Langerhans cell histiocytosis (LCH) is characterized by the accumulation of pathologic Langerhans cells, a type of histiocyte. Classically, LCH is grouped into three clinical syndromes: eosinophilic granuloma, Hand-Schuller-Christian syndrome, and Letterer-Siwe syndrome (1).

LCH can affect patients of any age from birth to adulthood, peaking during the first three years of life (2).

Orbital involvement of LCH is uncommon and accounts for less than one percent of all orbital tumors. Orbital LCH is found in about 20% of all LCH cases, commonly presenting as eosinophilic granuloma (3).

The role of CT in the evaluation of LCH has been described for pulmonary LCH, the pituitary stalk, and bony lesions involving the temporal bone or orbit (4-8). In order to describe the characteristic findings of LCH involving the orbit, we reviewed CT scans in children with orbital LCH.

Materials and Methods

During a four-year period we encountered six patients in whom LCH involving the orbit had been pathologically proven by incisional biopsy. Four were girls and t-
wo were boys, and all were aged between one and 3.4 (mean, 2.1) years.

Pre- and postcontrast CT scans obtained prior to specific treatment were available in all cases. Follow-up CT scans were available in five cases after chemotherapy (n = 4) or chemotherapy with radiation therapy (n = 1). In one patient, follow-up involved MRI imaging.

CT scanning involved the use of HiSpeed scanners (General Electric Medical System, Milwaukee, Wis), employing the parameters for routine orbit CT: 120 kVp, 200 mA, and slice thickness 3.0-5.0 mm. Postcontrast CT scans were obtained after bolus injection of contrast media (2 cc/kg body weight).

Soft tissue masses were evaluated for size, calcification, attenuation value, homogeneity, and enhancement pattern, while bone destruction was evaluated in terms of location, margin characteristics, and periosteal reaction. All images were reviewed by two radiologists who reached a consensus.

From the clinical record we obtained information about symptoms of presentation, treatment, and other organic or systemic involvement.

Results

Clinical data and CT findings are detailed in Table 1

### Table 1. Clinical Manifestations and Imaging Follow-up after Treatment in Six Patients with Orbital Langerhans Cell Histiocytosis

<table>
<thead>
<tr>
<th>Case No</th>
<th>Sex/Age (years)</th>
<th>Presenting Sign</th>
<th>Treatment Modality/Duration (weeks)</th>
<th>First follow-up Interval (months)</th>
<th>Modality</th>
<th>Findings size of bone</th>
<th>Second follow-up Interval (months)</th>
<th>Modality</th>
<th>Findings</th>
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<tbody>
<tr>
<td>1</td>
<td>F/1.5</td>
<td>Exophthalmos</td>
<td>CTx/44</td>
<td>3</td>
<td>CT</td>
<td>↓ R</td>
<td>12</td>
<td>CT</td>
<td>N</td>
</tr>
<tr>
<td>2</td>
<td>F/3</td>
<td>Eyelid swelling</td>
<td>CTx/40</td>
<td>2</td>
<td>CT</td>
<td>↓ R</td>
<td>10</td>
<td>CT</td>
<td>N</td>
</tr>
<tr>
<td>3</td>
<td>F/1</td>
<td>Exophthalmos</td>
<td>CTx/38</td>
<td>3</td>
<td>CT</td>
<td>↓ R</td>
<td>9</td>
<td>MR</td>
<td>Minimal</td>
</tr>
<tr>
<td>4</td>
<td>F/3</td>
<td>Poor oral intake</td>
<td>CTx+radiation/36</td>
<td>5</td>
<td>MR</td>
<td>↓ R</td>
<td>10</td>
<td>MR</td>
<td>N</td>
</tr>
<tr>
<td>5</td>
<td>M/2</td>
<td>Exophthalmos</td>
<td>CTx/46</td>
<td>2</td>
<td>CT</td>
<td>↓ N</td>
<td>12</td>
<td>CT</td>
<td>N</td>
</tr>
<tr>
<td>6</td>
<td>M/3.4</td>
<td>Exophthalmos</td>
<td>CTx/70</td>
<td>2</td>
<td>CT</td>
<td>↓ R</td>
<td>NP</td>
<td>NP</td>
<td>NP</td>
</tr>
</tbody>
</table>

↓, decreased; R, residual bone lesion; N, no residual bone lesion; Minimal, minimal residual bone lesion; NP, not performed; CTx, chemotherapy

Fig. 1. Case 1. 1.5-year-old girl presented with right exophthalmos. Unenhanced CT (A) and enhanced CT (B) show a large soft tissue mass (arrows) with bone destruction involving lateral wall of right orbit. Minimal peripheral enhancement is seen in B. There is intracranial extension (arrowheads) without intraconal extension. After 3 months, follow-up enhanced CT (C) shows marked improvement of soft tissue mass. Follow-up CT (D) after one year shows bone remodeling and reossification without residual soft tissue mass.
and 2.

