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Case report

Eosinophilic granuloma of bone: Two case reports

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Abstract

Eosinophilic granuloma (EG) is a benign, self-limiting disorder that usually involves a single bone. However, there is a growing evidence that the clinical picture of EG is protean. We report two cases with EG that showed rare presentations.

Case 1: A 14-year-old girl complained of headache in the left parietal region for several days. The initial examination was normal. During the next three weeks, her headache was progressive and she noticed a tender swelling on her head. Cranial computed tomography (CT) revealed an osteolytic lesion on the left parietal bone. On magnetic resonance imaging (MRI), the lesion corresponded to a tumor that arose from the intradiploic region, and showed both extracranial and epidural extension. She underwent tumor resection and a diagnosis of EG was made on pathological examination. An immunohistochemical study with Ki-67 suggested accelerated growth of the tumor cells.

Case 2: A 1.9-year-old boy suddenly complained of a pain in the back and soon had difficulty in walking without help. Several days later, he became unable to sit or walk. On examination, he had spastic paraplegia in addition to painful swelling on the back. A myelogram showed a block just below the T2 vertebra. Chest CT scanning disclosed that a tumor lying posterior to the T2 vertebra was causing marked cord compression and destruction of the posterior elements of the spine. The tumor extended at T1–T3 vertebral levels. He underwent tumor resection and recovered neurological ability.

EG should be considered as a differential diagnosis for patients with osteolytic lesions who exhibit aggressive clinical features.

Keywords: Eosinophilic granuloma; Langerhans' cell histiocytosis; Histiocytosis X; Bone; Neoplasm

1. Introduction

Langerhans' cell histiocytosis (LCH) is a rare, childhood disorder characterized by abnormal proliferation of Langerhans' cells, and affects many organs such as the skin, bone, liver or lungs [1,2]. LCH was previously known as histiocytosis X, and includes the clinical entities of eosinophilic granuloma, Letterer-Siwe disease, and Hand-Schullar-Christian disease [1].

In LCH, eosinophilic granuloma (EG) is the most common form and involves single or multiple bones [1]. EG is a benign disease, but recent studies indicate that it shows variable clinical expressions. We report the rare case of a young girl who showed progressive headache accompanied by rapidly expanding EG in a parietal bone. In addition, one of the authors (K.O.) experienced EG in an infant who showed rare clinical and radiological features, some 25 years ago. This patient is presented as case 2 in this report. EG may mimic malignant disorders that can cause osteolytic lesions [3].

2. Case report

Case 1: A 14-year-old female complained of left parietal headache for several days. At the first visit to our clinic, neurological examination was normal and there

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was no abnormality on blood examination or cranial computed tomography (CT) (Fig. 1A). During the next three weeks, her headache was progressive with nausea or malaise, and she noticed a tender swelling on her head. There was no history of head trauma. Physical examination revealed a soft mass in the left parietal region. Second CT scanning disclosed an osteolytic change in the left parietal bone (Fig. 1B). On magnetic resonance imaging (MRI), the lesion corresponded to a tumor that arose from the intradiploic region, and showed both extracranial and epidural extension (Fig. 1C and D). The outer table was more eroded than the inner one by the tumor, which showed marked enhancement after gadolinium injection (Fig. 1C and D). There was also extensive enhancement of the dura mater (Fig. 1D). The left parietal cortex was slightly depressed by the tumor. Bone scintigraphy demonstrated enhancement of the left parietal bone, but there was no other abnormality.

