Langerhans Cell Histiocytosis Mimicking a Meningeal Lesion with Temporal Bone and Muscle Compromise in an Adult Patient: A Case Report

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Abstract

Introduction Langerhans cell histiocytosis (LCH) is a rare proliferative systemic disease characterized by the growth of abnormal dendritic cells and wide-ranging organ involvement. This condition can affect individuals of all ages, but most commonly children, with a peak incidence in toddlers. Symptoms may vary depending on the affected organ or system.

Case Report A 43-year-old man presented with a left temporal stabbing headache unresponsive to management with therapy and nonsteroidal anti-inflammatory drugs. Initial evaluation revealed a contrast-enhanced left temporal extra-axial lesion with bone and muscle compromise. Differential diagnoses, including multiple myeloma, were explored. Initial laboratory tests and imaging studies showed no other abnormalities, except for splenomegaly and a residual granuloma in the left lung. En bloc resection of the lesion was recommended. The patient underwent surgical intervention, which included resection of the dural lesion and all borders of an infiltrating tumor within the temporalis muscle and the affected portion of the left temporal bone. Posterior pathological examination revealed LCH. Postoperative course was uneventful. Follow-up appointments were scheduled after pathology results confirmed the diagnosis. Patient has continued follow-up for the following 3 months after the surgical procedure. Further evaluations are pending.

Discussion This case report corresponds to a patient with LCH. These patients are individualized and stratified based on local or systemic involvement to determine the most appropriate type of management. This is a rare case as LCH is rare in older patients and the initial presented lesion initially mimicked a meningioma; however, its atypical behavior and associated lytic compromise led to consideration of possible differential diagnoses.

Keywords
- case report
- Langerhans cell histiocytosis
- adult
- bone lesions

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Conclusion LCH can present with lytic bone lesions, mimicking other conditions, including infiltrative neoplastic lesions. Early diagnosis and appropriate surgical management are essential for optimal patient outcomes. Long-term follow-up is crucial to monitor disease progression and response to treatment.

Introduction

Langerhans cell histiocytosis (LCH), previously referred to as histiocytosis X, is a rare clonal disorder characterized by the proliferation of clonal CD1a+/CD207+ (or Langerin+) immature cells derived from dendritic cells within the mononuclear phagocytic system.[1] While it was previously considered a reactive clonal proliferation of Langerhans cells, recent studies have shown that a significant number of cases of LCH exhibit extracellular signal-regulated kinase (ERK) phosphorylation and the BRAF V600E mutation in approximately 50% of cases, suggesting a shared mutagenic pathway with an inflammatory myeloid neoplasm.[1,2]

The discovery of the oncogenic BRAF V600E mutation or MAPK–ERK pathway activation in over 50% of LCH cases marked a significant advancement in our understanding of the disease's underlying mechanisms.[3,4] LCH was initially categorized into three distinct entities: (1) eosinophilic granuloma; (2) Hand–Schüller–Christian disease (characterized by the triad of diabetes insipidus, exophthalmos, and lytic bone lesions); and (3) Letterer–Siwe disease (characterized by generalized hyperplasia of nonlipid-storing macrophages in the liver, spleen, lymph nodes, skin, and bone marrow).[5]

Currently, LCH is classified as either single-system (SS) or multisystem (MS) and further categorized as unifocal or multifocal disease. The classification of MS disease depends on the involvement of risk organs (RO), which include the liver, lungs, spleen, and bone marrow. SS disease, RO-negative MS disease, and RO-positive MS disease correspond closely to eosinophilic granuloma, Hand–Schüller–Christian disease, and Letterer–Siwe disease, respectively.[6]

Clinical signs and symptoms vary depending on the affected organs. Bony lesions are the most common, occurring in approximately 80% of patients with LCH. In adults, the most commonly affected bone is the jaw, often causing pain and tooth loss. Temporal bone involvement may be unilateral or bilateral, with potential restriction to the petrous bone.[7] In children, the skull is the most commonly affected site, followed by the spine, extremities, pelvic bone, and ribs.[6–8] The overall 5-year survival probability postdiagnosis in adults was 92.3%.[9] It is highly unusual for adults to present initially with skull involvement and even more so with an associated meningeal component. During the literature review performed for the present manuscript, we only found one case report of meningeal involvement in an adult, specifically a 21-year-old man.[10] Dural involvement is still rare in children, but it appears to be more frequent than in adults.[11]

The following article presents a case report of a patient diagnosed with LCH, atypical in its presentation as it mimicked a meningeal lesion along with bone and muscle compromise in an adult patient.

