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W K Erly, R F Carmody and R M Dryden

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Orbital Histiocytosis X

William K. Erly, Raymond F. Carmody, and Robert M. Dryden

Summary: In three patients with histiocytosis X of bone with orbital involvement, CT scans were reviewed. Consistent findings included a destructive lesion of the lateral wall of the orbit with a large soft-tissue component that extended into the extraconal space, the ocular adnexa, and the infratemporal fossa. The greater wing of the sphenoid was eroded in all cases, with epidural extension into the middle cranial fossa. Cavernous sinus involvement and a second bone lesion were seen in two patients.

Index terms: Histiocytosis X; Orbits, diseases

Eosinophilic granuloma, Hand-Schuller-Christian disease, and Letterer-Siwe disease are three disorders within the general disease entity known as *histiocytosis X*, or more recently, *Langerhans cell histiocytosis* of bone. Although Langerhans cell histiocytosis is a rare entity, in patients with Langerhans cell histiocytosis, orbital histiocytosis is not uncommon and is generally associated with additional bone or soft-tissue lesions. Computed tomography (CT) scans of the orbit in three children with histiocytosis X were reviewed, and the findings are described.

Methods

The records of the radiology and pathology departments from 1981 to the present were reviewed for patients with a diagnosis of histiocytosis X in whom CT scans of the orbit were available. Twenty-five cases were diagnosed during this time period; 3 of these had Langerhans cell histiocytosis of the orbit.

Scans were performed using intravenous contrast and 2-mm-thick contiguous axial sections in two patients, and 2-mm-thick axial images at 3-mm section intervals in the third. The images were reviewed by two neuroradiologists.

Case Reports

Case 1

A 2-year-old girl was in good health until 3 weeks before her initial evaluation, at which time nasal conges-

tion developed. Subsequently, the parents noticed redness and bulging of her right eye and sought medical attention. On physical examination, obvious right-sided exophthalmos was found. The extraocular muscles were intact. She had no palpable adenopathy. The abdomen was nontender and without palpable mass. Laboratory findings all were normal.

Axial CT demonstrated a 2.5-cm enhancing mass centered in the lateral wall of the right orbit, with extension into the infratemporal fossa and into the anterior portion of the middle cranial fossa. Additional extension was present in the region of the lacrimal gland with soft-tissue thickening around the lateral margin of the globe and into the preseptal soft tissues. The preseptal disease extended along the fascial plane lateral to the frontozygomatic arch. The lateral rectus muscle could not be shown to be distinct from the mass (Fig 1).

Case 2

A 2-year-old girl had a brief history of right orbital proptosis. She was otherwise healthy. Initial physical examination revealed 6 mm of right orbital proptosis and a mild right temporal fullness. The remainder of the examination was unremarkable. Laboratory findings all were within normal limits.

CT of the brain and orbit showed a 3-cm enhancing mass centered in the right orbital wall with extension into the orbit, the middle cranial fossa, and the infratemporal fossa. There was extension into the superior orbital fissure and anterior cavernous sinus as well. A second, smaller lesion was identified in the squamous portion of the left temporal bone (Fig 2). CT findings of the chest and abdomen were normal.

Case 3

A 21-month-old girl had a left parietal scalp mass that had gradually enlarged over the 5 months before her initial presentation. The left eye became red and swollen 3 months before presentation. She was below the fifth percentile in height and weight, which was felt to be secondary to poor nutrition.

CT of the brain demonstrated a 2.5 × 5.0-cm mass centered in the left parietal bone. There was a large soft

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From the Department of Radiology, University of Arizona, Tucson (W.K.E., R.F.C.). Dr Dryden is in private practice in Tucson.

Address reprint requests to William K. Erly, MD, Department of Radiology, Grossmont Hospital, 5565 Grossmont Center Dr, La Mesa, CA 91942.

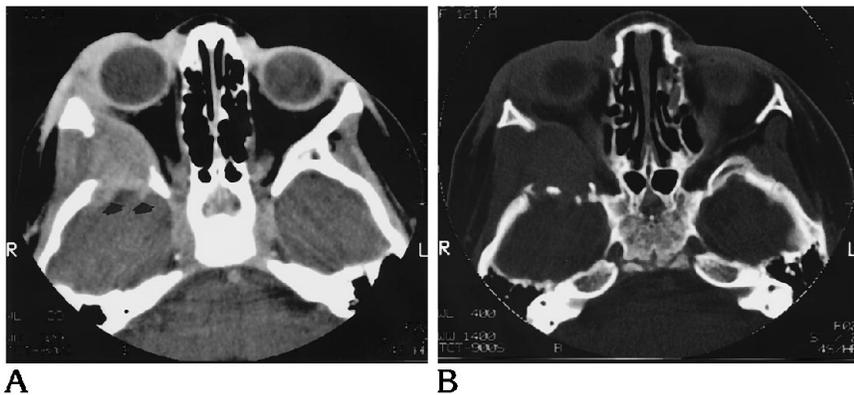


Fig 1. Soft-tissue mass centered in the lateral wall of the right orbit. Extension into the middle cranial fossa (*arrows*) and infratemporal fossa is seen. The lateral rectus muscle is not distinct from the mass.

