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Case Report

Calvarial Langerhans cell histiocytosis in an adult presenting rapid growth

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ABSTRACT

Background: Langerhans cell histiocytosis (LCH) arising in the skull is rare in adulthood.

Case Description: A 58-year-old woman experienced a durable headache. Cerebral magnetic resonance imaging (MRI) at presentation showed no abnormalities; however, MRI and computed tomography (CT) performed 6 weeks later revealed the emergence of a well-demarcated, heterogeneously enhancing calvarial tumor accompanied by irregular-shaped bone erosion. On MRI, the temporalis muscle and subcutaneous tissue adjacent to the tumor were extensively swollen and enhanced. The patient underwent *en bloc* resection. The microscopic appearance of the tumor was consistent with that of LCH. Postoperative systemic 18F-fluorodeoxyglucose positron emission tomography/CT did not detect any abnormal accumulation.

Conclusion: LCH may develop within a short period. It should be considered as a differential diagnosis when a rapidly growing calvarial tumor is encountered, even when the patient is an adult. Prompt histological verification is recommended in such cases.

Keywords: Adult, Calvarium, Langerhans cell histiocytosis, Rapid growth

INTRODUCTION

Langerhans cell histiocytosis (LCH) is thought to be a proliferative disorder of bone marrow-derived immature cells, presenting with diverse clinical manifestations, that commonly affect skeletal bones, skin, lymph nodes, and lungs.[1] Serum soluble interleukin-2 receptor (sIL-2r) level has been proposed as a prognostic factor or indicator of disease activity.[14,15] However, the mechanism underlying LCH development is not well understood. Demir *et al.* documented that low Ki-67 values may correlate with an aggressive clinical course and rapid expansion.[4] In general, patients with isolated LCH lesions are anticipated to have a good prognosis; however, those with multisystem involvement have a progressive disease course despite treatment.[6,16] LCH commonly develops in the pediatric population and is rare in adulthood. In addition, the differences and similarities in the long-term outcomes of pediatric and adult patients are not well known.[2,3,7,8,10] In the adult population, LCH lesions arising in the skull are very rare. Most present as osteolytic lesions or develop in association with the previous surgeries.[5,9,11-13,17]

Here, we present an isolated calvarial LCH in an adult patient that developed within a short period of time.

CASE PRESENTATION

A 58-year-old, previously healthy woman, presented to a local physician with a headache. Her medical history was unremarkable, and she had not experienced a previous craniotomy or head injury. Headache was neither distinctive nor localized to a specific cranial region. Cerebral magnetic resonance imaging (MRI) at the time did not reveal any abnormal findings in the intracranial cavity, skull, or scalp [Figure 1a]. As the headache persisted, the patient underwent a second MRI after 6 weeks, which revealed a mass lesion in the right calvarial convexity [Figure 1b]. The patient was referred to our hospital. At presentation, the patient presented with localized pain in the right parietal region, where peculiar superficial findings were not observed. She was afebrile, and the blood examination revealed normal findings. The serum sIL-2r level was 141 U/ml (normal range: 157–474 U/ml). Cranial computed tomography (CT) showed an isodense mass in the right parietal bone. It was accompanied by irregularly shaped bone erosion, mainly involving the diploe and outer table. The inner table was partially erosive [Figure 2]. On MRI, the tumor presented iso/low intensity on T1- and high intensity on T2-weighted images. It was well demarcated from the surrounding tissue, and heterogeneously enhanced. The temporalis muscle and subcutaneous tissue adjacent to the tumor were extensively swollen and enhanced compared to those on the contralateral side. The inner part of the tumor appeared adjacent to the dura mater [Figure 3]. No abnormal enhancements were observed in the hypothalamic-pituitary axis. The patient underwent a tumor resection. The tumor tissue was whitish, elastic hard in consistency, and mostly well demarcated from the surrounding tissue, while partially adhering to the inner surface of the temporalis muscle through fine defects in

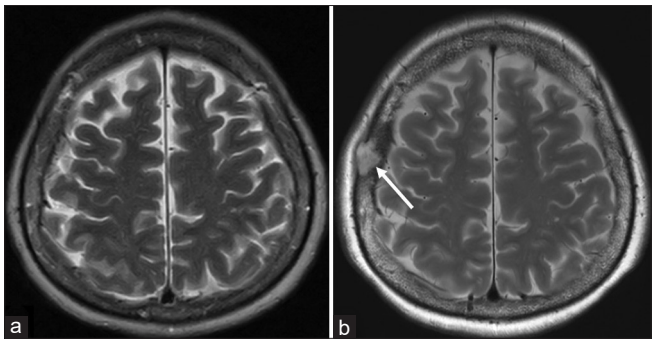

Figure 1: Axial T2-weighted magnetic resonance imaging performed near the same level, at the initial presentation to a local physician (a) and 6 weeks later (b), showing an emergence of a tumor in the right calvarial convexity (b, arrow).