Case Summary

Initial Assessment

We present a case of a 43-year-old man patient who consulted our outpatient clinic due to complaints of a persistent left frontal stabbing headache, which had led to multiple visits to the emergency room and the repeated use of nonsteroidal anti-inflammatory drugs for pain management. The patient had no significant prior medical history except for a surgical excision of a pterygium in his left eye. Initial physical examination was unremarkable.

The patient provided a gadolinium-enhanced brain magnetic resonance imaging (MRI), revealing a contrast-enhanced lesion with thickening of the dura in the left temporal convexity that invaded the squamous portion of the left temporal bone, as well as the temporalis muscle. Because of the location of the lesion and its radiological appearance, it was initially classified as a meningioma (►**Fig. 1**). The patient was then referred for inpatient hospitalization for a more detailed evaluation. Computed tomography (CT) scan confirmed a lytic bone lesion located in the left temporal bone (►**Fig. 2**). This lesion was narrowly margined, well-defined, involving the internal diploë, and associated with a solid component.

The patient also underwent an evaluation by the internal medicine department due to the inclusion of multiple myeloma among the differential diagnoses that could explain the lesion. Various studies were performed, with unremarkable results. Additional CT scans of the neck, thorax, and abdomen were performed to investigate the possibility of a primary malignancy, as it was deemed unlikely that the initial cranial lesion was an isolated primary lesion. Neck CT showed no abnormalities, whereas the thoracic CT revealed a residual granuloma in the lower left lung lobe. Abdominal CT reported splenomegaly without any other associated focal lesions. After ruling out a primary lesion, a decision was made to proceed with surgical management. The patient underwent a thorough assessment by the anesthesiology team, and surgery was scheduled.

Surgery and Perioperative Process

The patient was placed under general anesthesia. A frontotemporal incision was made, followed by posterior dissection of the temporalis muscle, revealing an infiltrative lesion and a lytic bone lesion in the left temporal bone with an adjacent friable solid tumor.

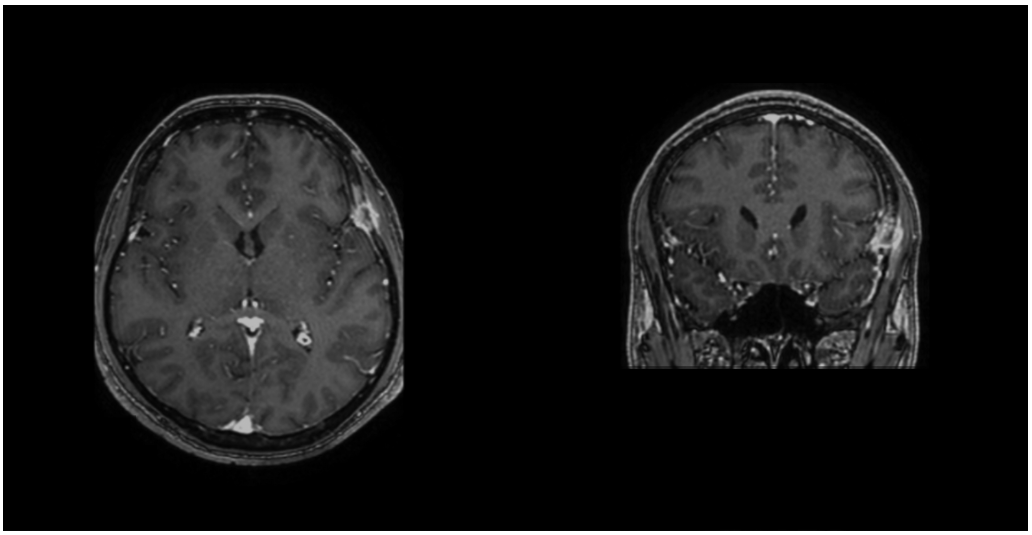

Fig. 1 Contrast-enhanced brain magnetic resonance imaging. Axial and coronal views are shown, in which a lesion with peripheral enhancement is seen, compromising the dura of the left temporal convexity as well as the temporalis muscle and the squamous portion of the temporal bone.

