

RADY 403 Langerhans Cell Histiocytosis

Erica Zeng 5/20/22

Focused patient history

- 3yo male patient presenting with a presumed forehead hematoma. Parents said that patient hit his head while playing around 2 months ago. The initial bump has since resolved but two other bumps on the forehead and temple were noticed. Patient has complained of intermittent headaches that are treated with ibuprofen. Other than increased swelling that interferes with his visual field, the patient has experienced no other neurological changes.

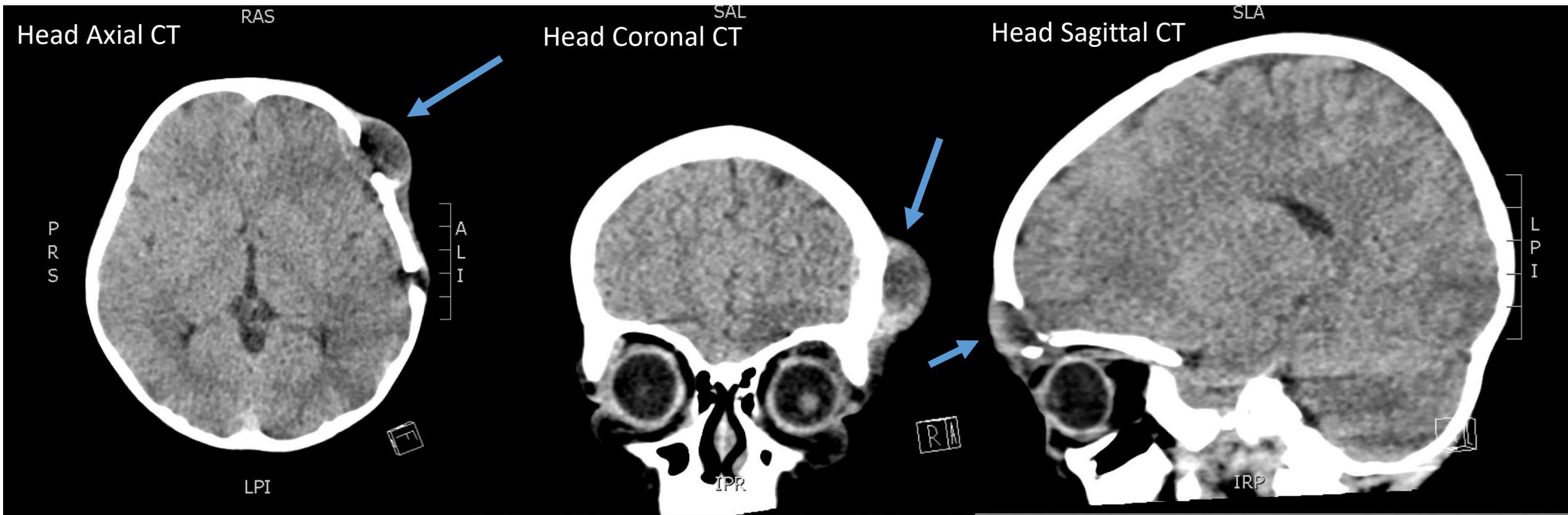
ACR Criteria

- Pertinent Positive on Physical Exam
 - 3 soft protrusions or swellings on head
- Obtained CT of head from skull base to vertex without contrast

Scenario	Procedure	Adult RRL	Peds RRL	Appropriateness Category	
Primary bone tumor suspected, skull, osteoid osteoma suspected on xray or clinical exam	CT head without IV contrast	1-10 mSv ○○○	0.3-3 mSv [ped] ○○○	Usually appropriate	●
	MRI head without IV contrast	0 mSv ○	0 mSv [ped] ○	May be appropriate	●
	MRI head without and with IV contrast	0 mSv ○	0 mSv [ped] ○	May be appropriate	●
	Bone scan whole body with SPECT or SPECT/CT brain	1-10 mSv ○○○	3-10 mSv [ped] ○○○○	May be appropriate	●
	CT head without and with IV contrast	1-10 mSv ○○○	3-10 mSv [ped] ○○○○	May be appropriate	●
	Bone scan whole body	1-10 mSv ○○○	3-10 mSv [ped] ○○○○	Usually not appropriate	●
	US transcranial	0 mSv ○	0 mSv [ped] ○	Usually not appropriate	●
	CT head with IV contrast	1-10 mSv ○○○	0.3-3 mSv [ped] ○○○	Usually not appropriate	●
	FDG-PET/CT whole body	10-30 mSv ○○○○	3-10 mSv [ped] ○○○○	Usually not appropriate	●

