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# Langerhans Cell Histiocytosis Involving the Sphenoid Sinus and Superior Orbital Fissure

J. S. Stromberg, A. M. Wang, T. E. Huang, F. A. Vicini, and P. A. Nowak

**Summary:** We report Langerhans cell histiocytosis involving the sphenoid sinus and extending to the orbital apex. MR and CT imaging, particularly with contrast, both were helpful in defining the extent of the lesion before treatment. MR was a useful examination for follow-up after treatment. Langerhans cell histiocytosis should be considered in the differential diagnosis of lesions of this region in the pediatric age group.

**Index terms:** Histiocytosis X; Paranasal sinuses, inflammation

The syndromes previously known as *histiocytosis X* recently have been reclassified as *Langerhans cell histiocytosis* based on contemporary pathologic examination (1). Radiologic findings in the central nervous system in patients with Langerhans cell histiocytosis have been described (2–9); however, there are few reports addressing the appearance of Langerhans cell histiocytosis in the sphenoid sinus and superior orbital fissure region. The use of magnetic resonance (MR), particularly with gadolinium, rarely has been characterized (10, 11). This report describes the radiologic findings of a case of Langerhans cell histiocytosis involving the sphenoid sinus and superior orbital fissure and reviews the literature regarding the imaging characteristics of Langerhans cell histiocytosis with central nervous system (CNS) involvement.

## Case Report

A 16-year-old boy presented with a 4-week history of progressive impaired vision of his left eye, particularly the nasal quadrants, associated with decreased light perception. A contrast-enhanced (Renograffin-60) computed tomography (CT) scan of the sinuses and orbits demonstrated a soft tissue mass in the sphenoid sinus with osteolytic bone destruction of the left lateral wall of the

sphenoid sinus (Fig 1A). The lesion displayed a mild degree of contrast enhancement and extended into the orbital apex. MR imaging demonstrated a mass filling the sphenoid sinus on the left extending into the orbital apex, which was isointense compared with brain parenchyma on T1-weighted imaging (500/15/2 [repetition time/echo time/excitations]), proton density-weighted imaging (3000/25/1) and T2-weighted imaging (3000/90/1). After intravenous administration of gadopentetate dimeglumine, the mass displayed a moderate degree of enhancement (Fig 1B). The neurohypophysis appeared normal on the axial T1-weighted postcontrast image and on the sagittal midline image (not shown). Clinically, the patient had no evidence of diabetes insipidus. A cerebral arteriogram done after left external carotid injection revealed a faint tumor blush. Tissue obtained from a transphenoidal biopsy demonstrated classical features of eosinophilic granuloma (now reclassified as Langerhans cell histiocytosis, class I). Infiltrates of large histiocytes with indented nuclei (Langerhans cells) accompanied by small eosinophils were seen (Fig 1C). Local external-beam radiotherapy was given to a total dose of 1000 cGy in 200 cGy/d fractions for 5 consecutive days with significant improvement of the visual acuity and light perception of the patient's left eye. MR imaging 2 months after completion of radiation therapy showed resolution of the previous soft tissue mass (Fig 1D); there was no abnormal enhancement after intravenous administration of gadopentetate dimeglumine.

## Discussion

The Langerhans cell histiocytoses are a group of clinically diverse syndromes that commonly can involve the CNS. The previous clinical syndromes, ranging in severity from solitary or multifocal eosinophilic granuloma of bone to chronic multiorgan involvement in Hand-Schuller-Christian disease to rapidly progressive and disseminated Letterer-Siwe disease, all have

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From the Departments of Radiation Oncology (J.S.S., F.A.V.), Diagnostic Radiology (A.M.W.), Pathology (T.E.H.), and Surgery (P.A.N.), William Beaumont Hospital, Royal Oak, Mich.

Address reprint requests to J. S. Stromberg, MD, Department of Radiation Oncology, William Beaumont Hospital, 3601 W Thirteen Mile Rd, Royal Oak, MI 48073.

