Langerhans Cell Histiocytosis

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Case Summary

A 6-month-old female with no significant medical history initially presented to the emergency department with acute right eyelid swelling and redness. Ophthalmology was consulted and the patient was diagnosed with orbital cellulitis, which was treated with antibiotics. The redness and swelling initially improved but gradually returned in a few weeks. On the return visit to ophthalmology, there was no conjunctival infection, eye discharge, or fever. Magnetic resonance imaging (MRI) of the brain and orbits was recommended.

MRI demonstrated a mass along the right frontal bone involving the orbital roof, as well as an additional mass in the left orbital roof. These findings were concerning for Langerhans cell histiocytosis (LCH) or metastatic disease, and the patient was referred to oncology for further workup. A subsequent bone survey did not show any additional masses within the skeletal system. Biopsy was then obtained of the right orbital mass.

Imaging Findings

MRI demonstrated a heterogeneous mass in the right frontal bone involving the orbital roof, with overall T1 hypointensity with facilitated diffusion and regions of T2 hyperintensity. The mass demonstrated enhancement following intravenous administration of a gadolinium-based contrast agent (GBCA) (Figure 1). Another, smaller, mass in the left orbital roof was best visualized with a postcontrast T1 fat-saturation sequence. There were no intracranial masses or additional calvarial lesions, nor was there abnormal leptomeningeal enhancement. MRI of the orbits showed normal extraocular muscles and optic nerves, with no abnormal enhancement.

A subsequent computed tomography (CT) exam demonstrated that the masses were associated with osseous erosions within the frontal bones, the right superolateral orbit, and the inner table (Figure 2). The smaller mass in the left frontal bone caused scalloping of the inner table and erosion of the outer table.

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 palpable masses, commonly involving 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. It is important to remember that LCH can present as single or multiple lesions.