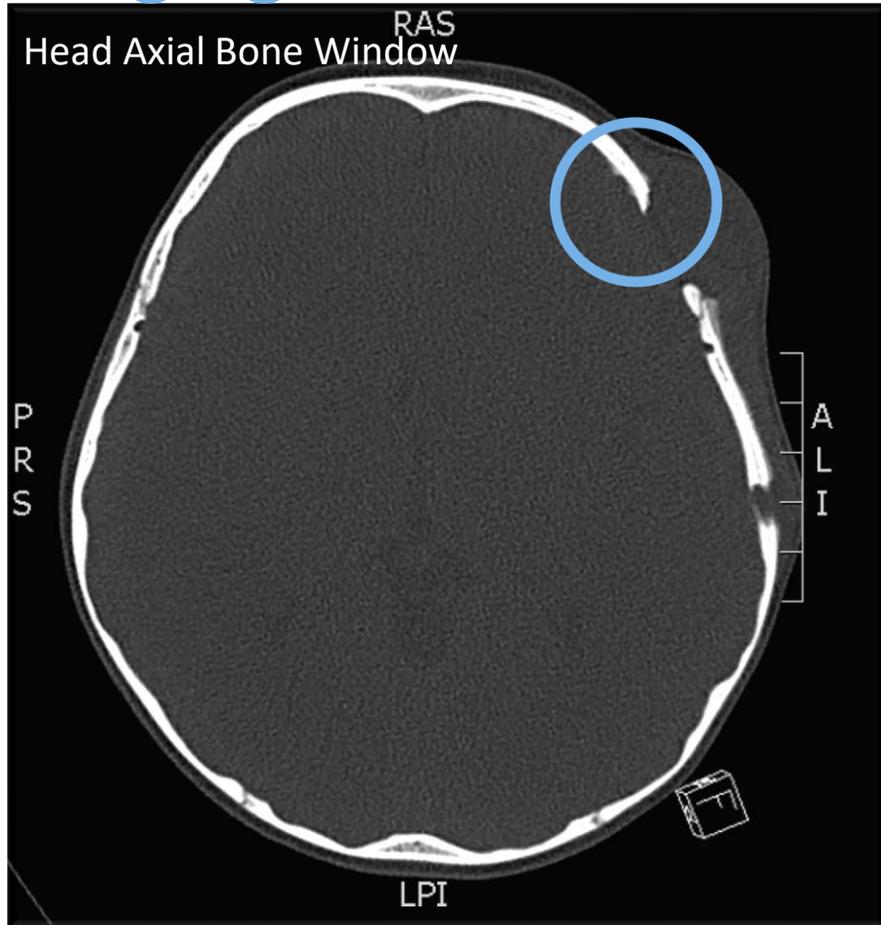
Image obtained from the American College of Radiology clinical resources page for 2022 ACR Appropriateness Criteria (<https://www.acr.org/Clinical-Resources/ACR-Appropriateness-Criteria>)