The most common presenting symptom was exophthalmos (n = 4) (Table 1). In two cases, redness and a sense of hotness in the periorbital region mimicked the symptoms and signs of periorbital cellulitis. Three patients showed other organic involvement including hepatosplenomegaly (n = 2) and suprasellar mass and rib destruction (n = 1). Final diagnoses were eosinophilic granuloma in four patients, Hand-Schulier-Christian disease with diabetes insipidus in one (case 4), and Letterer-Siwe disease with hepatomegaly, pancytopenia, and skin rash in one (case 3).

In all cases, CT revealed irregular bone destruction, with soft tissue mass (Figs. 1 and 2). This latter showed an average absorption value of 44 Hounsfield units (HU) and mild to moderate enhancement following contrast medium administration, with an average of 74 HU (Table 2). In three cases the enhancement pattern was inhomogeneous, with peripheral rim enhancement. In three cases the soft tissue margins were poorly defined, and in three they were well demarcated. In all cases, bony margins were irregular or smooth, and well demarcated. No calcification or periosteal new bone formation was seen, though in one case there was associated bony expansion.

The lateral or superolateral aspect of the bony orbit was most commonly involved (Table 2). Large soft tissue masses extended to the extraconal space of the orbit, the infratemporal fossa, middle cranial fossa, frontal sinus, or maxillary sinus. In cases with superolateral or-

<table>
<thead>
<tr>
<th>Case</th>
<th>Location</th>
<th>Intracranial</th>
<th>Size</th>
<th>Attenuation (H.U.)</th>
<th>Homogeneity*</th>
<th>Margin</th>
<th>Bony expansion</th>
<th>Periosteal Calciﬁcation</th>
</tr>
</thead>
<tbody>
<tr>
<td>No</td>
<td>orbital wall extension</td>
<td>(mm)</td>
<td>pre</td>
<td>post</td>
<td>pre</td>
<td>post</td>
<td>Soft tissue</td>
<td>Bone</td>
</tr>
<tr>
<td>1</td>
<td>Lateral</td>
<td>+</td>
<td>27</td>
<td>41</td>
<td>61</td>
<td>H</td>
<td>well</td>
<td>well, smooth</td>
</tr>
<tr>
<td>2</td>
<td>Superolateral</td>
<td>+</td>
<td>46</td>
<td>45</td>
<td>94</td>
<td>I</td>
<td>poor</td>
<td>well, irregular</td>
</tr>
<tr>
<td>3</td>
<td>Lateral</td>
<td>+</td>
<td>51</td>
<td>40</td>
<td>84</td>
<td>I</td>
<td>poor</td>
<td>well, irregular</td>
</tr>
<tr>
<td>4</td>
<td>Superolateral</td>
<td>-</td>
<td>multiple</td>
<td>47</td>
<td>75</td>
<td>H</td>
<td>well</td>
<td>well, irregular</td>
</tr>
<tr>
<td>5</td>
<td>Inferior</td>
<td>-</td>
<td>22</td>
<td>NP</td>
<td>NP</td>
<td>H</td>
<td>poor</td>
<td>well, irregular</td>
</tr>
<tr>
<td>6</td>
<td>Inferior</td>
<td>-</td>
<td>20</td>
<td>45</td>
<td>56</td>
<td>I</td>
<td>H</td>
<td>well, irregular</td>
</tr>
</tbody>
</table>

Note: CT Findings* H, homogeneous; I, inhomogeneous; NP, not performed

Fig. 2. Case 4. Three-year-old girl presented with poor oral intake. MR T1-weighted image (A) and T2-weighted image (B) show iso-signal intensity masses with bone destruction (arrowheads) involving superolateral wall of both orbits. Postcontrast T1-weighted coronal MR image (C) shows marked enhancement of soft tissue mass. Enhanced coronal CT (D) shows irregular bone destruction with soft tissue masses involving superolateral orbital wall bilaterally.
bital mass, this displaced the lateral rectus muscle medially without intraconal extension. Intracranial extension with epidural mass formation was found in three cases (Figs. 1 and 2).

Follow-up CT scans (n = 5) and MR images (n = 1) demonstrated that the soft tissue masses had almost completely disappeared, with bone remodeling and reossification (Figs. 1 and 2). The mass appeared isointense to muscles on T1- and T2-weighted images, with diffuse enhancement on postcontrast T1-weighted images (Fig. 2).

**Discussion**

Histiocytosis is the term used to denote a group of disorders whose common feature is an idiopathic proliferation of histiocytes. The histiocytic cell common to this constellation of lesions is known as the Langerhans cell (9). Hence the more pathologically precise term ‘Langerhans cell histiocytosis’ has been adopted (10).

Localized Langerhans cell histiocytosis, or eosinophilic granuloma is the most common form of LCH, and its prognosis is the most promising. These patients often do well with local therapy, such as curettage with or without low-dose radiation (300 or 600 cGy) (11).

