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Langerhans Cell Histiocytosis of the Skull Complicated with an Epidural Hematoma

Hsin-Chi Chen, Wu-Chung Shen, Der-Yang Chou, and I-Ping Chiang

Summary: Langerhans cell histiocytosis (LCH) is a rare disorder that affects the pediatric population. LCH complicated with a neurologic deficit due to the presence of epidural involvement is a rare condition. We describe the CT imaging features in a 2-year-old boy who presented with drowsy consciousness resulting from an epidural hematoma caused by spontaneous bleeding in an LCH of the skull. CT is an excellent means of depicting the full extent of bony destruction and the nature of the process.

Langerhans cell histiocytosis (LCH) is a rare disorder that affects the pediatric population. The most common site of involvement is the skull; LCH is especially prevalent in the frontal and parietal bones (1). LCH of the skull associated with an intracranial epidural hematoma is a rare condition. After a literature review, we found that only Lee et al (2) reported a case involving a patient with epidural hematoma as a major complication of LCH at the occipital bone after minor head trauma. Herein, we report findings in a 2-year-old boy with left occipital bone LCH complicated with spontaneous tumor bleeding and no history of head injury. In the present case, tumor bleeding resulted in rapid expansion of the epidural hematoma that caused a change in consciousness. CT imaging features of this rare manifestation of skull LCH is provided.

Case Report

A 2-year-old boy was admitted to our hospital because of a sudden onset of drowsy consciousness for 1 day. No prior head injury insult was present. His history revealed that a painless, tender, elastic lump had been present in the left occipital region for 3 months, but no treatment was given. Neurologic examination revealed that the Glasgow coma scale score was 11 (eye opening, 3; motor responsiveness, 5; verbal performance 3). On physical examination, a reddish elastic mass was seen beneath the left occipital scalp. No fever or neck stiffness was present. Laboratory evaluation demonstrated normal red blood cell, white blood cell, and platelet counts. The results of coagulation function tests, urinalysis, and biochemistry profile analysis were normal.

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From the Department of Radiology (H.-C.C., W.-C.S.), Department of Neurosurgery (D.-Y.C.), Department of Pathology (I.-P.C.), China Medical College Hospital, Taichung, Taiwan.

Address reprint requests to Hsin-Chi Chen, MD, Department of Radiology, China Medical College Hospital, No. 2, Yuh-Der Road, Taichung, Taiwan 407, Republic of China.

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Emergent nonenhanced and contrast material-enhanced CT scans demonstrated a well-defined osteolytic lesion in the left occipital bone, with an associated underlying large epidural mass ($2.2 \times 4.8 \times 4.8$ cm) and an adjacent subgaleal mass (Fig 1A and B). A small calcification near the bony defect was also depicted, and this was regarded as a residual bony fragment. A high-attenuating hemorrhage filled the space of the bony defect, which showed unequal bone destruction at its inner and outer tables. Both the epidural and subgaleal masses contained high-attenuating hemorrhage mixed with a component that had the same attenuation as that of gray matter. However, no abnormal enhancement was demonstrated. The isoattenuating component was presumed to indicate the presence of tumor tissue or unretracted liquid blood. The large epidural mass extended downward to the left posterior fossa and had compressed the fourth ventricle, resulting in mild hydrocephalus (Fig 1C). On the basis of the imaging findings, an osteolytic lesion with tumor bleeding complicated with epidural hematoma and subgaleal hematoma was considered.

Emergent craniotomy was performed after the CT study. At surgery, the osteolytic lesion and adjacent epidural and subgaleal masses were entirely filled with blood clot and some unretracted liquid blood (about 30 mL). No tumor tissues were visible. The underlying dura mater was intact. The left transverse sinus was not visible during the surgical approach.

Pathologic examination of the blood materials obtained from the osteolytic lesion and adjacent epidural and subgaleal masses revealed the proliferation of mononuclear cells embedded in the blood pool (Fig 2). These cells had abundant acidophilic cytoplasm and oval indented nuclei. These mononuclear cells were confirmed to be Langerhans cells by the positive immunohistochemical staining results with S-100 protein. LCH with tumor bleeding was diagnosed. A subsequent bone survey and bone scanning revealed that this was the only site with LCH involvement.

