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Case Report

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J Korean Neurosurg Soc 43 : 304-306, 2008

10.3340/jkns.2008.43.6.304

Eosinophilic Granuloma Presenting as an Epidural Hematoma and Cyst

Langerhans' cell histiocytosis (LCH) is a rare immunologic disorder characterized by histiocyte proliferation in multiple organ systems. Eosinophilic granuloma, a benign bone lesion, represents a focal form of LCH. We experienced a case of Langerhans' cell histiocytosis in a patient who presented with intracranial epidural hematoma and cyst on the midline of the frontal skull. A 10-year-old boy presented with a rapidly growing large scalp mass on the midline frontal area after mild head trauma. The scalp mass was painless and immobile. Plain skull x-ray showed a punched-out bone lesion. Computed tomography and magnetic resonance imaging showed a non-enhancing osteolytic lesion presenting with an epidural hematoma and cyst on the midline of the frontal skull. The lesion of the skull was completely resected and the patient's recovery was uneventful. The acute presentation of a solitary eosinophilic granuloma of skull with an epidural hematoma has been described in only five cases in the literature and we report the first case of LCH presenting as an intracranial epidural hematoma on frontal area.

KEY WORDS : Langerhans' cell histiocytosis · Epidural hematoma · Eosinophilic granuloma · Head trauma.

INTRODUCTION

Langerhans' cell histiocytosis (LCH) is a rare immunologic disorder characterized by histiocyte proliferation in multiple organ systems[2]. The LCH is used to describe a related group of three disorders of unknown cause ranging from solitary eosinophilic granuloma (EG) of the bone to more severe multifocal or disseminated forms, known as Hand-Schüller-Christian disease or Letterer-Siwe syndrome, respectively[4]. LCH usually manifests before puberty, and the skull is the most frequent site of involvement. The diagnosis of LCH can be confirmed if stains for CD68 antigen and protein S-100 are positive or when intracytoplasmic organelles (Birbeck granules) are seen with electron microscopy.

EG occurs predominantly in children and adolescents. It is a painless, destructive bone lesion that frequently involves the skull (in 43-80% of cases), followed by the femur, mandible, and ribs[7]. Patients typically present with a tender enlarging skull mass with an area of bone destruction underneath, which is surrounded by poorly circumscribed, but sharply defined margins on skull x-ray. The course is benign with a tendency towards spontaneous regression, but recurrences may also occur[7]. The acute presentation of a