tissue component with minimal intracranial extension. A second mass was present centered in the lateral wall of the left orbit. The mass extended into the middle cranial fossa, orbit, infratemporal fossa, lateral aspect of the clivus, and cavernous sinus. Within the orbit, the region of the lacrimal gland was involved. There was extension into the preseptal soft tissue (Fig 3).

Results

All three patients were approximately 2 years old, of Hispanic origin, and presented with proptosis. The following findings were common to all three cases: The mass was centered in the lateral wall of the orbit with erosion of the greater wing of the sphenoid. A large soft-tissue component was present with extension into the extraconal space of the orbit, the infratemporal fossa, and the middle cranial fossa. The intraorbital extension bowed the lateral rectus muscles medially. There was proptosis and thickening of the orbital adnexa. The lateral rectus muscle could not be separated by a distinct fat plane from the tumor. Infiltration of the muscle could therefore not be excluded.

In cases 2 and 3, the tumor extended to the orbital apex, with mild dilatation of the superior ophthalmic vein. In addition, epidural tumor extension to the cavernous sinus and erosion of the anterolateral aspect of the clivus was present. Second lesions were apparent in both cases.

None of the patients demonstrated definite extension into the intraconal space of the orbit. No parenchymal lesions of the brain were identified. The bone margins were indistinct in all cases, allowing the appearance to be confused with a more aggressive tumor. No calcification or periosteal new bone formation was present.

Discussion

Langerhans cell histiocytosis, previously called *histiocytosis X*, describes a spectrum of disease ranging from isolated eosinophilic granuloma of bone to disseminated Letterer-Siwe disease. Eosinophilic granuloma is a relatively uncommon entity, accounting for only 1% of all tumorlike lesions of bone (1). In addition, in-

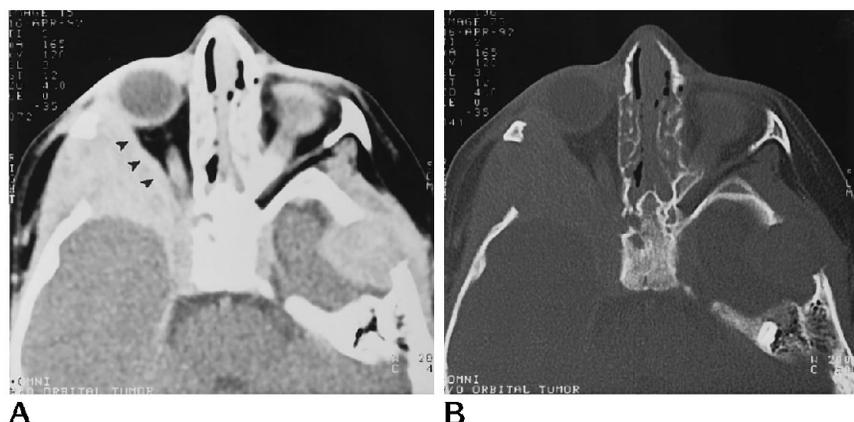


Fig 2. A, Soft-tissue mass in the lateral wall of the right orbit. The lateral rectus is displaced medially (*arrows*).

B, Extension into the clivus and middle cranial fossa is seen. A second lesion is apparent in the left squamous temporal bone.