Imaging studies from PACS 1

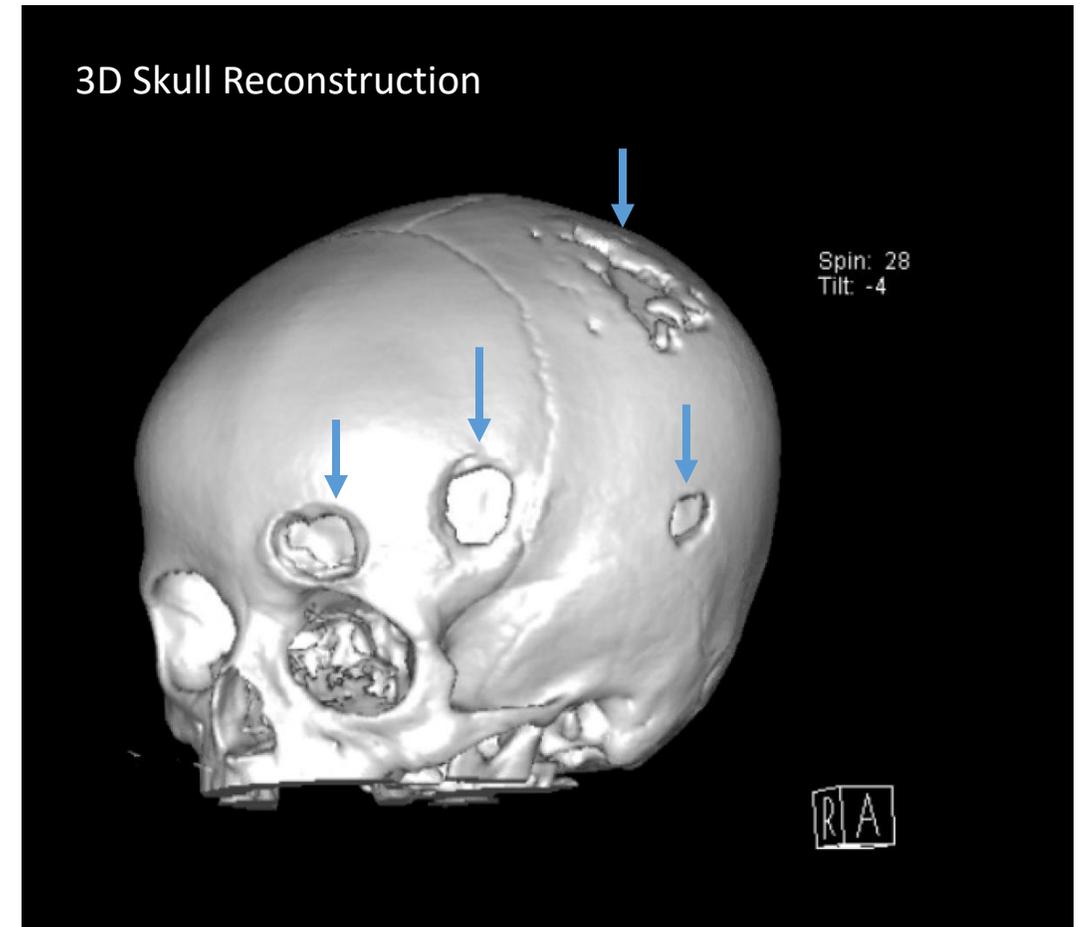


CT w/o contrast: No midline shift, mass lesion, acute infarct or hemorrhage. The sinuses are pneumatized. A large area of skull base dehiscence is seen involving the left parietal region as well as smaller areas in the left temporal, lateral left frontal, and anterior left frontal regions.

Imaging studies from PACS 2



Beveled edge observed on bone window imaging of the skull. A characteristic finding for the final diagnosis.



Skull base dehiscence observed in the 3D skull reconstruction, involving the left parietal region as well as smaller areas in the left temporal, lateral left frontal, and anterior left frontal regions.

Differential Diagnosis

- Immediate concern for head trauma resulting in growing skull fracture (leptomeningeal cyst)
- Multiple Skull Lesions
 - Langerhans Cell Histiocytosis (multiple skull lesions)
 - Metastases-Neuroblastoma, lymphoma, leukemia, infection (multiple skull lesions)
- Single Skull Lesions
 - Bone epidermoid cyst (single skull lesion)
 - Ewing's Sarcoma (single skull lesion)

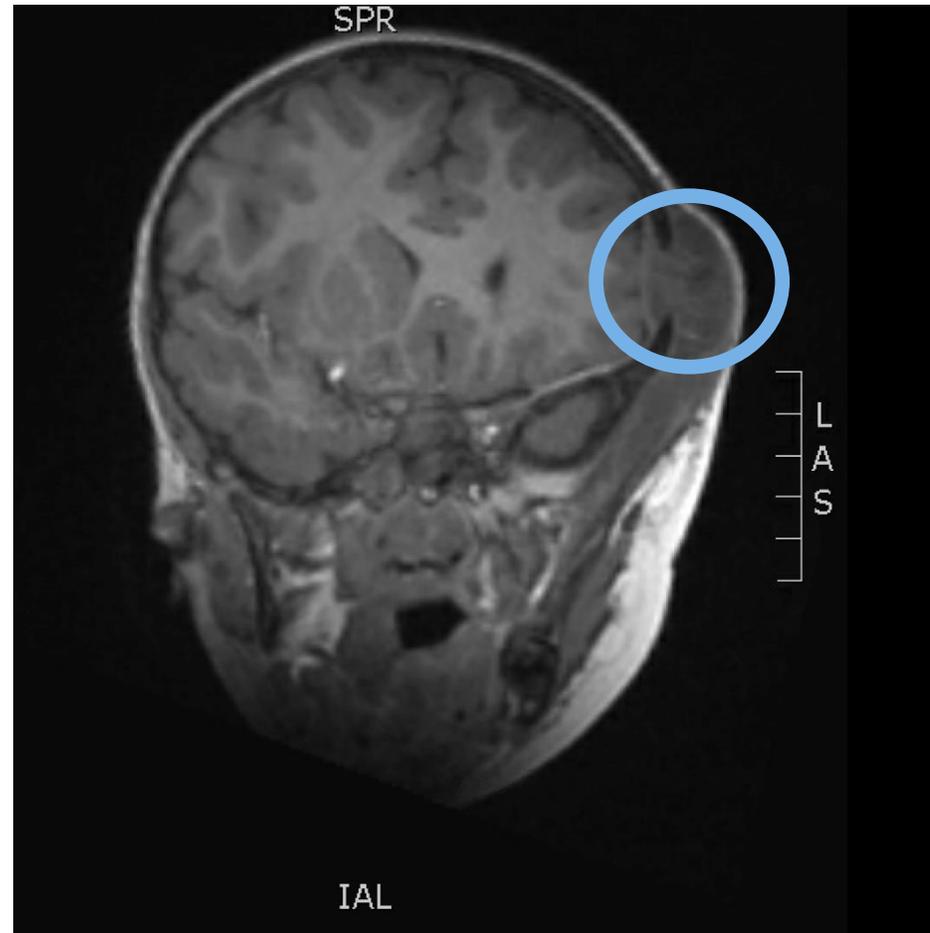
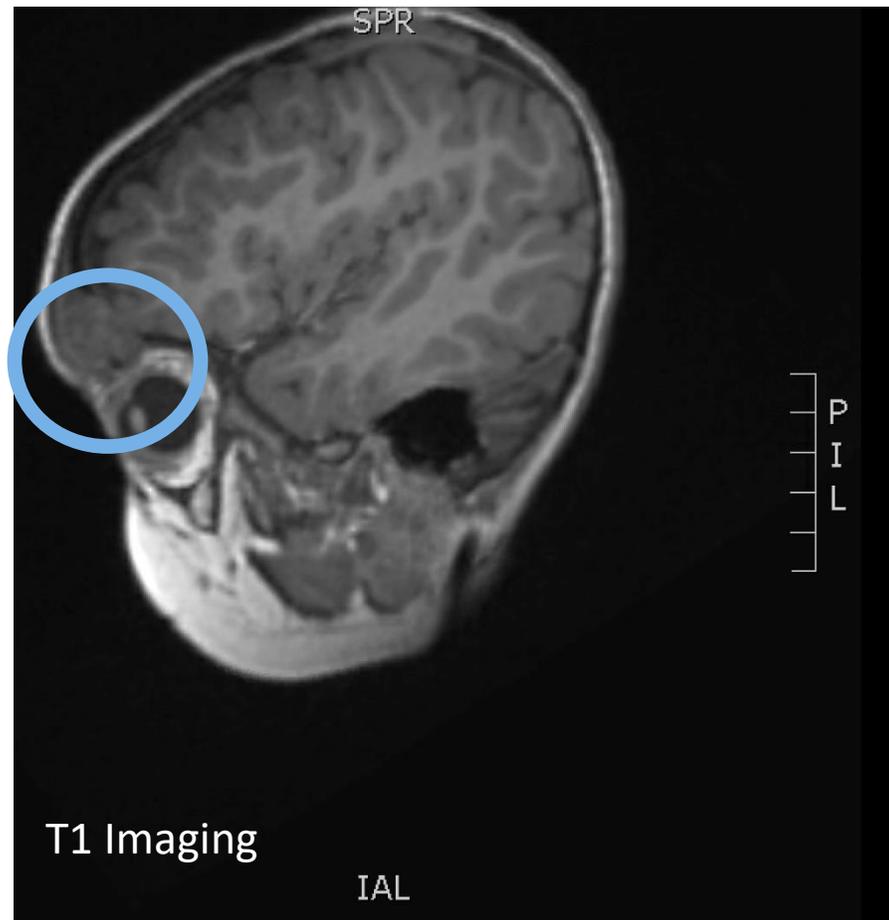
Further Steps