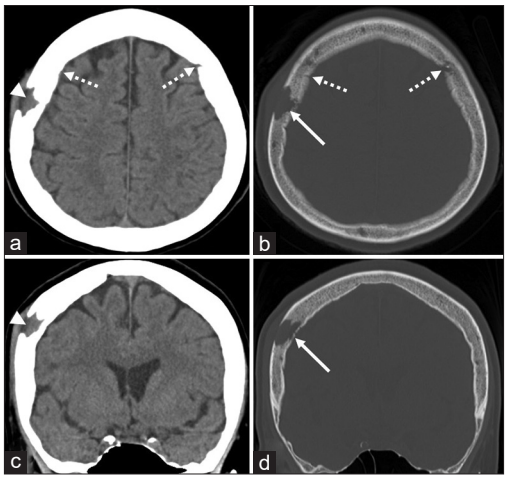

Figure 2: Axial (a) and coronal (c) computed tomography scans and their corresponding bone target images (b and d) showing an isodense mass in the right parietal bone (a and c, arrowhead) accompanied by irregularly shaped bone erosion mainly involving the diploe and outer table (b and d, arrow). The inner table is partially erosive (d, arrow). Dashed arrow: coronal suture.

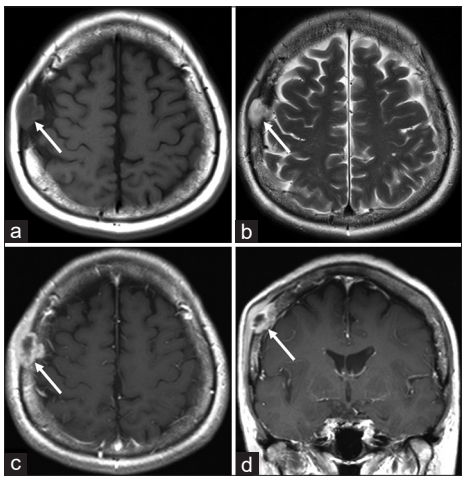

Figure 3: Axial T1- (a) and T2- (b) weighted magnetic resonance imaging showing a calvarial tumor presenting iso/low intensity on T1- and high intensity on T2-weighted images, and well demarcated from the surrounding tissue (arrow). Post contrast axial (c) and coronal (d) T1-weighted magnetic resonance imaging showing heterogeneous enhancement of the tumor with the inner part adjacent to the dura mater (arrow). Note that the temporalis muscle and subcutaneous tissue adjacent to the tumor are extensively swollen and enhanced compared to those on the contralateral side.

the inner table. *En bloc* resection was achieved as a bone flap with adhered temporalis muscle fibers. The size of the bone defect was 25 × 20 mm at the outer surface of the skull [Figure 4]. The dura mater underneath the bone flap appeared to be intact. The resulting calvarial defect was covered with a titanium plate. Microscopically, the tumor comprised proliferating neoplastic cells with cleaved nuclei, accompanied by infiltration of lymphocytes and eosinophils. Immunohistochemical examination showed positive staining for the S100 protein and CD68 [Figure 5]. In addition, subtle invasion of the subcutaneous tissue was observed. These results were consistent with those of LCH. Postoperative systemic 18F-fluorodeoxyglucose positron emission tomography (PET)/CT did not detect any abnormal accumulation. The patient was diagnosed with monostotic LCH. The patient is currently undergoing periodic PET/CT surveillance.

DISCUSSION

In the present case, rapid growth of the calvarial LCH was detected for 6 weeks. During this period, the LCH formed an osteolytic lesion measuring 25 × 20 mm in diameter. Osteolytic calvarial lesions are documented to be an infrequent entity. In a previous study involving 10 children younger than 15 years and 26 adult patients, the most common pathological diagnoses were metastasis and LCH (25.0%), followed by intraosseous hemangioma (13.9%).[5] Furthermore, patients with delayed diagnoses are likely to relapse after successful initial treatment.[16] Therefore, prompt surgical intervention should be performed for histological verification, assuming LCH as a differential diagnosis, in adult patients with osteolytic calvarial lesions.

