Langerhans' cell histiocytosis of the temporal fossa: A case report

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Abstract. Langerhans' cell histiocytosis (LCH) is a rare disease with a wide spectrum of clinical manifestations, varying from an isolated lesion to systemic involvement. The etiology of this disease remains to be elucidated. The present study reports a case of LCH with temporal fossa localization in an 8-year-old male patient, who had exhibited left temporal pain and headache for 1 month. Physical examination revealed slight exophthalmos and conjunctival hemorrhage in the patient's left eye, and non-contrast computed tomography imaging of the head revealed a soft tissue mass with unclear margins located in the left temporal fossa, as well as a wide bony defect. Magnetic resonance imaging revealed a heterogeneously contrast-enhanced mass near the left temporal pole, which eroded into the patient's left orbit and maxillary sinus. The lesion was totally excised and confirmed to be LCH through biopsy.

Introduction

Langerhans' cell histiocytosis (LCH) is a rare disease with a wide spectrum of clinical manifestations (1). The primary pathogenesis of this disease is an abnormal proliferation of Langerhans' cells, which are typically present only in the dermis (1). A total of three different terms, namely eosinophilic granuloma, Hand-Schüller-Christian disease and Letterer-Siwe disease, were used in the past to describe a group of disorders characterized by increased numbers of histiocytes (2). In 1953, Lichtenstein (3) summarized these disorders as histiocytosis X, due to their similar histopathologies, and in 1987, the term LCH was recommended by The Writing Group of the Histiocyte Society as the official definition for this syndrome (4). The clinical appearance of LCH is dependent on the location of the lesion. The abnormal accumulation of histiocytes may occur in almost every organ, including the central nervous system, skin, bone, bone marrow, lung, liver, spleen and lymph nodes, and may cause associated signs and symptoms (4). Therefore, LCH may be classified as monosystemic or plurisystemic, according to the number of organs involved (5). The diagnosis of LCH is based on pathological examination. Typical accumulation of histiocytes, electron microscopic observation of Birbeck granules and the presence of Langerhans' cell-associated markers, including cluster of differentiation (CD)1a and S-100 protein, are crucial for establishing a diagnosis of LCH (6). LCH is typically considered to be an extremely rare disease of childhood, with an incidence rate of ~2-5/1,000,000 children/year (7). There are no specific signs and symptoms associated with LCH involving the skull, and the most common presentation is a painful and immobile scalp mass, which may be palpable in certain cases (5,8). Epistaxis or otorrhagia may be exhibited when the lesion invades the paranasal sinuses or external ear canal (5,8).

The present study reports the case of an 8-year-old male patient with an LCH lesion in his left temporal fossa, and aims to provide clinical experience in the diagnosis and treatment of intracranial LCH.

Case report

An 8-year-old male patient presented at the Department of Neurosurgery of the First Affiliated Hospital of Xi'an Jiaotong University (Xi'an, China) in July 2014 with a 1-month history of left temporal pain and headache. Physical examination revealed slight exophthalmos and conjunctival hemorrhage in the left eye, with no other positive signs. Non-contrast computed tomography (CT; Brilliance 64; Philips Medical Systems, Inc., Bothell, WA, USA) imaging of the head revealed a soft tissue mass with unclear margins located in the left temporal fossa. The mass extended into the patient's left orbit and maxillary sinus. Bone window CT imaging revealed a wide bony defect, including part of the greater wing of the left sphenoid bone, the left lateral orbit and the posterior wall of the left maxillary sinus (Fig. 1). Magnetic resonance imaging (MRI; Signa HDxt 3.0T; GE Healthcare Bio-Sciences, Pittsburgh, PA, USA) revealed a heterogeneously contrast-enhancing mass close to the patient's left temporal pole, eroding into the left orbit and maxillary sinus (Fig. 2). There was a clear plane between the mass and the left temporal pole. The imaging findings suggested a diagnosis of meningioma.

The patient underwent resection of the tumor of the middle cranial fossa. A pterional craniotomy was performed. The bony defect of the sphenoid bone was observed, and the tumor extended outside the temporal fossa through this defect.