- MRI of the brain
- Full body imaging to rule out additional sites of disease
- Biopsy lesion

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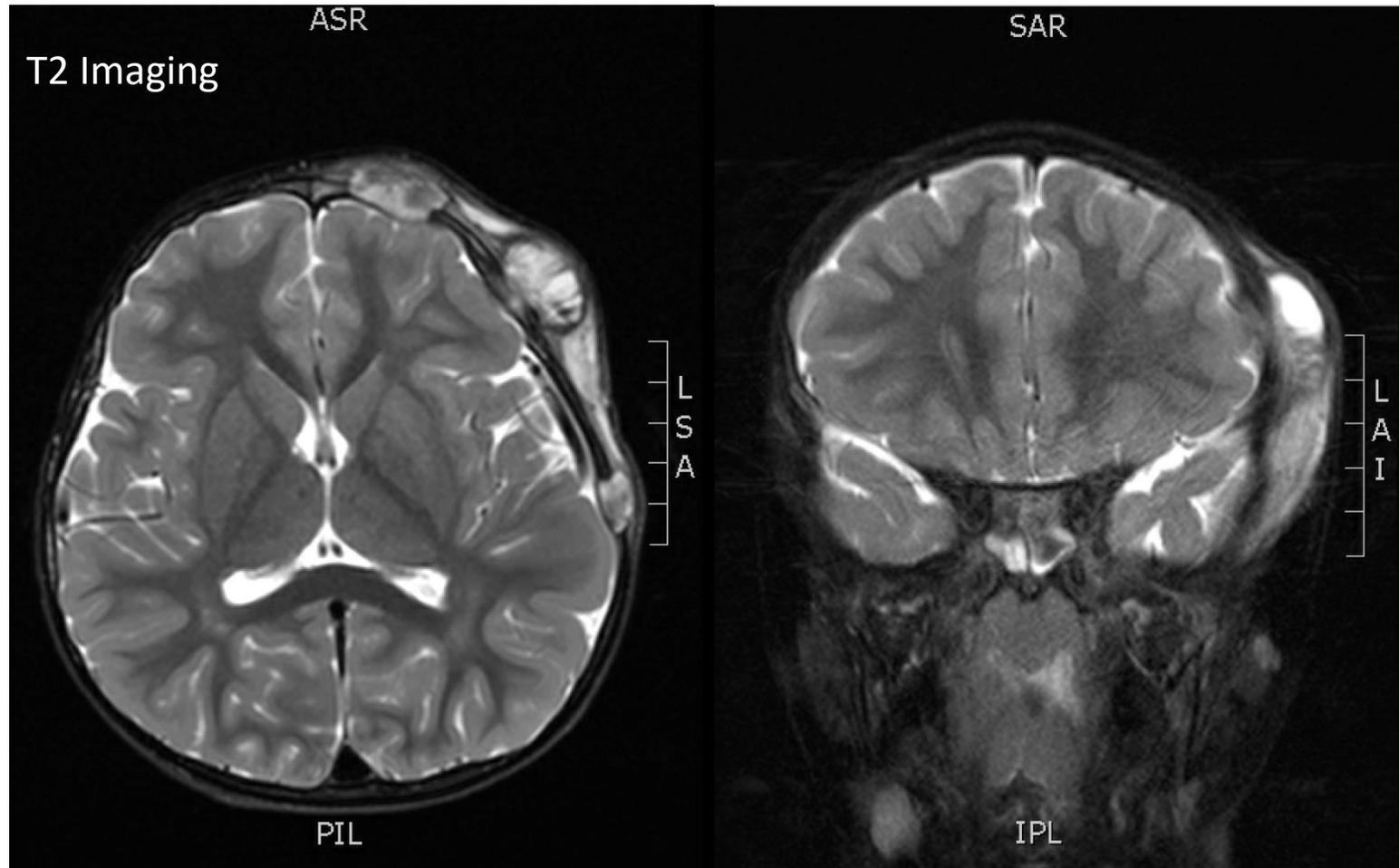
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Imaging studies from PACS 3