Discussion

LCH is characterized by localized proliferation of Langerhans cells in bone and/or soft tissue. The precise cause and pathogenesis of LCH remain unclear. More recently, clonal characteristics have been demonstrated for the Langerhans cells in LCH. Therefore, LCH is thought to be a clonal nonneoplastic disorder (3). Bone lesions are the most common manifestations of LCH; they occur in 80%–95% of children with LCH. A predilection for hematopoietically active medullary sites exists; the skull is most frequently involved (4). Most children with bony involvement present with symptoms related to bone pain or a localized mass (5). The treatment of LCH depends on the extent of the disease. Single bone lesions are best treated with surgical curettage (6). In general, the prognosis is excellent when the disease is limited.

In the case presented herein, a diagnosis of cranial tumor rupture with bleeding into the epidural space

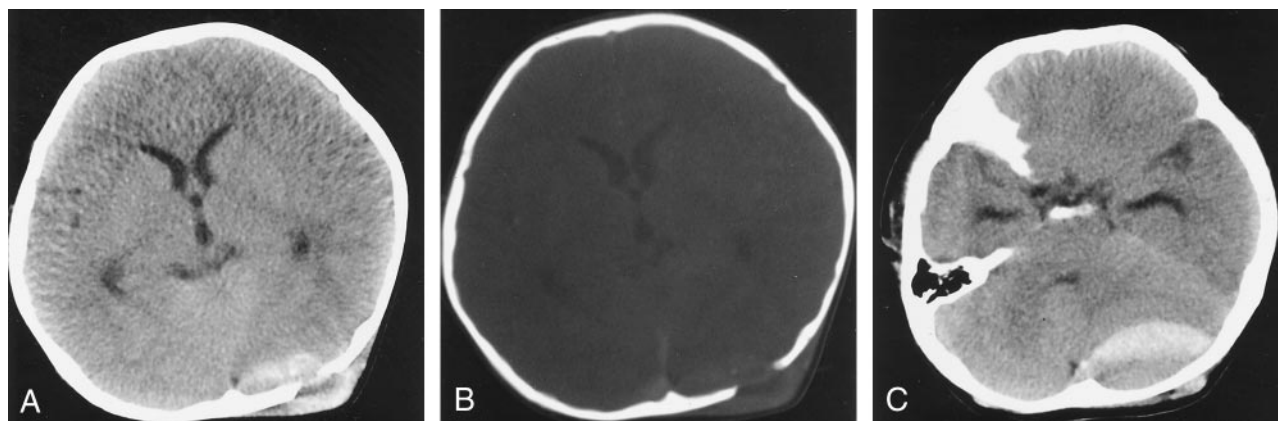


Fig 1. CT images obtained in a 2-year-old boy with drowsy consciousness resulting from an epidural hematoma caused by spontaneous bleeding in an LCH of the skull.

A, Nonenhanced CT scan demonstrates an osteolytic lesion in the left occipital bone, which is associated with an underlying epidural hematoma and an overlying subgaleal hematoma. Hemorrhage fills the space in the skull bone defect. A small residual bony fragment near the bone defect is also depicted. Both the epidural and subgaleal hematomas contain high-attenuating hemorrhage mixed with isoattenuating unretracted liquid blood. No abnormal enhanced component was present within the osteolytic lesion or adjacent epidural and subgaleal hematomas (not shown).

B, CT scan obtained with a bone window setting shows a well-defined osteolytic lesion in the left occipital bone, with unequal bone destruction at its inner and outer table sides that causes the so-called beveled-border appearance.

C, Nonenhanced CT scan shows the downward extension of the large epidural hematoma to the left posterior fossa and compression of the fourth ventricle, with resultant mild hydrocephalus. The epidural hematoma contains a high-attenuating blood clot and isoattenuating unretracted liquid blood.

