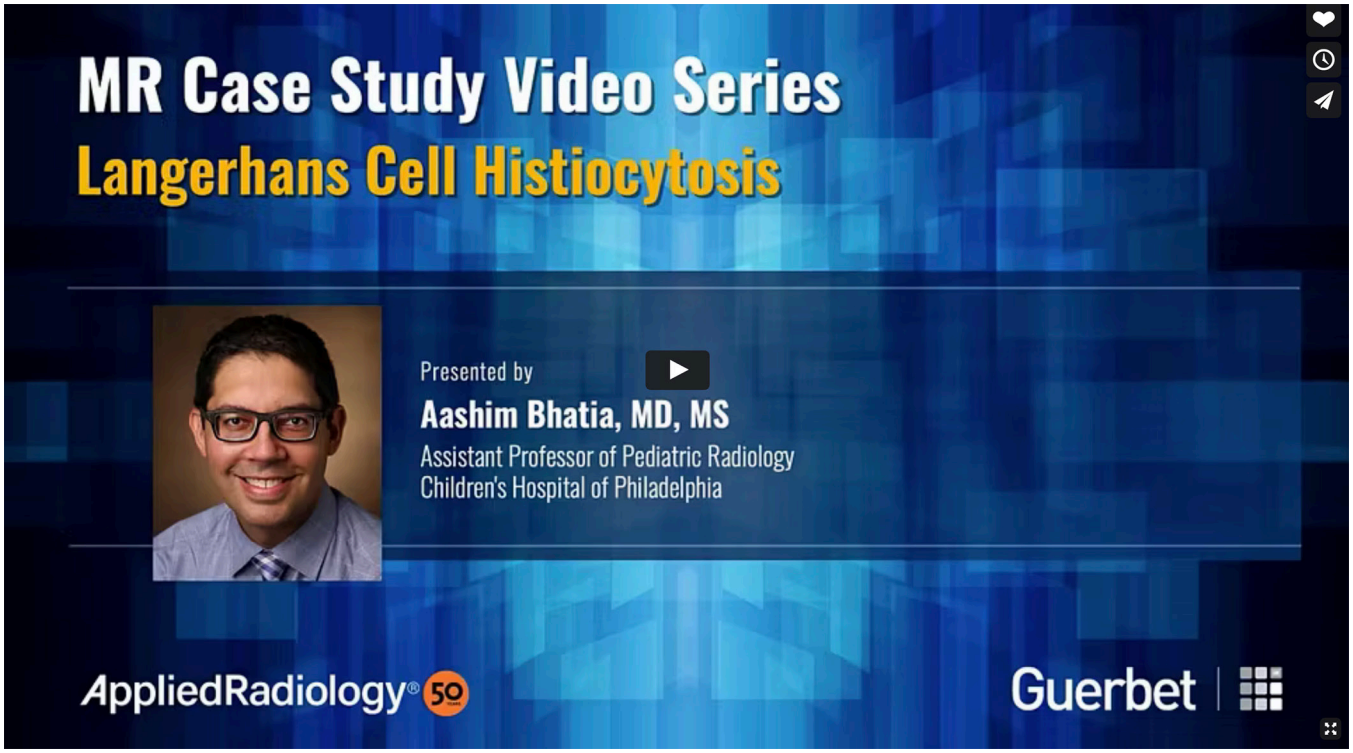

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Langerhans Cell Histiocytosis | Case 2

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Case Summary

An otherwise healthy 4-year-old presented to the emergency department with a lump on his head. A noncontrast computed tomography (CT) scan and both noncontrast and contrast-enhanced magnetic resonance imaging (MRI) scans of the head were performed.

Imaging Findings

The CT scan demonstrated a large heterogeneous scalp mass extending into the calvarium within the right frontal bone. There was erosion of the right frontal bone involving the outer and inner table of the calvarium, from the lytic lesion. Noncontrast and contrast-enhanced MRI scans of the brain also revealed

the mass to be heterogeneous and showed fluid levels containing blood. Contrast-enhanced MRI showed that the mass extended through the calvarium and involved the dura but did not involve the brain parenchyma.

Diagnosis

Langerhans Cell Histiocytosis

Discussion

Langerhans Cell Histiocytosis (LCH) is a rare, granulomatous systemic disease that is more common in children than in adults. It can present as single or multiple palpable masses that often involve the calvaria, skull base, and maxillofacial bones.[1] The most common imaging finding is a well-defined skull lesion demonstrating significant GBCA enhancement on MRI and contrast-enhanced CT imaging, appearing as osteolytic masses on the latter. A bone survey should be performed to determine if other bones are involved.

Gadolinium-based contrast agents (GBCA) are helpful when imaging any mass, whether intracranial or extracranial. Cases like this one require further work-up; the differential diagnosis in the pediatric patient include LCH, primary bone tumors, lymphoma, and metastatic disease. The work-up should include consultation with oncology and a full-body bone survey for additional lesions.

In this case, the bone survey revealed only a single, right frontal calvarial lytic lesion, which was confirmed by pathology as LCH.

Langerhans cell histiocytosis involvement of the central nervous system (CNS) is less common; patients can be asymptomatic for years.[2,3] Imaging findings in the CNS can include an enhancing mass within the tuber cinereum or infundibulum and absence of the T1 hyperintense posterior pituitary bright spot. These are best appreciated with a dedicated MRI scan of the pituitary that includes thin-section, T1 postcontrast sequences with a GBCA. The clinical presentation typically accompanies diabetes insipidus.

Brain lesions can involve multiple regions and represent demyelination; they are best seen on T2/FLAIR sequences. The posterior fossa is a commonly associated region for LCH-related neurodegeneration; T1 hyperintense lesions of the dentate nuclei and globi pallidi can also be seen. While other histiocytic disorders affect the CNS, LCH is the most common. Other demyelinating processes such as acute disseminated encephalomyelitis are more common in children, and the history of LCH is important in accurately diagnosing neurodegenerative LCH.[4]

Conclusion

A well-defined lytic lesion involving the calvaria, skull base, or maxillofacial bones and displaying enhancement in pediatric patients should include LCH in the differential. Familiarity with CNS involvement of LCH can aid neuroradiologists in making an accurate diagnosis.

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