The rapid growth of the present LCH and its lymphocytic infiltration suggests that the inflammatory reaction might be associated with the local pain. However, the correlation between the patient's headache that had persisted before radiological emergence of the LCH and its pathology is unknown.

In this case, the temporalis muscle and subcutaneous tissue adjacent to the tumor appeared thick and were extensively enhanced on the presurgical MRI. In contrast, intraoperative findings showed that the outer surface of the tumor was mostly well demarcated from the surrounding tissue. Microscopically, only a subtle tumor invasion was observed in the adjacent subcutaneous tissue. Therefore, on MRI, inflammatory mechanisms were thought to be predominantly associated with the appearance of the temporalis muscle.

The patient was diagnosed with monostotic LCH. In general, such LCH is anticipated to have a good prognosis

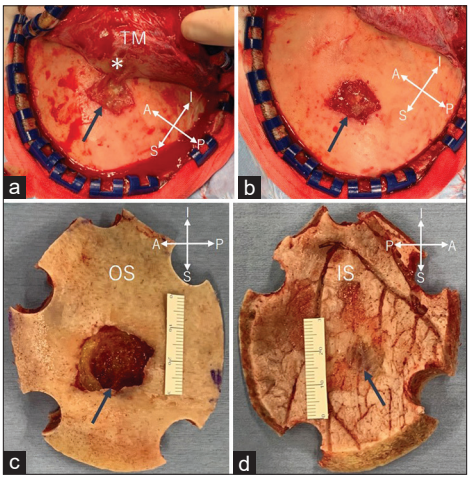

Figure 4: (a-d) Intraoperative photos. (a and b) Reflection of the scalp flap showing a whitish tumor protruding through the completely eroded outer table (arrow). The tumor is partially adhered to the inner surface of the temporalis muscle (a, asterisk). (c) *En bloc* tumor resection has been achieved with surrounding bone. (d) Fine defects are found in the inner table adjacent to the tumor (arrow). *A:* Anterior, *I:* inferior, *IS:* inner surface of the skull, *OS:* outer surface of the skull, *P:* posterior, *S:* superior, *TM:* temporalis muscle, *Arrow* in (c): bone defect, 25 × 20 mm in diameter at the outer surface of skull; *Asterisk:* tumor tissue adhered to the temporalis muscle fibers.

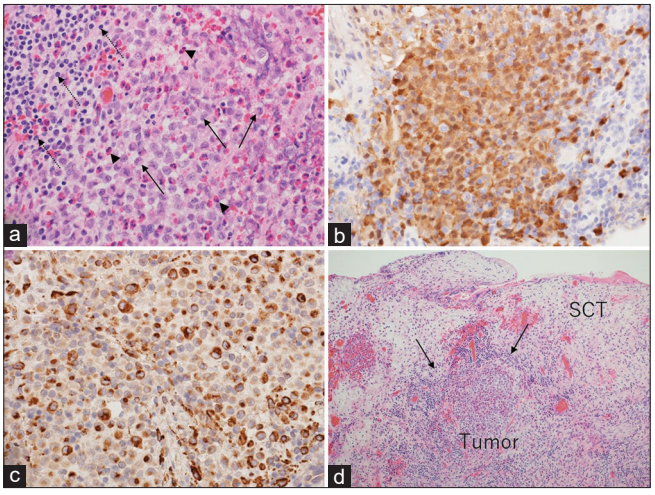

Figure 5: (a) Photomicrograph of the resected specimen showing proliferation of neoplastic cells with cleaved nuclei (arrows) accompanied by infiltration of the lymphocytes (dashed arrows) and eosinophils (arrowheads). Immunohistochemical examination showing positive staining for the S100 protein (b) and CD68 (c). Tumor invasion of the subcutaneous tissue is observed (d, arrows). SCT: Subcutaneous tissue. (a and d): hematoxylin and eosin stain. (a): × 200, (d): × 40, and (b and c): × 200.

without the need for systemic therapy after successful initial treatment.[2,6,16,17] However, LCH in adults is rare and not well understood. Furthermore, most of our knowledge about its diagnosis and treatment comes from pediatric studies.[2,10,11] Given that LCH invasion was found in the subcutaneous tissue, careful long-term observation is necessary for the patient.

CONCLUSION

LCH may develop within a short period. It should be considered as a differential diagnosis when a rapidly growing calvarial tumor is encountered, even when the patient is an adult. Prompt histological verification is recommended in such cases.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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