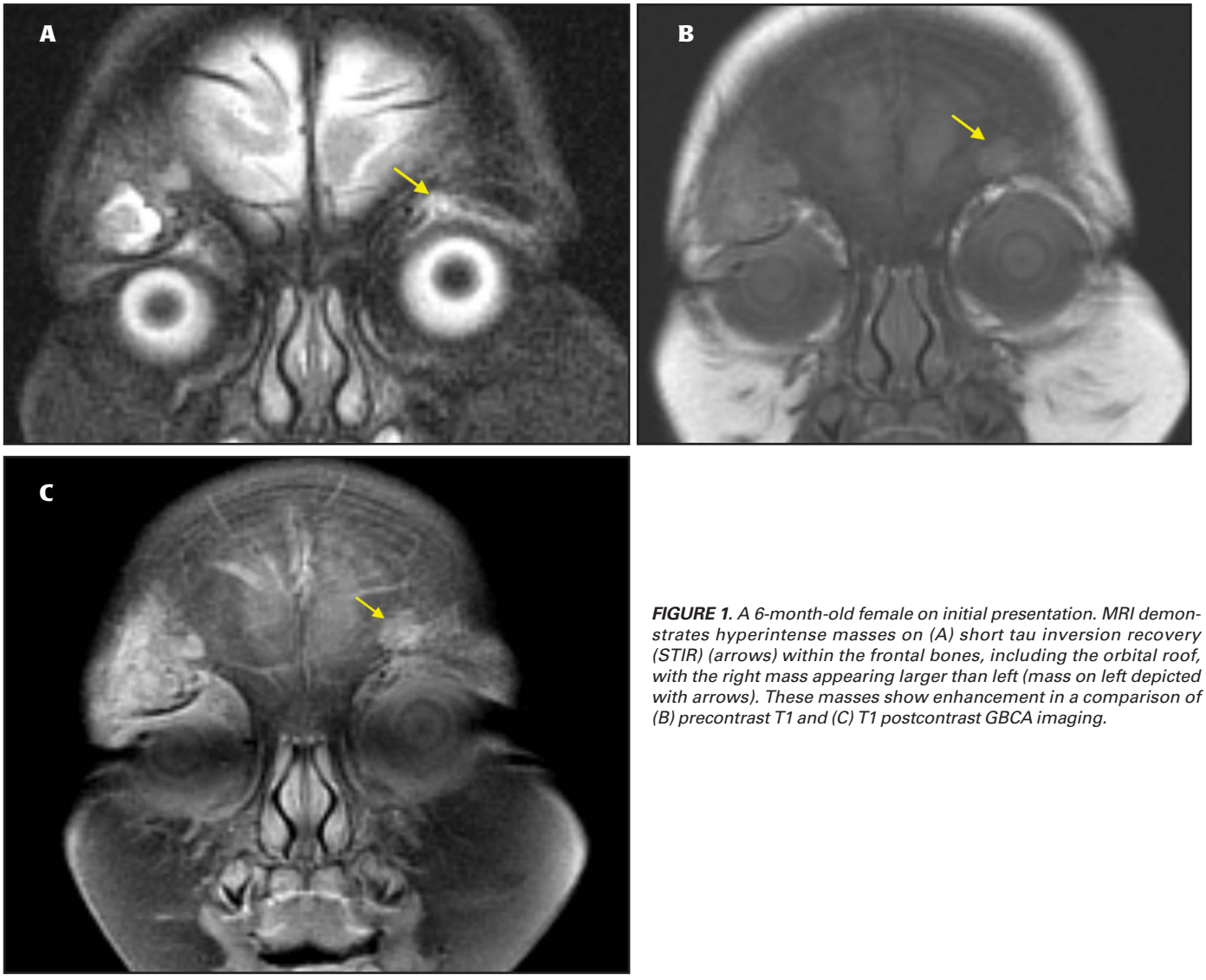

FIGURE 1. *A 6-month-old female on initial presentation. MRI demonstrates hyperintense masses on (A) short tau inversion recovery (STIR) (arrows) within the frontal bones, including the orbital roof, with the right mass appearing larger than left (mass on left depicted with arrows). These masses show enhancement in a comparison of (B) precontrast T1 and (C) T1 postcontrast GBCA imaging.*

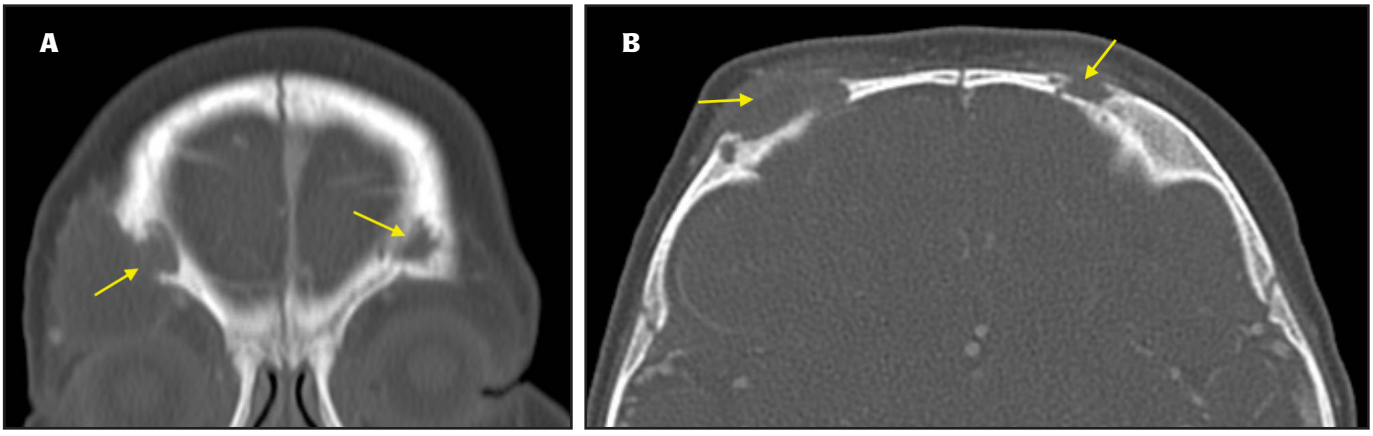

FIGURE 2. *At presentation, CT performed after the MRI shows (A) lytic lesions in the frontal bones, including the orbital roof on coronal reconstruction (arrows) with associated enhancing masses, right larger than left, also seen in the axial view (B).*