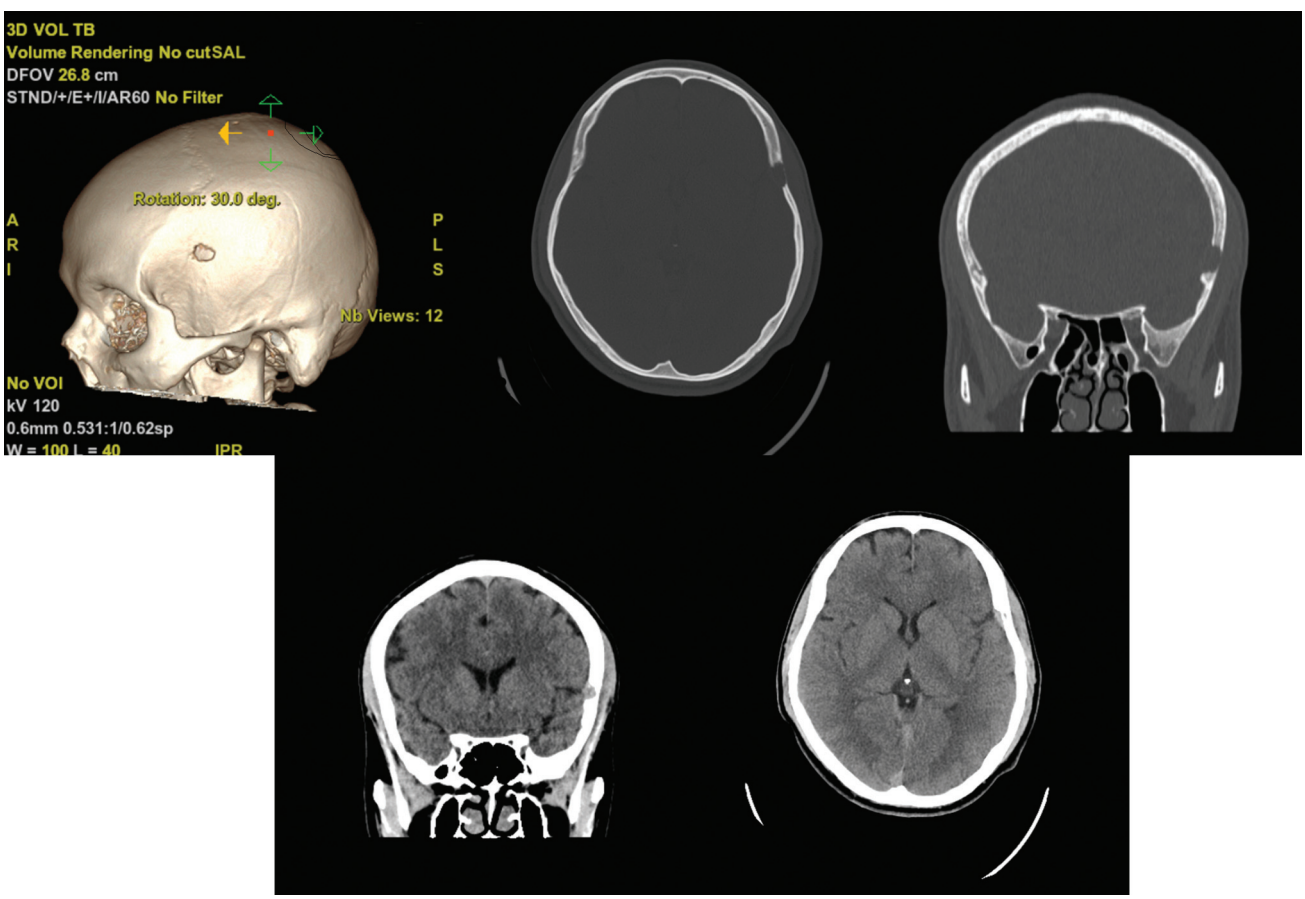

Fig. 2 Head computed tomography scan with 3D reconstruction. Axial and coronal views of the bone and brain windows are shown, in which a lytic lesion measuring approximately 17 mm in its anteroposterior diameter that affects the left temporal bone is visualized. A solid component of the lesion is also observed, which initially seems to arise from the adjacent meninges.