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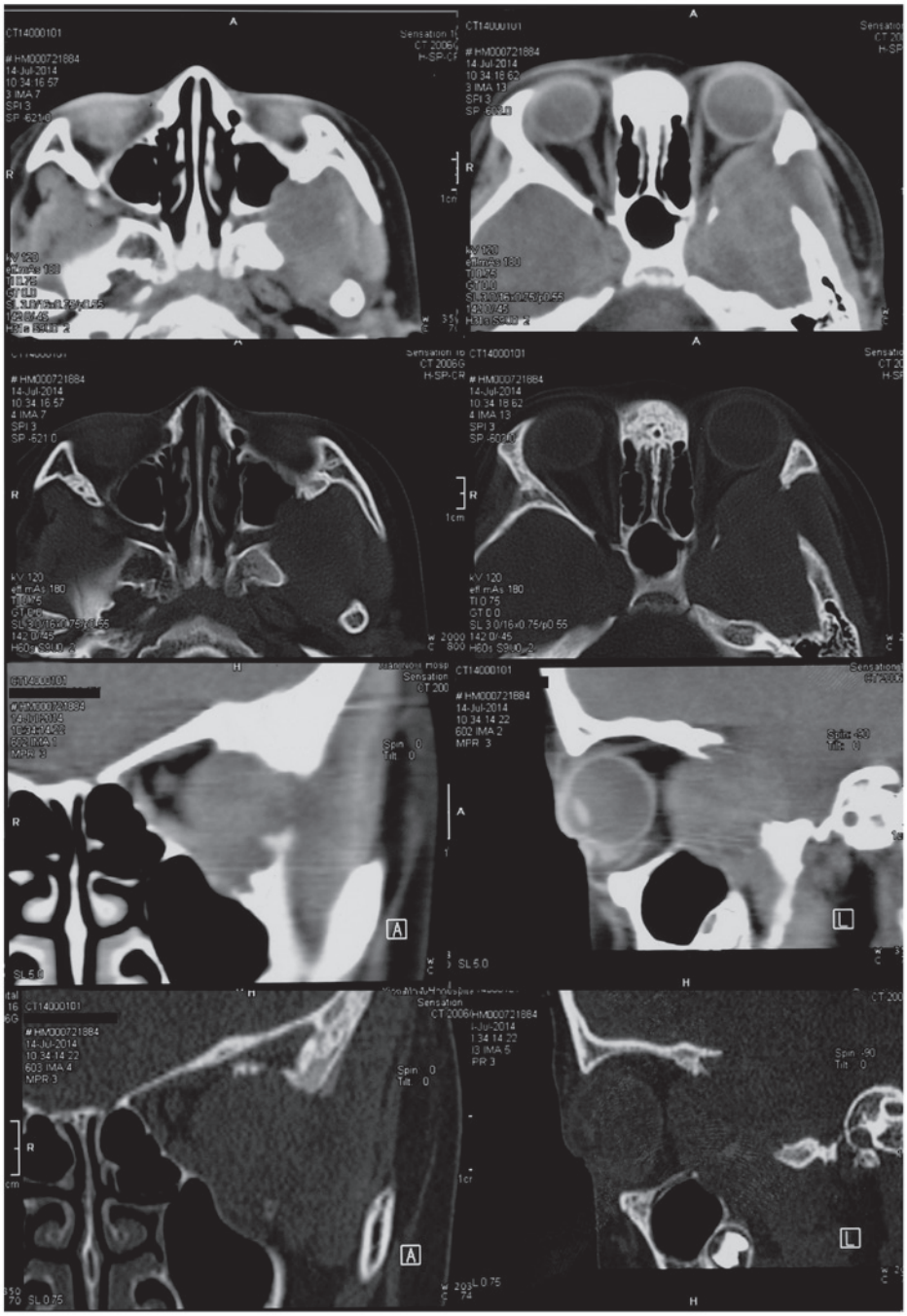

Figure 1. Non-contrast computed tomography image revealing a soft tissue mass with unclear margins located in the left temporal fossa, and a wide bony defect including part of the greater wing of the left sphenoid bone, the left lateral orbit and the posterior wall of left maxillary sinus.

Following removal of the bone flap, the tumor was revealed to be completely extradural, but adhered to the dura. Although the patient's left lateral orbit and the posterior wall of his left maxillary sinus were eroded by the tumor, the orbital fascia and the mucous membrane of the maxillary sinus remained complete, and did not adhere to the tumor. The tumor was removed from around the normal anatomical structures, and was completely resected under a surgical microscope (OPMI Pentero; Zeiss AG, Oberkochen, Germany).

Tissues were fixed with 10% formalin (Xi'an Chemical Reagent Factory, Xi'an, China) for 24 h at room temperature. The fixed tissues were trimmed and placed in embedding cassettes with paraffin (Shanghai Huayong Paraffin Wax Co., Ltd, Shanghai, China). The paraffin-embedded, trimmed blocks were cut into 4-μm sections and adhered to microscope slides. The paraffin sections were deparaffinized by xylene (Xi'an Chemical Reagent Factory) and hydrated routinely, then hematoxylin and eosin staining (ZSGB-BIO, Beijing, China) was performed.

