components, and incomplete primary resection allowing for malignant transformation. Unique to this case, a 10-15% yolk sac component was noted which upstages the tumor, making it potentially malignant. Monitoring of serum alpha fetoprotein (AFP) and MRI is required for long-term surveillance. SCTs are very rare, and a malignant component is exceedingly rare with few case reports. The patient's case was presented at the oncology tumor board which decided to forgo chemotherapy or radiation. Instead, close clinical surveillance was chosen including physical examinations every 3 months for 2-3 years and then every 6 months until 5-6 years post-resection.

Discussion (cont.)

Prenatal imaging will mainly involve ultrasound of the fetus as a safe, initial approach. More recently, MRI may also be used adjunctively for further SCT characterization and assessment. In the neonatal period, although non-invasive and without ionizing-radiation, MRI may be challenging with newborn infants due to the need for sedation.

Discussion (cont.)

The initial imaging modality for a soft tissue mass such as the SCT found in this patient may include ultrasound or radiograph. Afterwards, if the ultrasound or radiograph is nondiagnostic, a CT with contrast may be considered next. MRI may also be considered as an appropriate alternative to CT. For this case, ultrasound was decided initially, followed by CT. The team felt the CT was the safer option for the patient as it would require less sedation. If CT did not demonstrate the necessary anatomy, MRI would be the next option. After diagnosis, complete surgical resection is key for tumor control.

References:

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