Pathological tissue was resected, including samples of the epidural lesion and the affected portion of the left temporal bone, which were sent for further histological examination. Dural infiltration was evident, necessitating a durotomy. The resulting intracranial defect was covered with absorbable hemostatic gelatin sponge. Duraplasty was performed using an autologous pericranial patch secured with 4–0 polyglactin sutures.

The osseous defect was covered with a set of plates and miniplates with a mesh and four self-drilling screws. The

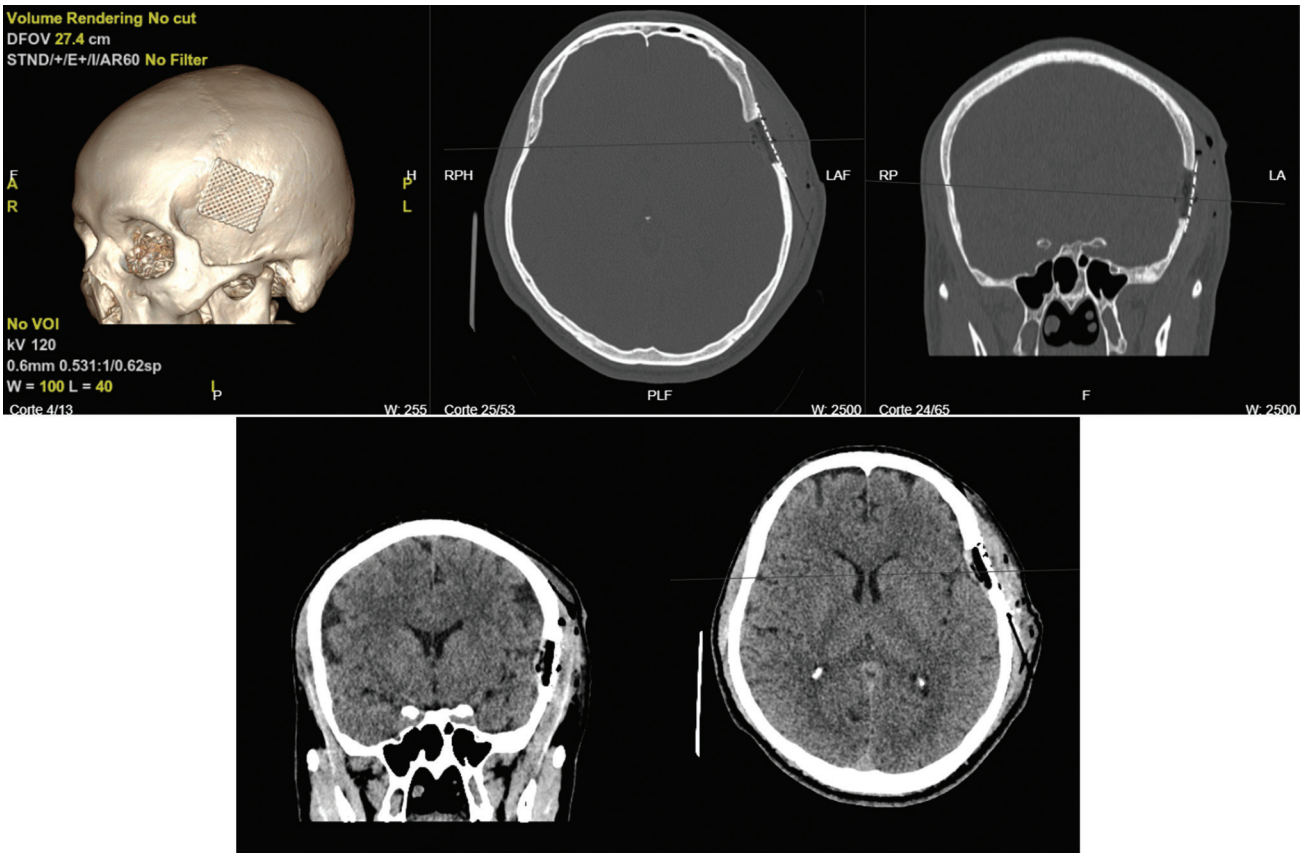

Fig. 3 Postoperative head computed tomography scan with 3D reconstruction. Axial and coronal views of the brain and bone windows are shown. Complete resection of the solid and lytic component of the lesion was achieved, with coverage of the resulting osseous defect.

surgical cavity was irrigated with vancomycin powder. Closure was performed in layers.