Histological section of the tumor tissue stained with hematoxylin and eosin revealed the presence of histocytes, with moderate amounts possessing eosinophilic cytoplasms and pale staining nuclei (Fig. 3). Immunohistochemistry of the tumor tissue revealed positivity for S-100 (monoclonal mouse anti-human; catalog no., ZM-0224; ZSGB-BIO, Beijing, China), CD1a (monoclonal rabbit anti-human; catalog no., ZA-0544;

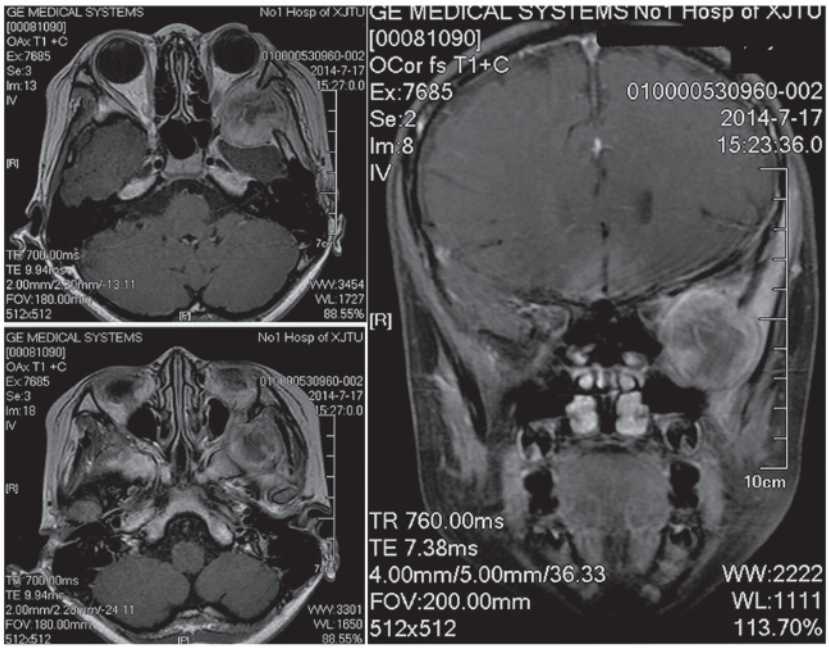

Figure 2. Magnetic resonance imaging revealed a heterogeneously contrast-enhancing mass near the left temporal pole, which eroded into the left orbit and maxillary sinus.

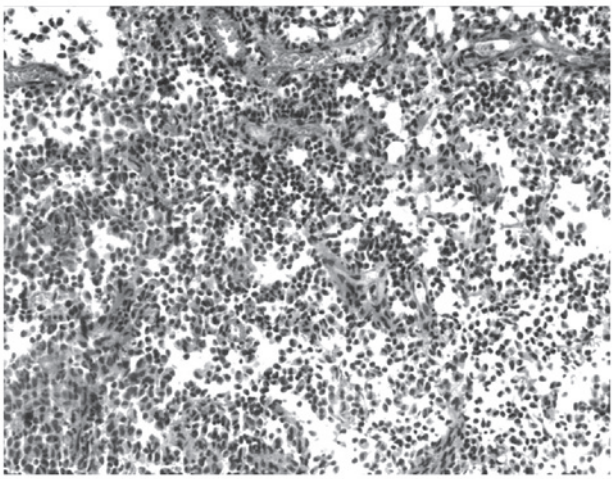

Figure 3. Langerhans-like cells demonstrate diffuse distribution (hematoxylin and eosin staining; magnification, x10).

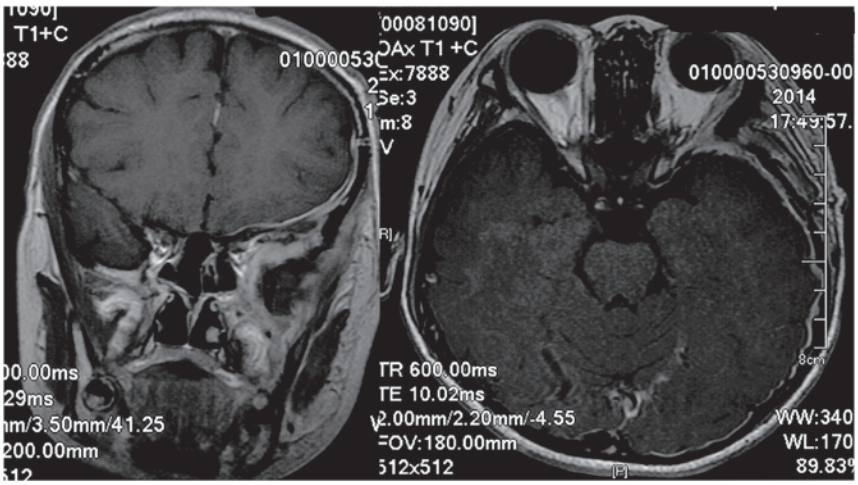

Figure 4. Follow-up magnetic resonance imaging revealed no residual tumor matter or recurrence.

