

20 year-old Male with Destructive Lesion of Orbital Roof

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Patient History

Mr. G.R. is a 20-year-old man with no significant past medical history. He presented in July of 2013 with two months of worsening pain over the right orbit. Periorbital edema was also present. Initial treatment with eye drops, antibiotics (erythromycin followed by cephalexin), and steroids (prednisone) failed to control his symptoms.

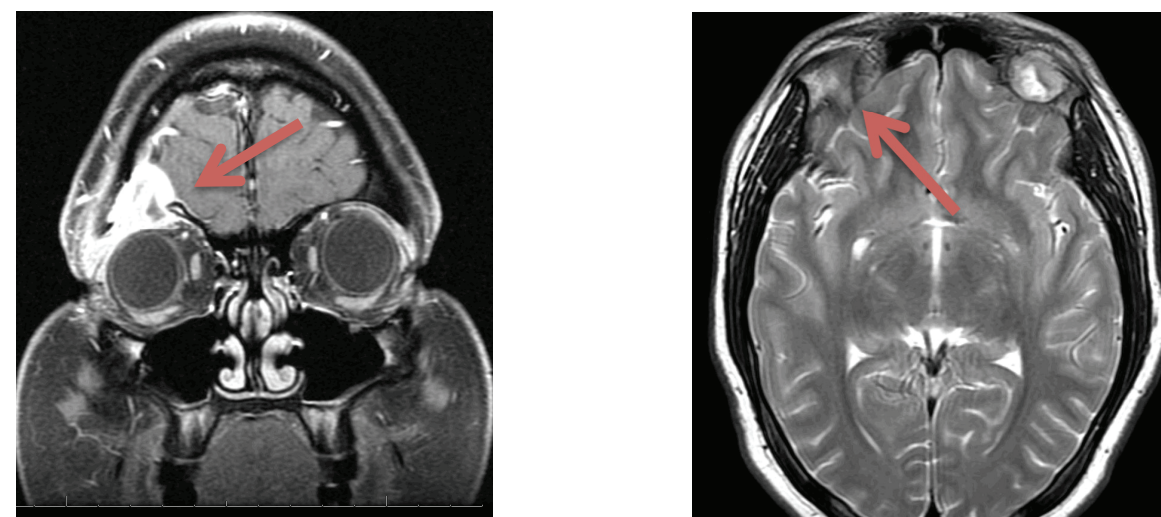
In October of 2013, CT and MRI scans revealed a permeative destructive lesion centered in the right orbital roof with extension into the superior orbit and the floor of the anterior cranial fossa. There was no abnormal enhancement in the visualized brain parenchyma.

The radiologic appearance of the mass suggested a differential diagnosis of a lymphoma, a metastasis, an eosinophilic histiocytosis, or an infection.

In November of 2013, the patient underwent a frontal craniotomy for resection of a 2.4 cm mass in the orbital rim. Langerhans cell histiocytosis was diagnosed by frozen section and confirmed by immunohistochemistry.

In December of 2013, the patient was seen for surgical follow-up at which time his pain, swelling, and erythema had completely resolved. There were no visual abnormalities present.

Magnetic Resonance Imaging



Figures 1 & 2. Pre-operative coronal T₁-weighted and horizontal T₂-weighted MRI images with contrast show an enhancing mass in the right orbit.