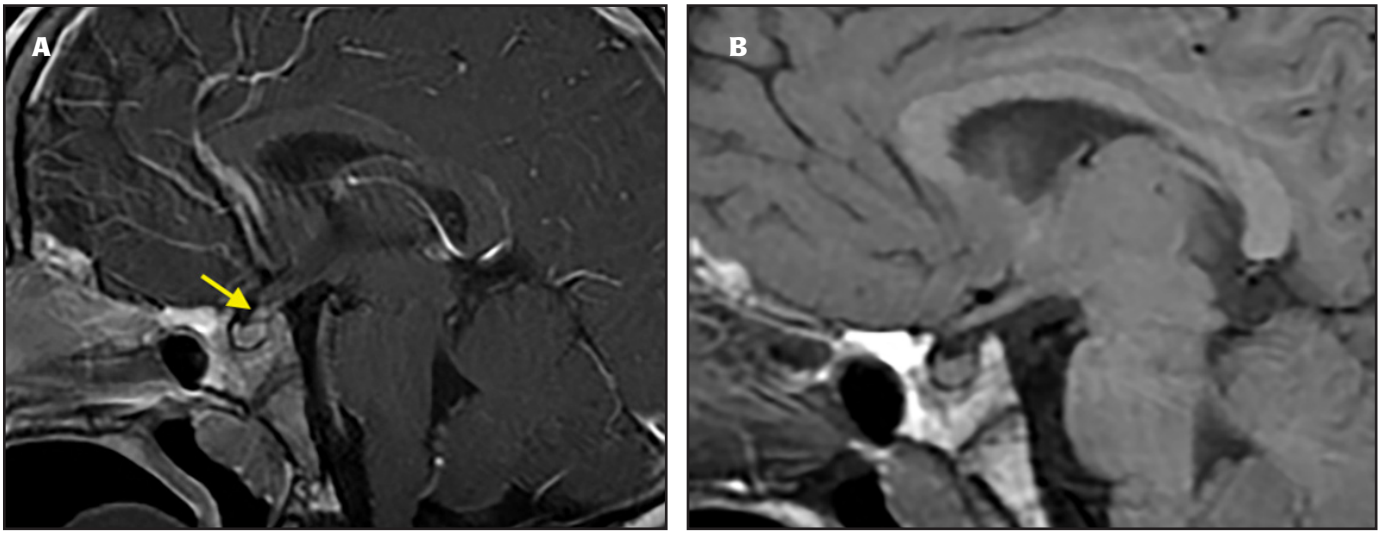

FIGURE 3. *Initial presentation of a different patient shows CNS involvement of LCH at 2 years of age. (A) Thickening and enhancement of the infundibulum (arrow) on T1 postcontrast GBCA imaging. (B) Precontrast T1 image shows absence of the posterior pituitary bright spot.*

LCH involvement of the central nervous system is less common; patients can be asymptomatic for years.[2,3] These findings can include an enhancing mass within the tuber cinereum or infundibulum and absence of the T1 hyperintense posterior pituitary bright spot (Figure 3). This is 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, best seen on T2/FLAIR sequences (Figure 4). 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 central nervous system, LCH is the most common. Other demyelinating processes such as acute disseminated encephalomyelitis (ADEM) 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 the pediatric population should include LCH in the differential. Familiarity with CNS involvement of LCH can aid neuroradiologists in making an accurate diagnosis.

