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CT Findings of Orbital Langerhans Cell Histiocytosis[1]

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Purpose: To evaluate the CT findings in patients with Langerhans cell histiocytosis (LCH) involving the orbit.

Materials and Methods: Orbital CT scans of six children with pathologically proven LCH were retrospectively analyzed. Follow-up CT (n = 5) and MR (n = 1) imaging findings were also reviewed.

Results: Initial CT scans revealed varying degree of bone destruction with soft-tissue masses, and on nonenhanced images the mean attenuation value was 44 Hounsfield units (HU). All masses showed mild to moderate enhancement with a mean attenuation value of 74 HU. The bony margins abutting onto soft tissue masses were irregular but clearly demarcated. No evidence of calcification or periosteal reaction was noted. Suprasellar mass and rib involvement was noted in one patient and hepatosplenomegaly in two. Follow-up CT and MR images showed that the soft tissue masses were almost completely resolved, with bone remodeling and reossification.

Conclusion: A soft tissue mass with irregular but clearly demarcated bone destruction is thought to be a characteristic finding of LCH involving the orbit. Follow-up images after treatment showed bone remodeling and reossification.

Index words : Histiocytosis
Neoplasms, in infants and children
Orbit, CT

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, 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-

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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 MR 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

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

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

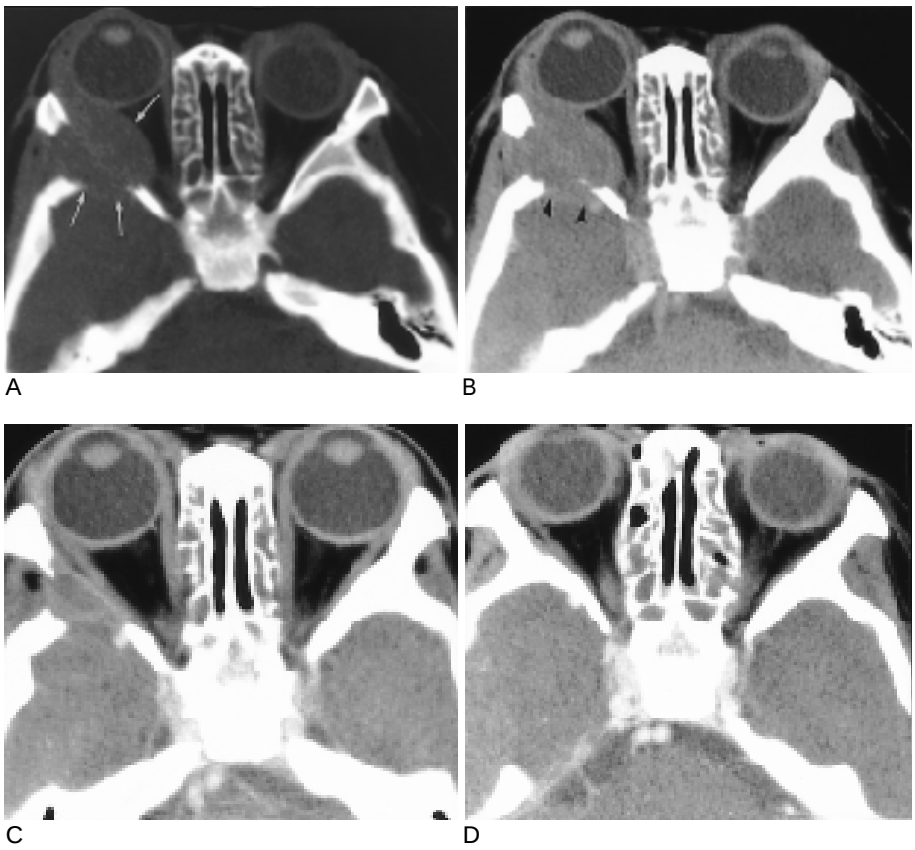

Fig. 1. Case 1. 1.5-year-old girl presented with right exophthalmos. Unenhanced CT (A) and enhanced CT (B) show a large soft tissue mass (arrows) with bone destruction involving lateral wall of right orbit. Minimal, peripheral enhancement is seen in B. There is intracranial extension (arrowheads) without 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-Schuller-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 2. CT Findings in Six Patients with Langerhans Cell Histiocytosis