Microscopic Pathology and Immunohistochemical Stains

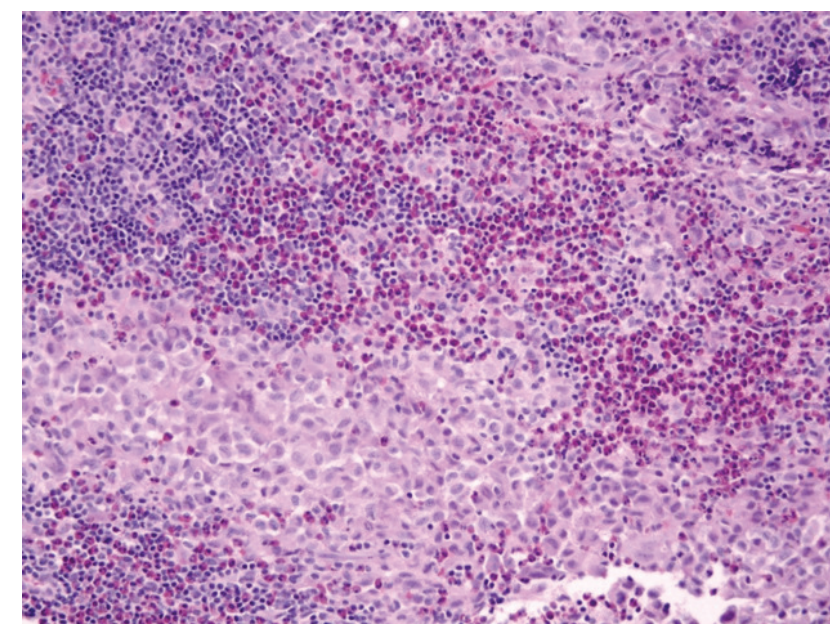


Figure 3. H&E stain of the tumor at 200x shows abundant eosinophils and a proliferation of histiocytes.

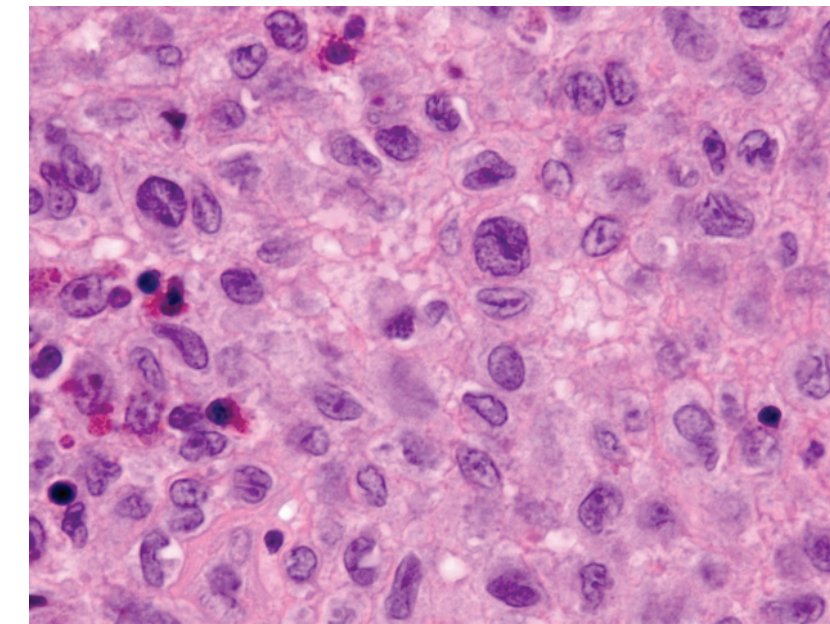


Figure 4. H&E stain of the tumor at 1000x reveals large histiocytic cells with grooved nuclei.