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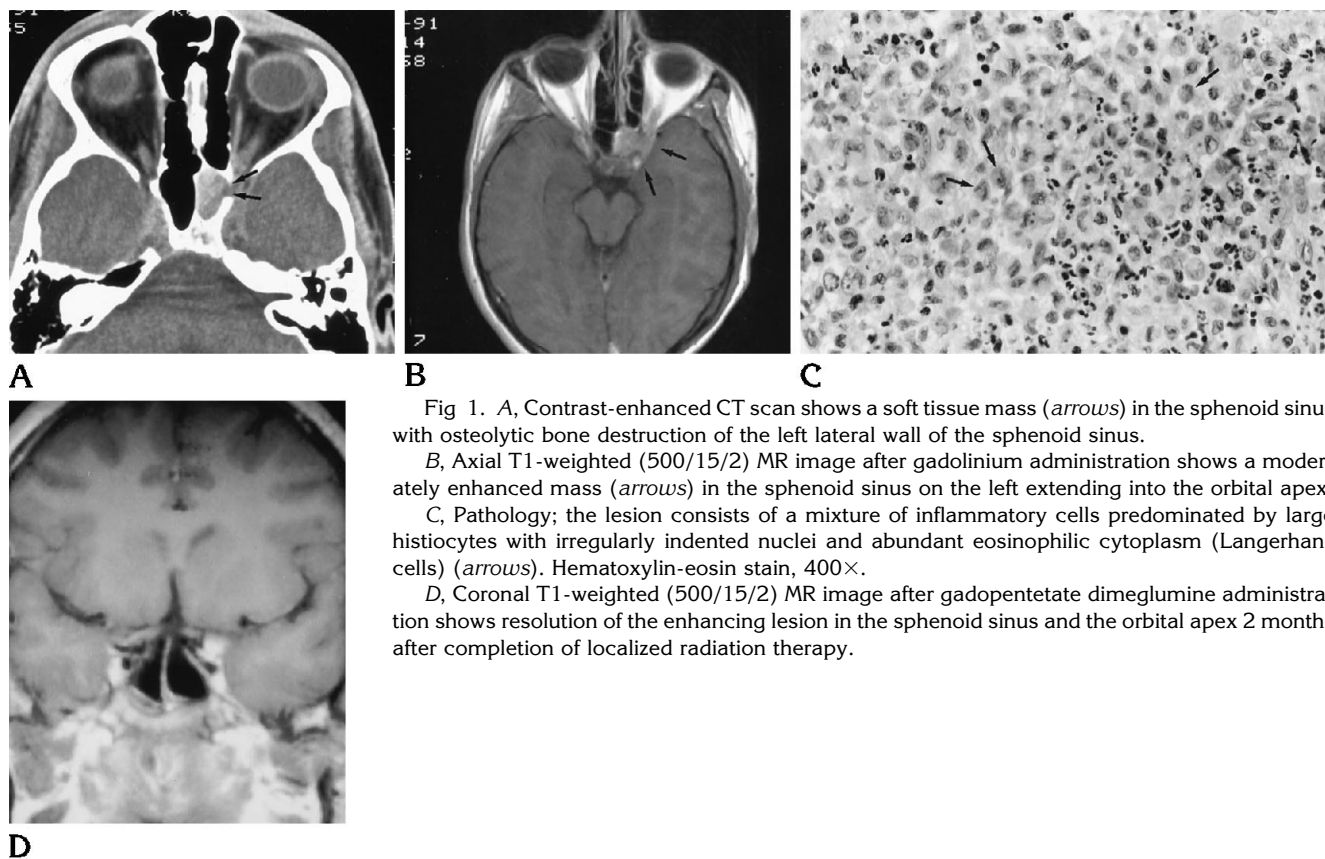


Fig 1. A, Contrast-enhanced CT scan shows a soft tissue mass (arrows) in the sphenoid sinus with osteolytic bone destruction of the left lateral wall of the sphenoid sinus.

B, Axial T1-weighted (500/15/2) MR image after gadolinium administration shows a moderately enhanced mass (arrows) in the sphenoid sinus on the left extending into the orbital apex.

C, Pathology; the lesion consists of a mixture of inflammatory cells predominated by large histiocytes with irregularly indented nuclei and abundant eosinophilic cytoplasm (Langerhans cells) (arrows). Hematoxylin-eosin stain, 400 $\times$ .

D, Coronal T1-weighted (500/15/2) MR image after gadopentetate dimeglumine administration shows resolution of the enhancing lesion in the sphenoid sinus and the orbital apex 2 months after completion of localized radiation therapy.

been reclassified as Langerhans cell histiocytosis, class I (1, 12). Langerhans cell histiocytosis commonly involves the CNS; however, rarely is this the only site. The most common CNS locations of involvement are the hypothalamic/pituitary axis and cerebellum (13, 14). Uncommonly, the meninges or parenchyma, usually the temporal or occipital lobes, may be involved either primarily or from direct extension of a calvarial lesion to the epidural space (2). Diabetes insipidus is the most common endocrine manifestation of Langerhans cell histiocytosis and is attributable to decreased secretion of antidiuretic hormone. The prevalence of diabetes insipidus varies. However, it was recently reported in 23% of patients with Langerhans cell histiocytosis and was observed more commonly in association with multisystem disease, especially that with skull and orbit involvement (15).