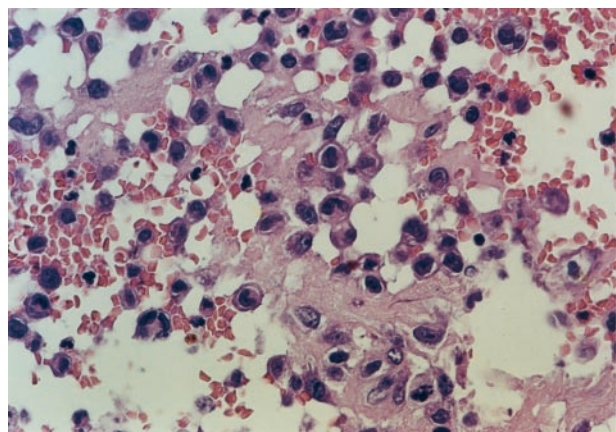


Fig 2. Photomicrograph obtained at pathologic examination reveals proliferation of Langerhans cells with twisted and indented nuclei and conspicuous acidophilic cytoplasm. Scattered eosinophils are also depicted (hematoxylin-eosin, original magnification $\times 200$). The Langerhans cells had immunohistochemically positive results with S-100 protein staining (not shown).

was made according to the clinical setting involving no trauma history and CT scans that demonstrated an epidural hematoma that was contiguous to a skull defect. In this case, CT revealed that the skull lesion had an unequal destruction in the outer and inner tables of the vault; this finding indicated the presence of a beveled border, which is a typical feature in the diagnosis of LCH. In addition, a small bony fragment near the skull defect was seen; this suggested the presence of a button sequestrum, which also provided clues for the diagnosis of LCH. The authors speculate that the bony fragment was situated in the dipole space of the osteolytic lesion before tumor bleeding occurred. It was then pushed into the underlying

epidural space and embedded in the epidural hematoma during the episode of tumor bleeding.

Possible complications of LCH depend on the local situation and the extent of the lesion. Neurologic dysfunction may occur in patients with LCH that involves the central nervous system and spinal cord. Nonetheless, LCH complicated with neurologic deficit because of the presence of epidural involvement is a very rare condition. A literature review revealed that only two cases (2, 7) involved this unusual manifestation. One case occurred in a 4-year-old boy with LCH that involved the body of C6 and the pedicle of T1; this was associated with tumor infiltration in the adjacent anterior and posterior epidural spaces, as detected at CT and MR imaging (7). No hemorrhage was present in these epidural masses. The other case involved an 8-year-old boy with LCH in the right occipital bone that was complicated with tumor hemorrhage leading to epidural hematoma formation, which occurred after two episodes of minor head trauma 4 weeks and 1 day before the CT study (2). In the 8-year-old boy, CT scans showed a supratentorial epidural hematoma consisting of acute and chronic hemorrhage contents.

In contrast, in our case, spontaneous tumor hemorrhage in LCH at the occipital bone occurred in a patient without a history of head trauma. LCH with spontaneous hemorrhage is rare; to our knowledge, it was previously reported in only a single article by Hindman et al (7). They analyzed CT and MR imaging features in 34 children with 262 LCH skeletal lesions; only one 8-month-old boy with multiple frontal and parietal bone lesions had intralesional fluid-fluid levels, which were due to spontaneous tumor hemorrhage. In this 8-month-old boy, the innermost inner table of the frontal and parietal bones was still

preserved. Thus, no intracranial soft-tissue component was contiguous to these skull lesions. However, in our case, the entire inner and outer tables of the occipital bone were destroyed, and intracranial and extracranial components accompanied the skull lesion. Furthermore, LCH with tumor bleeding resulted in rapid expansion of the epidural hematoma, which not only existed in the supratentorial location but also extended to the infratentorial location. The large infratentorial hematoma even compressed the fourth ventricle, with resultant neurologic deterioration.

Conclusion

Cranium LCH with spontaneous hemorrhage may accompany a large epidural hematoma. CT scans can better define a calvarial osteolytic lesion and more

precisely delineate the extra- and intracranial soft-tissue components of the calvarial lesions.

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