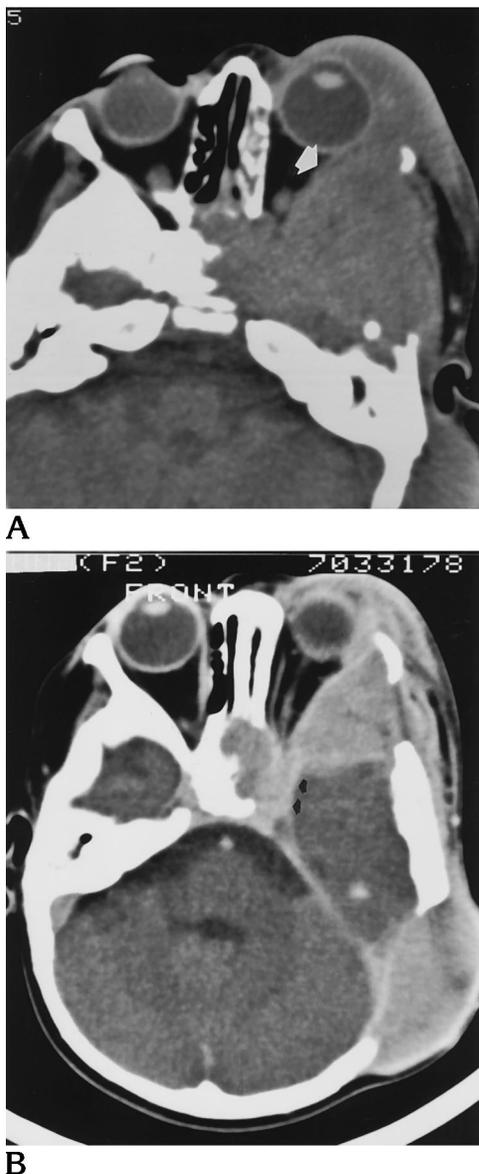


Fig 3. A, Large soft-tissue mass centered in the lateral wall of the orbit. The mass abuts the lateral rectus muscle (*white arrow*). B, Higher cut demonstrating second lesion in the parietal bone. Cavernous sinus extension is seen (*black arrows*).

involvement of the orbit by Langerhans cell histiocytosis is uncommon and accounts for fewer than 1% of all orbital tumors (2). However, when the orbit is involved, it is usually the superolateral aspect (3). Radiologic evidence of disease may be present without clinical signs (2). However, if clinical signs are present, a lytic lesion of the orbital wall usually is found. Proptosis is the most common symptom, but dislocation of the globe has been reported (4).

In our three cases, the orbital mass was uniformly extraconal, apparently arising from the

orbital wall. Despite the medial bowing of the lateral rectus, no intraconal or intraocular involvement was seen. In all cases, the lesion extended into the infratemporal fossa as well as into the middle cranial fossa. Previous reports have described intraocular involvement and involvement of the brain parenchyma, but these are rare findings. Orbital masses are usually extraconal and are thought to be of bone origin (2, 5).

The cause of Langerhans cell histiocytosis is uncertain. Currently, it is considered a disease of immune regulation with histiocytic proliferation and granuloma formation. The Langerhans cell is present in all forms of the disease, from isolated eosinophilic granuloma to Letterer-Siwe disease. The Langerhans cell is a dendritic cell of the epidermis which is characterized by a unique organelle, the Langerhans, or Birbeck, granule (6, 7).

The prognosis of Langerhans cell histiocytosis varies with the age of the patient and with the extent of the disease. Generally, the younger the patient at presentation, or the greater the extent of the disease, the worse the prognosis. Persons with organ dysfunction have the worst prognosis (6, 8).

Localized Langerhans cell histiocytosis, or eosinophilic granuloma, is defined as those cases in which the disease is confined to the bone or lung. These lesions favor the flat bones but may occur at any site. Eosinophilic granuloma is the most common form of Langerhans cell histiocytosis and carries the best prognosis. These patients often do well with local therapy, such as curettage with or without low-dose radiation (300 to 600 cGy) (9).

Hand-Schuller-Christian disease, or chronic recurring Langerhans cell histiocytosis, is classically described as the triad of diabetes insipidus, exophthalmos, and destructive bone lesions. This triad occurs in only 10% to 15% of patients with Langerhans cell histiocytosis (9).

Letterer-Siwe disease is the acute disseminated form of Langerhans cell histiocytosis and accounts for approximately 10% of Langerhans cell histiocytosis cases. Clinically, these patients are younger than 2 years of age at presentation, and have fever, hepatosplenomegaly, thrombocytopenia, anemia, and a skin rash. Death usually occurs within 2 years of diagnosis (10).

Of the 25 cases of Langerhans cell histiocytosis we reviewed, 3 (12%) had orbital involve-

ment. The overall incidence of orbital Langerhans cell histiocytosis is estimated to be 20%, most commonly as eosinophilic granuloma (2).

The diagnosis of Langerhans cell histiocytosis usually can be made by biopsy of tissue from other sites. However, in patients with primary orbital involvement, other lesions, such as osteomyelitis, metastatic neuroblastoma, Ewing sarcoma, chloroma, lymphoma, and rhabdomyosarcoma need to be excluded. Although the CT appearance may look aggressive, benign Langerhans cell histiocytosis should be considered in the differential diagnosis of primary orbital bone lesions in a child.

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