Hand-Schuller-Christian disease, or chronic recurrent Langerhans cell histiocytosis, is classically described as a triad of diabetes insipidus, exophthalmos, and destructive bone lesion, though the three conditions co-occur in only 10% to 15% of patients with LCH (11). In our study, one patient without exophthalmos showed clinical symptoms and signs of diabetes insipidus as well as bony lesions (case 4).

Letterer-Siwe disease is the acute disseminated form of LCH and accounts for approximately 10% of LCH cases. Clinically, patients are aged less than two years at presentation, and their symptoms include fever, hepatosplenomegaly, thrombocytopenia, anemia, and a skin rash. Death usually occurs within two years of diagnosis.

The conventional radiographic findings of eosinophilic granuloma are well known (12). CT more clearly demonstrates the presence of an osteolytic lesion and associated soft tissue mass, and in addition, may help differentiate LCH from other orbital diseases including osteomyelitis, lymphoma, leukemia, retinoblastoma, rhabdomyosarcoma, or metastatic neuroblastoma. Malignant tumors often cause irregular and permissive bone destruction, and a malignant-type periosteal reaction. In our cases, on the other hand, the bone destruction occurring in orbital LCH was clearly marginated and there was no periosteal reaction. In addition, the soft tissue mass in LCH was more homogeneous and well-marginated than in cases of malignant tumor.

Previous reports of orbital LCH have described the replacement of a focal bony defect by soft tissue lesion, without calcification or periosteal reaction, as in our cases (7, 8). The bony margins of LCH lesions were reported to be indistinct, mimicking a more aggressive tumor (8). In our cases, on the other hand, demarcated bony margins were clearly visible in all cases. Following bolus injection of the contrast medium, moderate inhomogeneous enhancement of the soft tissue mass was seen, with an average absorption value of 74 HU; this was slightly higher than in previously described cases. MR images are expected to demonstrate marked enhancement after intravenous administration of the contrast agent (13, 14). In one of our cases, the soft tissue mass showed iso-signal intensity on T1-weighted and T2-weighted MR images and marked enhancement after intravenous administration of the contrast agent.

In terms of tumor location, the superolateral bony orbit was most commonly involved, followed by the inferior bony orbit. No cases showed involvement of the medial orbital wall. To determine whether a specific site is more frequently involved than the rest of the bony orbit, further study involving a large series of orbital LCH cases may be needed.

In summary, the characteristic CT findings of orbital LCH were an enhanced soft tissue mass, irregular but clearly demarcated bone destruction, and an absence of calcification or periosteal reaction. Follow-up imaging revealed marked interval decreases in mass size as well as bone remodeling and reossification.

**References**

1. Lichtenstein L. Histiocytosis X: Integration of eosinophilic granuloma of bone” Letterer-Siwe disease” and “Schuller-Christian Disease” as related manifestations of a single nosologic entity. Arch Pathol 1953;56:84-102

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17TH SYMPOSIUM FOR COMPUTER APPLICATIONS IN RADIOLOGY (SCAR) (2000 6-9 June)  
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ANNUAL MEETING AMERICAN SOCIETY OF RADIOLOGIC TECHNOLOGISTS (2000 3-8 June)  
venue: Convention Center, Albuquerque, USA.  
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venue: Hotel Lotte World, Seoul, Korea.  
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venue: Four Seasons Resort Hotel, Las Vegas, Nevada, USA.  
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venue: Jacob K. Javits Conv. Ctr., New York, New York, USA.  
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63RD ANNUAL SCIENTIFIC MEETING OF THE CANADIAN ASSOCIATION OF RADIOLOGISTS (2000 10-14 June)  
venue: Metro Toronto Conv. Ctr., Toronto, ON, Canada.  
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87TH CONGRESS OF THE SWISS SOCIETY FOR MEDICAL RADIOLOGY(SSMR) (2000 11-13 June)  
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THE 5TH INTERNATIONAL WORKSHOP ON DIGITAL MAMMOGRAPHY (2000 11-14 June)  
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6TH ANNUAL INTERNATIONAL CONFERENCE ON FUNCTIONAL MAPPING OF THE HUMAN BRAIN OF THE ORGANIZATION FOR HUMAN BRAIN MAPPING (2000 12-16 June)  
venue: San Antonio, TX, USA.  
contact: Sylvia Gonzalez, UT Health Science Center SA, 7703 Floyd Curl Drive, San Antonio, TX 78284-6239, USA.  
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TENTH ANNUAL CONFERENCE ON MUSCULOSKELETAL ULTRASOUND (2000 12-16 June)  
venue: Orchard Hotel, Singapore  
contact: Manix van Holsbeek, MD, Michael A. Di Pietro, M.D, J. Antonio Bouffard, MD 3588 Plymouth Road, PMB#499 Ann Arbor, MI 48105 USA  
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8TH ANNUAL MEETING EUROPEAN SOCIETY OF THORACIC IMAGING(ESTI) (2000 18-20 June)  
venue: Salzburg, Austria.  
contact: Prof. Dr. D. Hahn, Institut f. Röntgendiagnostik, Josef-Schneider-Strasse 2, D-97080 Würzburg, Germany.  
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