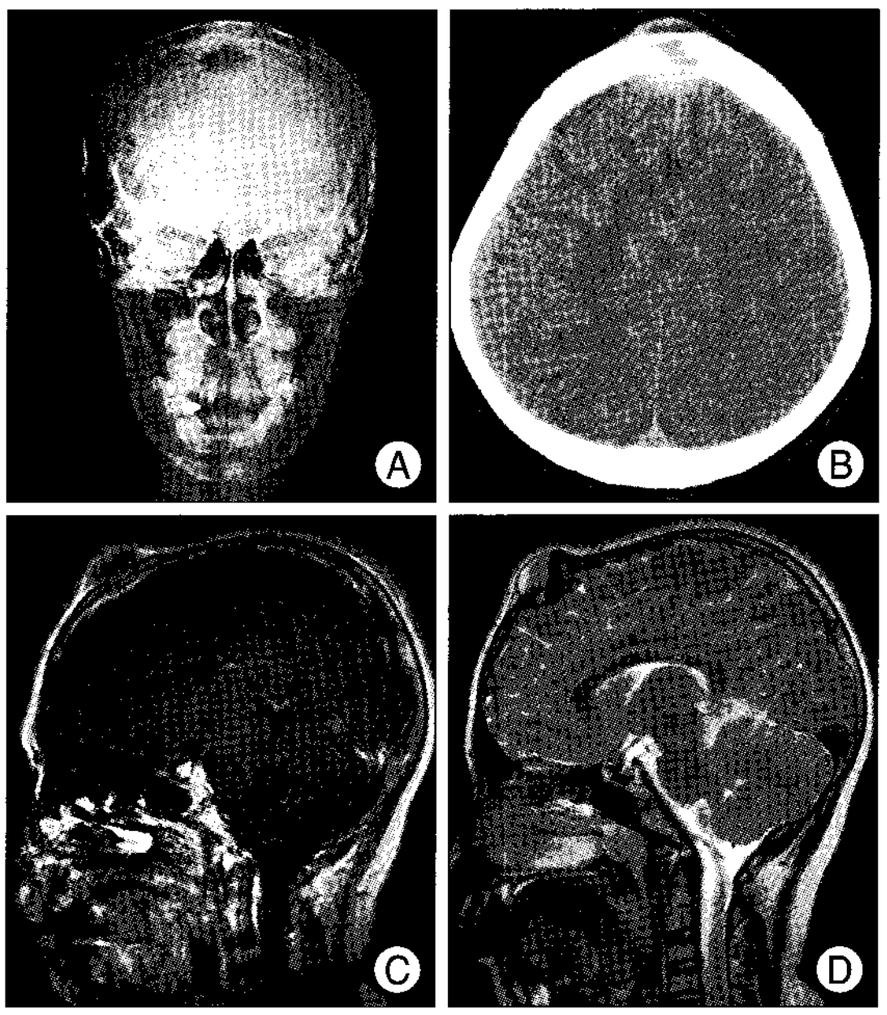

Fig. 1. Skull x-ray (A) showing a 2.5 x 2.0 cm-sized lytic bony defect in the mid-frontal region. Computed tomography (B) showing an epidural hematoma with acute and subacute components. Sagittal enhanced magnetic resonance imaging (MRI) (C) showing an epidural hematoma. Sagittal T2-weighted MRI (D) showing mixed cystic fluid with intracystic hemorrhage.

• Received : March 6, 2008
• Accepted : June 13, 2008
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solitary EG of the skull with an epidural hematoma and cyst is extremely rare. The causes of epidural hematomas in these cases were suggested that a rupture of the tumor cyst communicating with the epidural vein and the sinus pericranii or a head trauma, and intratumoral bleeding into the epidural space.

Herein, we report a case of midline frontal EG associated with an epidural hematoma.

CASE REPORT

A 10-year-old boy was referred to our hospital because of a progressive mass in his frontal region that had lasted for one month and three days prior to admission after struck of his head on horizontal metallic bars. The findings of physical and neurological examination were unremarkable, except for a tender mass, 3×3 cm in diameter, in his frontal region. The mass was painless, soft and elastic, and fluctuation or pulsation was not noted. There was no vascular bruit. Skull x-ray films documented a 2.5×2 cm round and lytic bony defect with sharp edges and no sclerosis in the frontal region overlying the sagittal sinus.(Fig. 1A). Cranial computerized tomography (CT), with and without contrast enhancement showed an epidural hematoma and cyst on the frontal bone (Fig. 1B). The epidural hematoma was mainly located on the midline and included subacute components. Magnetic resonance imaging (MRI) revealed an epidural hematoma with a cystic mass depressing the sagittal sinus. It was as thick as 1.7 cm in some areas. The hematoma showed heterogeneous signals, which implied that cystic fluid and blood had accumulated at different times (Fig. 1C, D). No other lesions were observed on a whole-body scan and a bone survey. The results of peripheral blood tests and a bone marrow biopsy were normal with no eosinophilia.

The mass was removed surgically. Intraoperatively, a protruding mass extending from the scalp through the defect in the frontal bone to the epidural space was encountered when the scalp flap was reflected. While removing the bone flap, it was noted that the epidural

Table 1. Summary of six cases of eosinophilic granuloma associated with epidural hematoma

Case	Age (years)	Sex	Symptoms	Trauma history	Site
1[5]	4	F	Headache, nausea, vomiting	Yes	Occipital
2[3]	8	M	Headache, vomiting	Yes	Occipital
3[1]	2	M	Sudden loss of consciousness	No	Occipital
4[6]	9	M	Headache, vomiting, somnolence	Yes	Occipital
5[7]	9	M	Headache, nausea, vomiting	No	Occipital
6 (present case)	10	M	Progressive mass	Yes	Frontal

Fig. 2. A : After the scalp flap is removed, an osteolytic and cystic bone mass is exposed. B : An epidural hematoma and fluid are observed in the cystic mass . C : Cranioplasty has been performed using bone cement.