The immediate postoperative period transpired uneventfully. Control head CT showed expected postsurgical changes, with complete surgical resection of the lesion and with proper positioning of the surgical material in the left parietal bone (►**Fig. 3**). The patient did not require additional monitoring in the intensive care unit and was transferred to a regular hospitalization room, where he remained under observation for the following 24 hours. He was discharged with analgesics and instructions for an outpatient consultation in the following 2 weeks.

Follow-up

The patient attended two follow-up appointments: 2 and 7 weeks after the procedure. During both consults he reported no neurological symptoms. Surgical wound was dry, with no evidence of cerebrospinal fluid leakage, bleeding, or signs of infection. Pathology results were obtained. The microscopic description reported cortical bone with foci of histiocyte aggregates accompanied by frequent eosinophils and a sparse monomorphic lymphoid population. Immunohistochemical studies were performed, with markers CD1A and S100 reported as positive.

The pathological diagnosis was LCH. Based on these pathology findings, the patient was scheduled for an outpatient internal medicine evaluation and a subsequent follow-up appointment with neurosurgery, which included a gadolinium-enhanced brain MRI and a head CT with three-dimensional (3D) reconstruction. As of time of publishing, the patient had yet to return for further consultation.

Discussion

LCH is an orphan disease, defined as a clonal proliferation of CD1a+/CD207+ abnormal myeloid precursor cells, derived from dendritic cells that exhibit the same antigens (Langerin, CD1a, S100 protein), and same intracytoplasmic organelles (Birbeck granules, unique in this cells) as the normal Langerhans cells found in mucosa and skin.[12,13] Incidence in adults has been estimated around 1 to 2 cases per million, likely indicating underdiagnosis.[14]

Its classification as a neoplastic or reactive disorder has been debated. However, recent discoveries of mutations in somatic components of the MAP/ERK pathway, particularly the BRAF V600E mutation, suggest a neoplastic origin and are associated with disease progression. Recent classifications have grouped histiocytoses into five categories: (1) Langerhans-related; (2) cutaneous and mucocutaneous; (3) malignant histiocytosis; (4) Rosai–Dorfman disease; and (5) hemophagocytic lymphohistiocytosis and macrophage activation syndrome,[15] with LCH falling under the Langerhans-related subgroup.

While most common in childhood, LCH can manifest at any age, with varying degrees of systemic involvement. It can be classified based on the number and type of affected

organs, with SS HCL involving only one organ/system and MS disease involving two or more organs/systems. Bone involvement is frequent. Although rare, central nervous system, bone marrow, liver, and vascular involvement are associated with worse prognosis.

Evaluating location and assessing for SS or MS disease is paramount in establishing treatment.[16,17] The petrous portion of the temporal bone has been described as a "current" location of LCH-associated lesions.[7] The patient in this case had SS disease, with an affection of the squamous portion of the temporal bone associated with a soft tissue mass infiltrating the temporalis muscle. Because of the characteristics associated with unifocal lesions, surgical resection is often preferred.[16] Strict postsurgical follow-up is essential to detect new lesions or recurrences.

Conclusion

LCH can present with lytic bone lesions, mimicking other conditions, including infiltrative lesions of the central nervous system such as meningiomas. Early diagnosis and appropriate surgical management are essential for optimal patient outcomes. Long-term follow-up is crucial to monitor disease progression and response to treatment.

Ethical Considerations

We addressed ethical considerations in the publication of this case report in adherence to the Declaration of Helsinki. Patient confidentiality is preserved, and the patient fully understands the nature, benefits, and risks of the research, as well as the academic interest in publishing his medical history. Any potentially identifying information has been omitted or altered. Informed consent was obtained from the patient. No additional institutional review board approvals were required.

Conflict of Interest

None declared.

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