As seen on prior head CT, there are multiple lytic skull lesions along the left frontoparietal calvarium.

Imaging studies from PACS 4



Enhancing soft tissue lesions with bony destruction. Lesions causing mild mass effect on the underlying parenchyma without intraparenchymal involvement. Ventricles are normal in size, no extra axial fluid collection visualized.

Imaging studies from PACS 5

Multiple lytic lesions visualized in the skull. Lytic lesion in the right iliac with mild FDG uptake. Both findings concerning for LCH. Mild uptake in spleen and bowel may be indicative of inflammation.



Diagnosis: Langerhans Cell Histiocytosis

- Peak age 1-3, but also diagnosed in adults
- A malignancy due to uncontrolled monoclonal proliferation of Langerhans cells accompanied by inflammation and granuloma formation
- characteristic Birbeck granules on electron microscopy (previous gold standard, outdated modality)
- Immunohistochemistry- HLA-DR, CD1a, CD207, S100
- Most common lesion observations:
 - Bone most often affected is the skull (calvaria > maxillofacial > skull base)
 - Spinal lesions (thoracic > lumbar > cervical)
 - Long bone lesions most often in the femur, affecting the metaphysis or diaphysis
 - Cranial lesions often described as round/ovoid, lytic, clear boundaries
 - Characteristic bevelled edges
 - Lesions often accompanied by soft tissue masses
 - Bones can also have more permeative changes

Langerhans Cell Histiocytosis Cont.