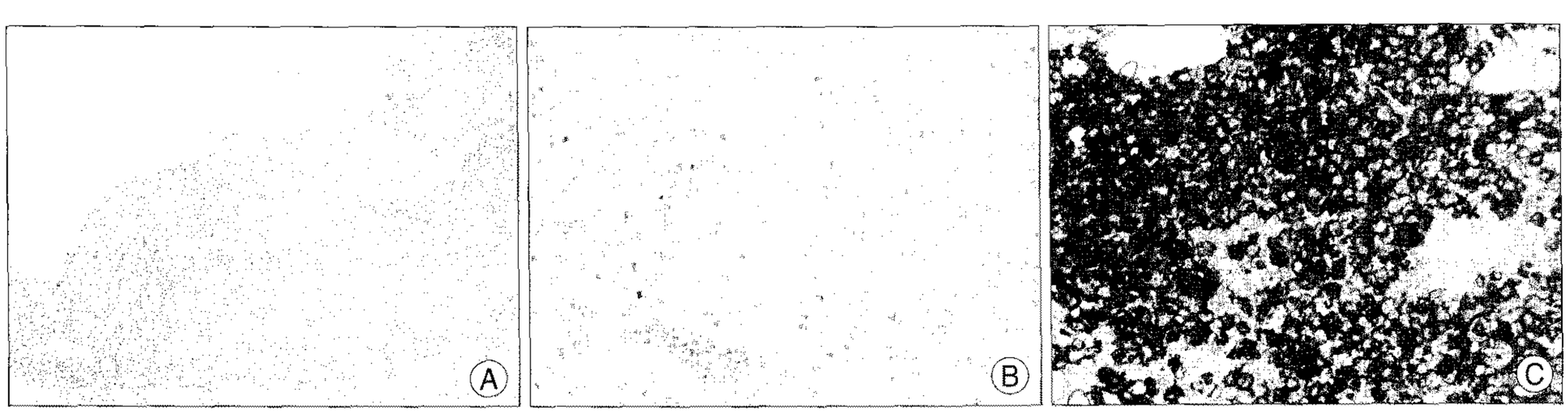

Fig. 3. A : The cystic mass is consisted of a sheet of tumor cells and hemorrhage (H-E, x100). B : The tumor cells has irregularly contoured nuclei, abundant cytoplasm admixed with eosinophils (H-E, x400). C : Immunohistochemical staining for S-100 was strongly positive (x400).

hematoma and cyst fluid were surrounded by a cystic membrane. The epidural cyst and hematoma were removed. The dura mater was intact, and the superior sagittal sinus was depressed without tumor invasion. There were no definite bleeding foci. The tumor and surrounding bony edges were completely removed. The bony defect was replaced with bone cement (Fig. 2). The postoperative course was uneventful.

Histologically, the tumor had a cystic portion. The wall of the cyst was densely infiltrated by histiocytes and showed hemorrhagic change. The outer surface of cyst was densely fibrotic, and tumor cells were clustered below the cyst. The tumor cell nuclei were grooved and irregularly contorted. The cytoplasm was abundant and eosinophilic. Mitosis was not present. The tumor cells were stained for S-100. These findings were consistent with Langerhans' cell histiocytosis (Fig. 3).

DISCUSSION

The incidence of LCH is estimated at 0.54 per 100,000 annually[4]. EG, the mildest form of LCH, is usually benign and accounts for 70% of all cases of LCH. Hand-Schüller-Christian disease and Letterer-Siwe syndrome represent multifocal EGs and a malignant fulminant lymphomatous disease of infancy, respectively[7].

The most common presentation of EG is a painful, immobile scalp mass in the parietal and frontal bones. Skull radiographs show a characteristic punched-out, sharply defined, lytic lesion of the skull vault. Although unifocal involvement of the skull is common, multifocal involvement occurs in more than 20% of patients and should be further investigated with imaging. The acute (or subacute) presentation of EG with an epidural hematoma seems to be exceptional.

Treatment of unifocal histiocytosis of the skull should involve the use of split-thickness cranioplasty to completely resect the lesion and obtain clean bone margins. This treatment is associated with a very low recurrence rate, and adjuvant therapy is not indicated. However, patients with multifocal lesions, recurrent cases or progressive disease may be treated with low-dose radiation or chemotherapy[4]. Although it is considered a benign lesion, the ultimate outcome of EG is not fully predictable. Both spontaneous regression and recurrence after surgical excision have been reported in the literature. There are frequent local and distant recurrences in series with longer follow-up periods. Therefore, follow-up evaluations should be carried out for at least 10 years.

There have been five reported cases of EG associated with epidural hematoma (Table 1). The authors of these studies speculated that the possible mechanisms for the development of epidural hematoma were rupture of the tumor cyst, communicating with the epidural vein and the sinus pericranii[5] and trauma related intratumoral hemorrhage, which subsequently penetrated into the epidural space[1,3,6,7]. All five cases except our case had occipital lesions, and four of the six patients had a previous history of head trauma. Our case also had had a minor head injury. Although it is not clear whether trauma plays a role in the development of EG or whether the injury is the event that attracts attention to a previously unnoticed lesion, head injury may precipitate intratumoral bleeding and subsequent symptom presentation.

CONCLUSION

The present case shows the characteristic feature of frontal bone involvement with an epidural hematoma and intracystic hemorrhage. It is suggested that frontal bone EG might present acutely with an epidural hematoma. We report the first case of LCH presenting as an intracranial epidural hematoma on frontal area.

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