ZSGB-BIO), vimentin (monoclonal rabbit anti-human; catalog no., ZA-0511; ZSGB-BIO) and cytokeratin (monoclonal mouse anti-human; catalog no., ZM-0067; ZSGB-BIO), and negativity for epithelial membrane antigen (monoclonal mouse anti-human; catalog no., ZM-0095; ZSGB-BIO), desmin (monoclonal rabbit anti-human; catalog no., ZA-0610; ZSGB-BIO), CD68 (monoclonal mouse anti-human; catalog no., ZM-0060; ZSGB-BIO), glial fibrillary acidic protein (monoclonal rabbit anti-human; catalog no., ZA-0529; ZSGB-BIO), synaptophysin (monoclonal rabbit anti-human; catalog no., ZA-0236; ZSGB-BIO) and chromogranin A (monoclonal rabbit anti-human; catalog no., ZA-0507; ZSGB-BIO). The final pathological diagnosis of the tumor was LCH.

The patient experienced a good recovery with no neurological deficits following surgery, and was treated with chemotherapy (vinblastine 6 mg/m^2 intravenously administered once a week for 6 weeks following surgery; prednisone 40 mg/m^2 once a day administrated orally for 4 weeks following surgery and then gradually at a reduced dosage over 2 weeks) and radiation treatment (three dimensional-conformal radiotherapy; total dose, 15 Gy in 1.5 Gy increments). Follow-up MRI revealed no residual tumor matter or recurrence (Fig. 4). Written informed consent was obtained from the family of the patient for the publication of the present study.

Discussion

As mentioned previously, there are no specific signs and symptoms for LCH involving the skull, and the most common presentation is a painful and immobile scalp mass, which may be palpable in certain cases (5,8). In the present study, the patient experienced left temporal pain and headache, however, there was no obvious palpable mass. Therefore, imaging examinations of the cranium were essential in order to locate the lesion.

LCH lesions may develop in the diploic space and are round or oval-shaped with well-defined margins, which is described as a 'punched-out' image on an X-ray (6). CT scanning is useful for identifying bony defects that may be eroded by the lesion, while MRI may be used to identify the intracranial lesion (9). Due to the absence of LCH diagnostic criteria for imaging examination, meningioma, rhabdomyosarcoma and Ewing's sarcoma should be considered during differential diagnosis (9). In the present case, the lesion was located in the patient's left temporal fossa, and a wide bony defect was detected. As 60-80% of LCH cases have skull involvement (8), it was appropriate to hypothesize that the lesion originated from the skull. In recent years, positron emission tomography/CT has proven to be the most sensitive test available for the identification of LCH lesions and in the evaluation of patient responses to therapy (10,11).

Once the diagnosis of LCH has been confirmed, it is important to assess the involvement of other organs, as LCH is typically a systemic disease. According to the current guidelines, full blood count, liver function, electrolytes, erythrocyte sedimentation rate, abdominal ultrasound, coagulation studies, chest radiograph and skeletal radiographic survey should been performed during this phase (9). In the present case, all these tests were performed, which confirmed that no other organs were involved (data not shown).

Due to the potential for development of sequelae, according to the current guidelines, systemic therapy is recommended for the treatment of patients with lesions involving the skull base, orbits and temporal bone (9). Therefore, chemotherapy and radiation treatment were performed following surgery in the present case. The standard chemotherapy treatment is based on vinblastine and steroids, and the clinical response should be evaluated following the first 6 weeks of treatment (9).

There remains controversy regarding the use of radiotherapy to treat LCH patients. Certain experts no longer recommend radiotherapy due to the risk of long-term sequelae (12). However, other experts consider radiotherapy to be an effective and safe treatment option for LCH patients (13,14). Thus, the overall safety of radiotherapy to treat LCH patients requires additional investigation in future studies.

In conclusion, the rare case described in the present report highlights the importance of neurosurgeons to be familiar with LCH, as this disease frequently involves head tissues and organs. LCH should be considered during differential diagnosis in children with imaging examination results suggestive of an intracranial lesion associated with a bony defect.

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