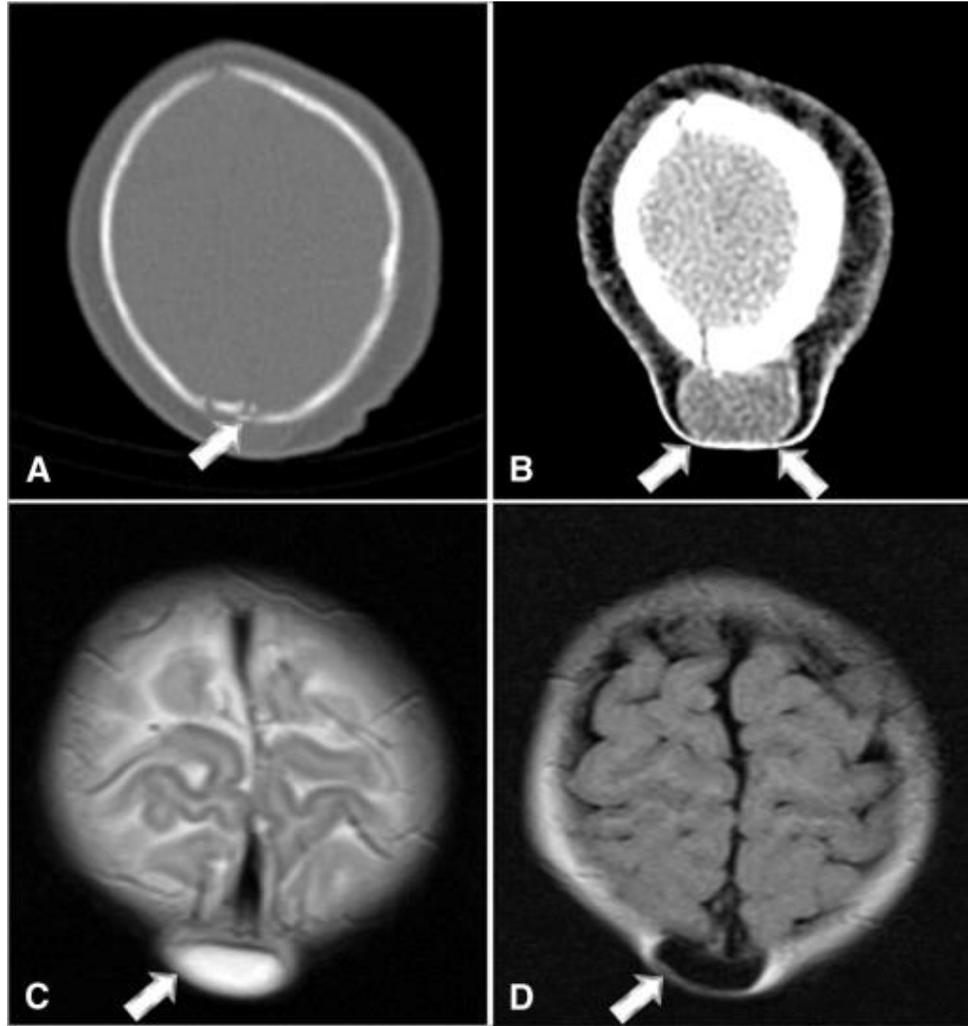
- Historically 3 Syndromes:
 - Letterer Siwe Disease
 - rare
 - Generally: Multi-organ systems, multiple sites involved
 - Hand-Schuller-Christian Disease
 - Often accompanied by diabetes insipidus
 - Generally: Single organ system, multiple sites involved
 - Eosinophilic Granuloma
 - Most common, m>f
 - Common locations: spine, clavicle, pelvis
 - Generally: Single lesions
- Today:
 - Single system single site (SS-s)
 - Single system multi-site (SS-m)
 - Multisystem (MS)

Patient treatment or outcome

- Most cases resolve on their own
- Treatment ranges between low vs high risk LCH patients and depends on where lesions are located
 - High risk organ: spleen, liver, bone marrow
 - Observation
 - Physical or medical reinforcement of bone structure depending on the extent of bone lesions
 - For low risk: generally localized therapy or surgery +/- steroids
 - For high risk: chemotherapy and steroid therapy

Similar Appearing Diagnoses

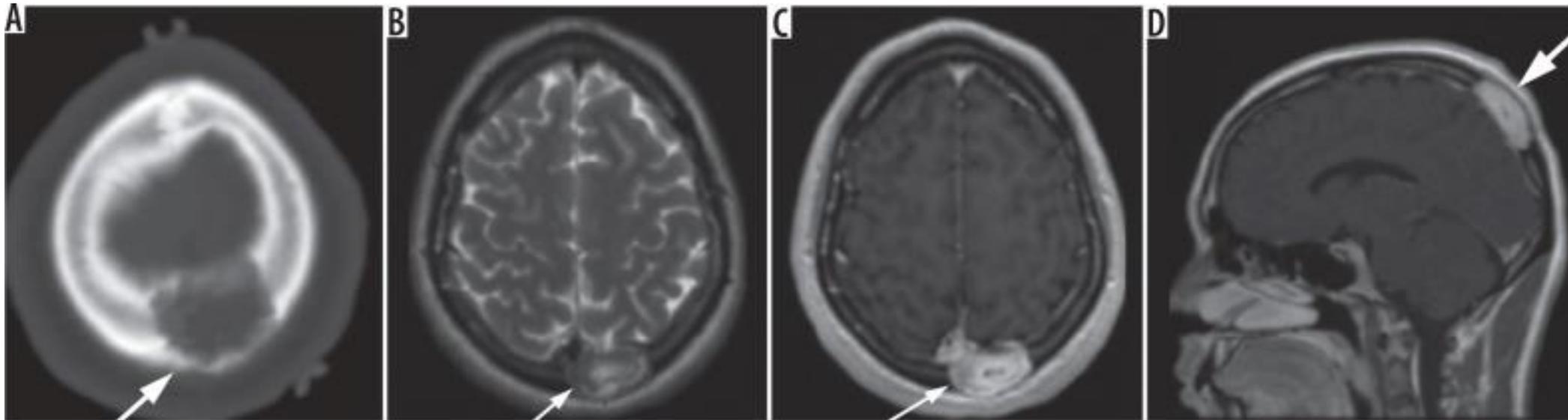
- Post traumatic leptomenigeal cyst has dural laceration and skull fracture
- Often see associated pathologies such as subdural fluid collection, ventricular dilation



Ibrahim Guler, Meral Buyukterzi, Ozgur Oner, Ismet Tolu, Post-Traumatic Leptomeningeal Cyst in a Child: Computed Tomography and Magnetic Resonance Imaging Findings, The Journal of Emergency Medicine, Volume 48, Issue 5, 2015, Pages e121-e122, ISSN 0736-4679, <https://doi.org/10.1016/j.jemermed.2014.12.042>.