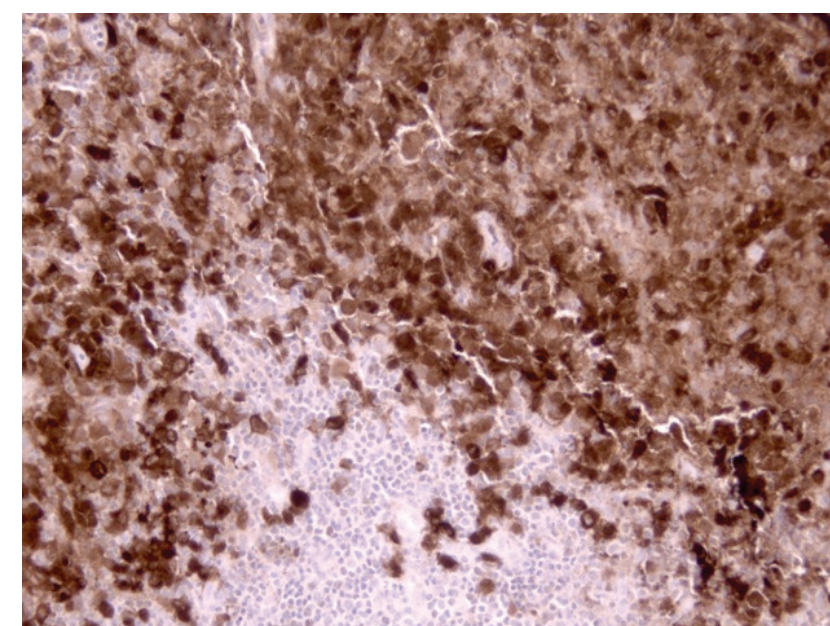


Figure 5. Immunohistochemistry for S100 stain at 200x

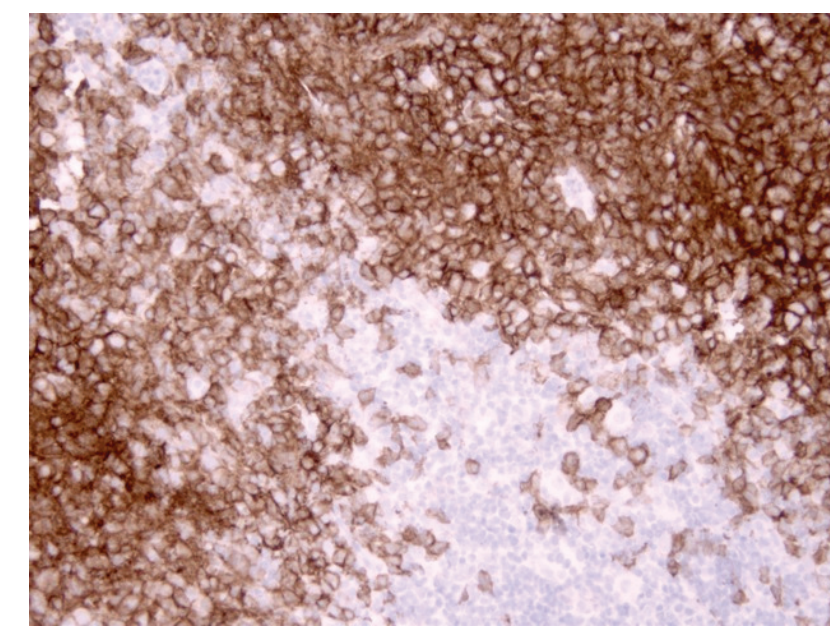


Figure 6. Immunohistochemistry for CD1a stain at 200x.

Diagnosis

The differential diagnosis for a 2.4 cm mass causing bony destruction of the superior orbital rim includes a histiocytic lesion, lymphoma, a metastasis, or an infection. Hematoxylin and Eosin and immunohistochemical stains such as CD1a, S-100 protein, and CD68 are helpful in distinguishing amongst these entities.

H&E stained sections revealed a histiocytic lesion with the presence of large cells with grooved nuclei in addition to an abundance of eosinophils. Erdheim-Chester disease, Langerhans cell histiocytosis, and Rosai-Dorfman disease are distinct histiocytic lesions that can be differentiated by their patterns of S-100, CD1a and CD68 expression. CD68 expression is a measure of lysosomal activity and marks all histiocytes. S-100 is a protein expressed in Langerhans cells and the histiocytes of Rosai-Dorfman disease, but not the histiocytes of Erdheim-Chester disease. CD1a is a specific marker of dendritic cells including Langerhans cells. Strong immunoreactivity for CD1a and S-100 along with low atypia and few mitoses on H&E stain confirmed the diagnosis of Langerhans cell histiocytosis.

Discussion

Langerhans cells are mononuclear phagocytes derived from precursor cells in the bone marrow. They are found in the epidermis, lymph nodes, spleen, thymus and mucosal tissues. Langerhans cells ingest, process, and present antigens to T lymphocytes. Langerhans cells have distinctive characteristics, including cytoplasmic tennis racquet-shaped "Birbeck granules" on electron microscopy.

Langerhans cell histiocytosis (LCH) represents a lesion of proliferating histiocytes in the background of numerous eosinophils. LCH can present either as an isolated bone lesion or a systemic process. An isolated lesion is more common in children and young adults. Surgical excision of solitary lesions is often curative.

References

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