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

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

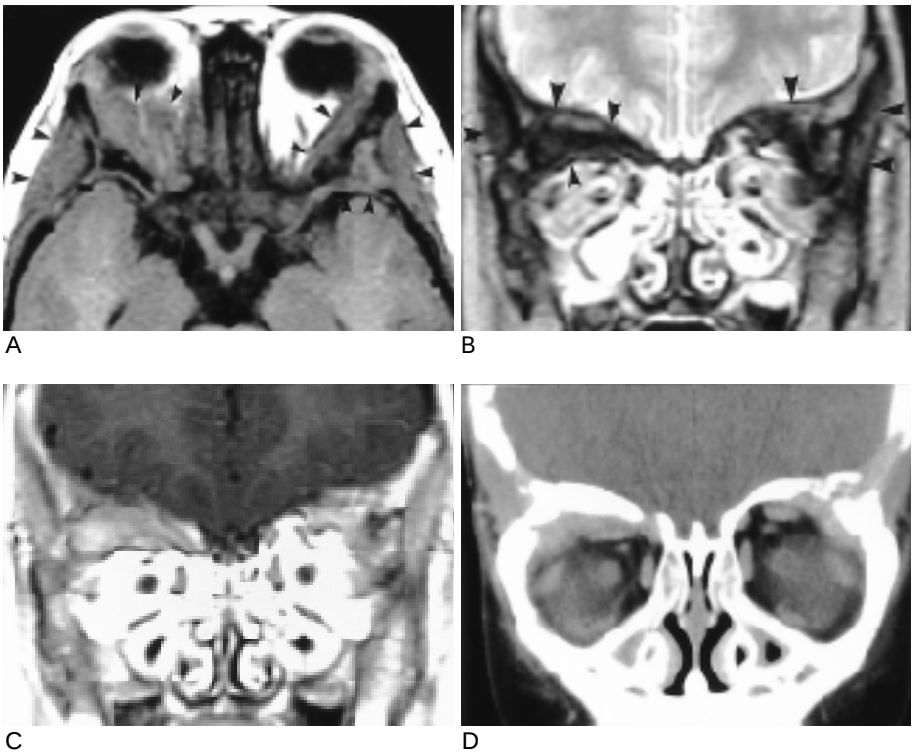

Fig. 2. Case 4. Three-year-old girl presented with poor oral intake. MR T1-weighted image (A) and T2-weighted image (B) show iso-signal intensity masses with bone destruction (arrowheads) involving superolateral wall of both orbits. Postcontrast T1-weighted coronal MR image (C) shows marked enhancement 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 permeative 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.

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안와 랑게르한스세포조직구증의 CT 소견[1]

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이진아 · 윤혜경 · 김보경 · 김경아 · 주인욱

목적: 안와를 침범한 랑게르한스세포조직구증(Langerhans cell histiocytosis)의 CT소견을 알아보고자 하였다.
대상과 방법: 병리조직학적으로 확진된 6명의 안와 랑게르한스세포조직구증 환아를 대상으로 초기 CT 및 추적 관찰한 CT (n = 5) 또는 MR (n = 1)소견을 후향적으로 분석하였다.
결과: 초기CT 에서 다양한 골 파괴와 연부조직종괴를 보였으며 조영증강 전 CT에서 평균 하운스필드 수치는 44 였다. 조영증강 후 연부조직 종괴는 경도에서 중등도의 조영 증강을 보였으며 평균 하운스필드 수치는 74였다. 골파괴는 불규칙하고 경계가 잘 그려지며 골막반응은 보이지 않았다. 종괴내 석회화 소견도 없었다. 2명에서 간 비종대가 동반되었고 한 환아에서 말안장등위(suprasellar) 종괴와 늑골병변이 같이 있었다. 치료 후 추적 관찰한 CT 및 MR에서 연부조직 종괴는 소실되고 파괴된 골 부위는 재골화와 재형성을 보였다.
결론: 불규칙하지만 경계가 명확한 골파괴를 동반한 연부조직 종괴가 안와를 침범한 랑게르한스세포조직구증의 특징적인 소견이라고 생각된다. 치료 후 추적 관찰 시 골 파괴 부위의 재형성과 재골화를 볼 수 있다.

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