References

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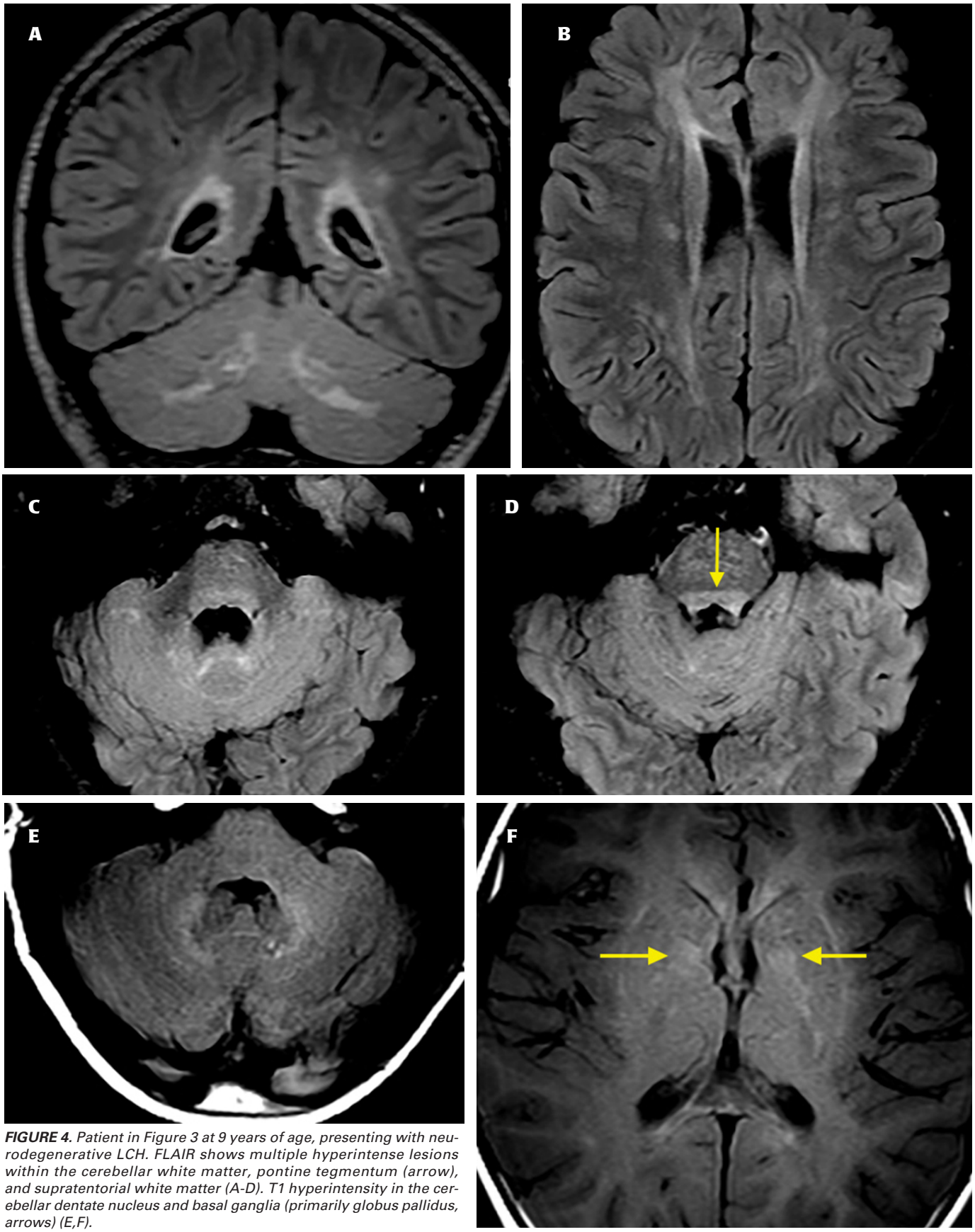

FIGURE 4. *Patient in Figure 3 at 9 years of age, presenting with neurodegenerative LCH. FLAIR shows multiple hyperintense lesions within the cerebellar white matter, pontine tegmentum (arrow), and supratentorial white matter (A-D). T1 hyperintensity in the cerebellar dentate nucleus and basal ganglia (primarily globus pallidus, arrows) (E,F).*