Similar Appearing Diagnoses

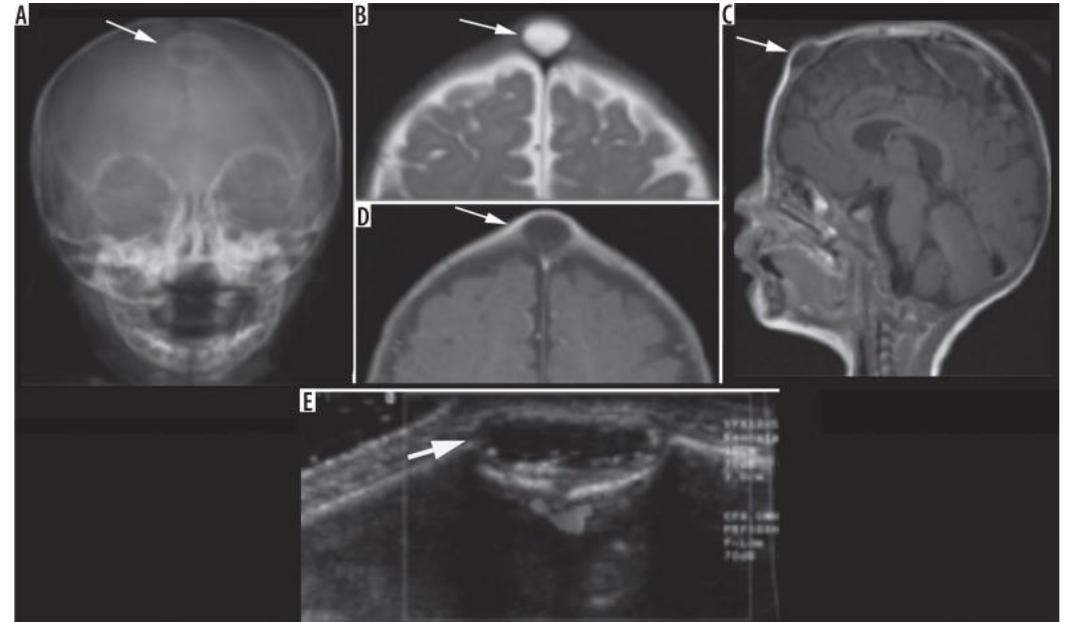
- **Intraosseous hemangiomas** are also well defined osteolytic lesions affecting the outer table however periosteal reactions are rare
- MRI can depict vascular channels



Choudhary G, Udayasankar U, Saade C, Winegar B, Maroun G, Chokr J. A systematic approach in the diagnosis of paediatric skull lesions: what radiologists need to know. *Pol J Radiol.* 2019;84:e92-e111. Published 2019 Feb 8. doi:10.5114/pjr.2019.83101

Similar Appearing Diagnoses

- **Bone epidermoid cyst** depicted in the image to the right
 - also has beveled edge and lytic lesions of the skull, but inner table more affected than outer
- **Myeloma**
 - in adults, so unlikely in this patient
 - also has beveled edge and described punched out lesions but often + osteoporosis