Although the clinical and pathologic involvement of the CNS has been described, the CNS radiologic findings of Langerhans cell histiocytosis have not been extensively reported. We have reported the MR and CT findings of a case of Langerhans cell histiocytosis involving the

extradural location of the sphenoid sinus and superior orbital fissure. Analysis of the MR features of this case revealed the lesion to be best evaluated on a T1-weighted image with administration of gadopentetate dimeglumine, which demonstrated a moderate degree of enhancement of the lesion and better delineated the boundaries of the mass. The lesion in our report was isointense to surrounding white matter on T1-weighted, T2-weighted, and proton density-weighted imaging. These findings are generally consistent with other findings reported in the literature (5, 11), although often T2-weighted images in extrahypothalamic/parenchymal sites of Langerhans cell histiocytosis involvement will show increased signal intensity (2, 5, 6, 10). The extradural location of the lesion described in this report may account for the lack of increased T2 signal intensity. The histiocytosis lesion sampled in this case report revealed moderate vascularity on histologic examination, which correlates well with the radiographic enhancement observed.

CNS Langerhans cell histiocytosis lesions involving the hypothalamus/pituitary stalk region with associated diabetes insipidus require spe-

Typical CNS imaging features in patients with Langerhans cell histiocytosis

	Extrahypothalamic/Parenchymal	Hypothalamic/Pituitary
CT without contrast	Isodense to hypderdense mass	Isodense to hypderdense mass
CT with contrast	Uniform enhancement	Uniform enhancement
MR without gadopentetate dimeglumine	Lesions isointense to hypointense on T1-weighted images; lesions often hyperintense on T2-weighted images	Can see loss of normal high-intensity signal on T1-weighted images in patients with diabetes insipidus
MR with gadopentetate dimeglumine	Moderate to strong enhancement usually seen	Strong enhancement of lesion seen in patients with diabetes insipidus

cial attention. Normally, MR demonstrates increased signal intensity in the posterior lobe of the pituitary on T1-weighted images (16, 17). When Langerhans cell histiocytosis arises in the infundibulum, loss of this high signal can occur and may represent a disruption of the integrity of the hypothalamic hypophyseal tract (2, 5, 10, 18). It is important in such cases to search for other findings such as enlargement of the infundibulum and associated clinical diabetes insipidus. Gadopentetate dimeglumine administration also may be particularly helpful in revealing abnormalities in the hypothalamic and pituitary regions in patients with Langerhans cell histiocytosis and deficient antidiuretic hormone (10, 11). In one recent report, all five patients with chiasmic-hypothalamic or posterior pituitary abnormalities and deficient antidiuretic hormone demonstrated lack of the usual increased signal in the posterior pituitary on T1-weighted images. Three of these five patients underwent intravenous administration of gadopentetate dimeglumine, and all three demonstrated enhancement of the involved areas (10). Although the normal infundibulum also enhances after gadolinium administration, it usually does not do so to the extent seen with Langerhans cell histiocytosis and diabetes insipidus. Gadopentetate dimeglumine administration also has been previously reported to result in enhancement of involved extrahypothalamic/parenchymal areas of CNS histiocytosis (10), and our report is in agreement with these findings. The Table outlines the CT and MR features of CNS histiocytosis in both the extrahypothalamic/parenchymal and hypothalamic/pituitary regions.

MR imaging may also be a useful modality for assessment of treatment response. The patient presented in our report demonstrated complete resolution of his mass as assessed by MR 2 months after the completion of radiation ther-

apy. Other reports also have suggested MR as a useful modality for assessment of treatment response. CT and MR both are useful studies for delineating the boundaries of disease involvement for radiation therapy treatment planning purposes (6, 19). Both contrast-enhanced CT images and gadolinium-enhanced MR images provide improved location of anatomic abnormalities (10).

CT has an important role in demonstrating CNS histiocytosis involvement. It can be particularly helpful in revealing bone destruction as demonstrated in this case report. Early lesions, however, often can be difficult to demonstrate on CT, because they often are of similar density to surrounding brain and can cause little mass effect without much contrast enhancement. MR has been reported to be superior to CT in demonstrating earlier lesions because of its improved soft tissue contrast and potential gadolinium enhancement (2, 3). MR may also be the study of choice in young children with Langerhans cell histiocytosis and associated diabetes insipidus, because these lesions can be particularly difficult to demonstrate on CT.

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