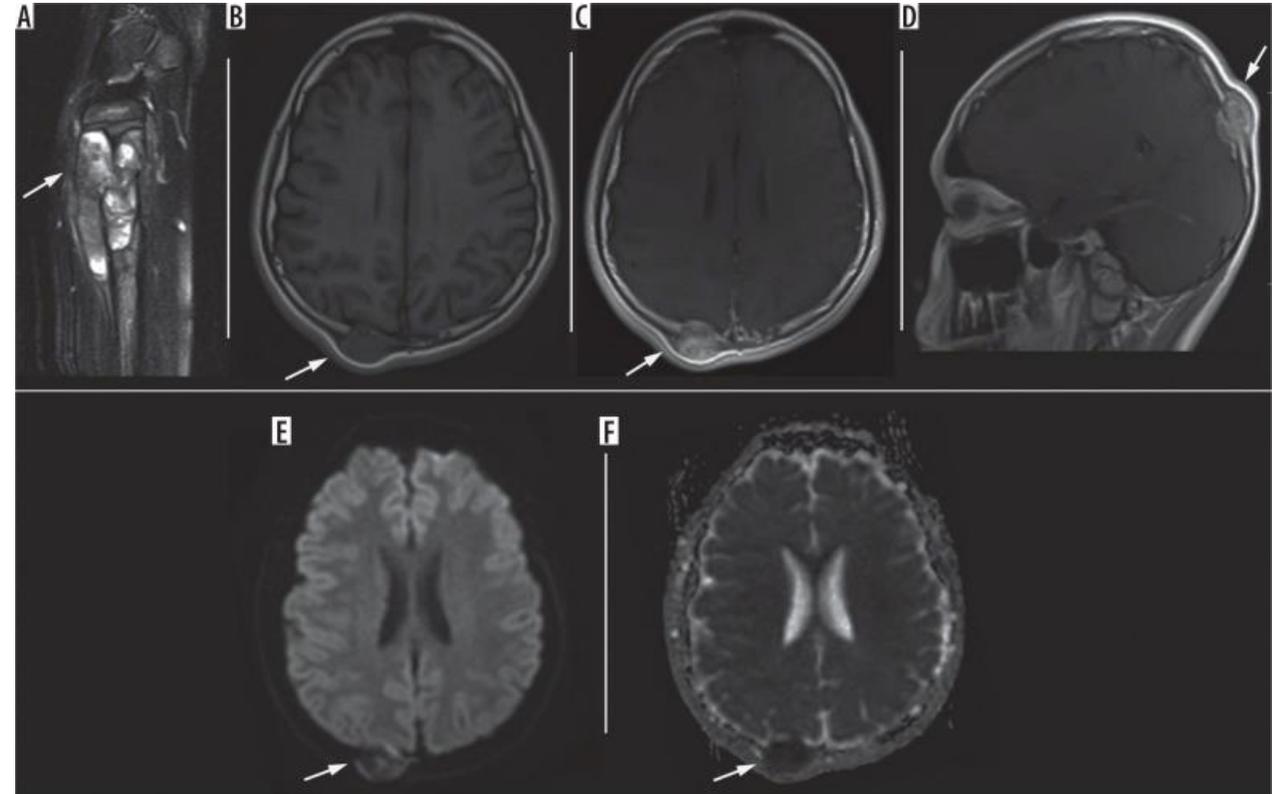


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Similar Appearing Diagnoses

- Ewing's Sarcoma

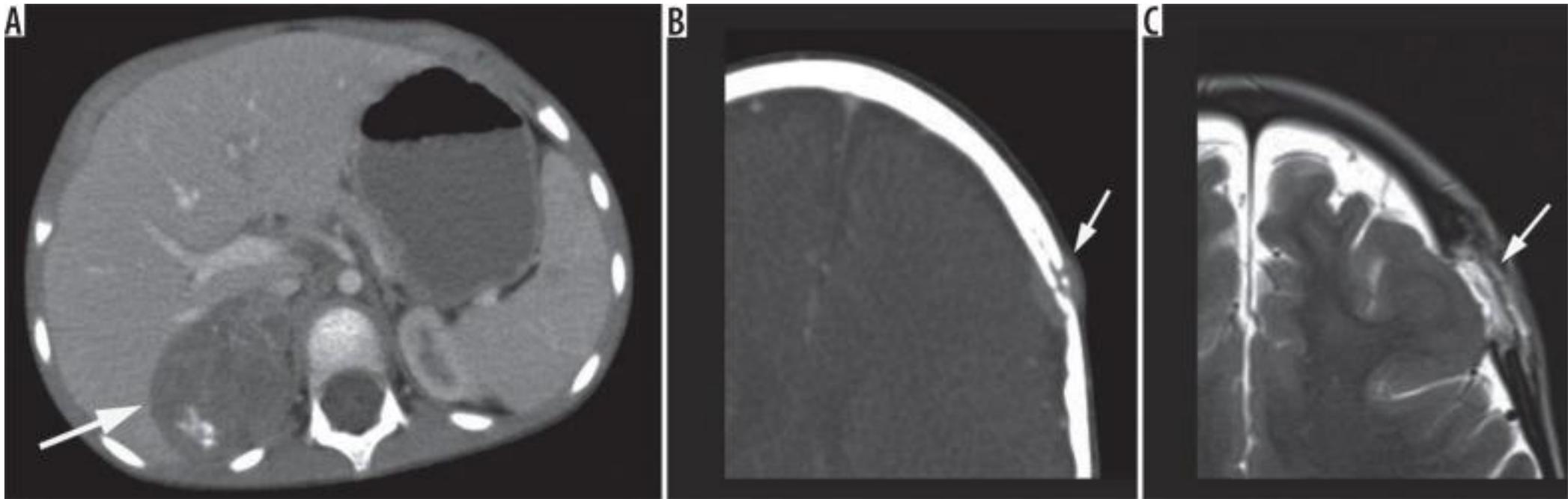
- Often see a similar periosteal reaction with onion skin appearance
- But less likely to have lesions surrounded by marrow and soft tissue edema
- Lesions are also usually poorly margined, and more commonly singular



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Similar Appearing Diagnoses

- **Metablasic neuroblastoma**
 - Has the “hair on end” periosteal reaction, lytic defects and separation of sutures
 - Tends to metastasize to the dura and favors its external surface



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