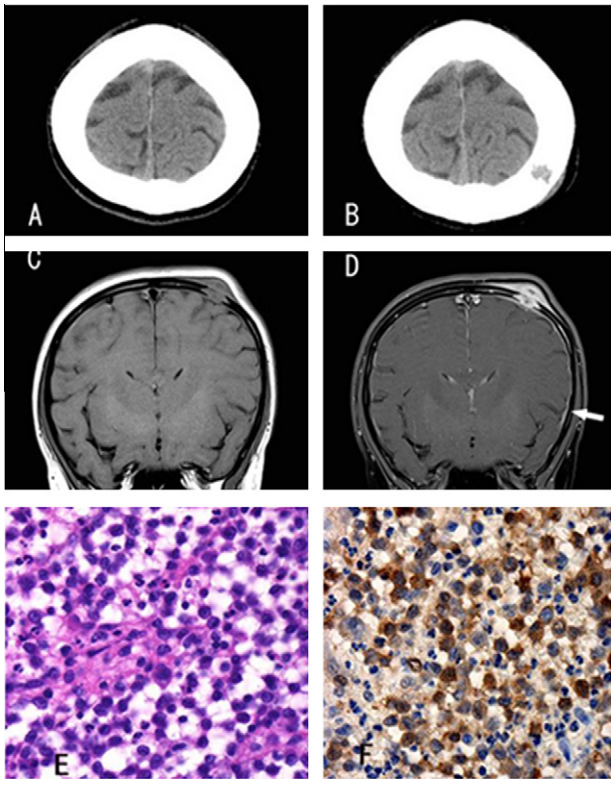

Fig. 1. Results for case 1. (A) Cranial CT scan performed at first visit. There is no bony defect. (B) Second CT scan performed three weeks after the first visit. An osteolytic change is seen in the left parietal bone. There is also slight extracranial swelling. (C) T1-weighted MRI, coronal image. Tumor is heterogeneous in signal intensity. (D) T1-weighted MRI after gadolinium administration. Note marked contrast enhancement of the tumor, which shows both extracranial and epidural extension. There is also extensive enhancement of the dura mater (arrow). (E) Photomicrograph of a pathological specimen. There are infiltrates of histiocyte-like cells that have single, irregularly contoured nuclei. Lymphocytes and neutrophils are also seen (H&E). (F) Histochemical staining with S100 protein. A significant portion of the tumor cells are S100-positive.

The tumor was totally resected operatively and a pathological specimen showed features of EG: There were sheets of histiocyte-like cells in the lesion that had irregularly contoured or folded nuclei (Fig. 1E) and were stained positively with S100 protein (Fig. 1F). Infiltration of lymphocytes and neutrophils was also seen (Fig. 1E). Moreover, an immunohistochemical study with Ki-67 showed that the tumor cells had a labeling index of 20%. She has now been free of symptoms for 7 months.

As was mentioned in the introduction, one of the authors (K.O.) previously experienced a rare case of EG in a male infant, who will be presented as case 2 hereafter. The clinical description of this patient is based on the medical records at the time.

Case 2: A 1.9-year-old boy suddenly complained of a pain in the back and soon had difficulty in walking without support. Over the following several days, his neurological condition progressively worsened, and eventually he could not sit or walk alone. There was no history of trauma. On examination, he exhibited painful swelling on the right upper part of the back, which was close to the upper thoracic vertebrae. He had weakness of the legs. The muscle tone in the lower limbs was increased, and deep tendon reflexes were hyperactive on both sides. Plantar responses were extensor, and ankle clonus was sustained bilaterally. Temperature and pain sensation were diminished in the legs, although it was difficult to determine the exact sensory levels. Abdominal reflexes were diminished bilaterally. Kernig's sign was absent, but there was nuchal rigidity. Movement of the craniofacial and arm muscles was normal. Other physical examination was not remarkable. After admission, he gradually took the equinovarus position.

Laboratory investigation showed that the white blood cell count was 10,300/mm^3 and the erythrocyte sedimentation rate was increased to 75 mm/hour. C-reactive protein was 1(+). In addition, there was elevation of urinary dopamine excretion (1.43 mg/day vs. 0.15–0.95 mg/day for controls), but other test results including catecholamine levels were normal. Cerebrospinal fluid examination revealed cell-protein dissociation: cells 2/3 mm^3, protein 111 mg/dl, sugar 54 mg/dl, Cl 129 meq/l.

A plain radiograph of the spine was lost. A myelogram disclosed a block just below the T2 vertebra (Fig. 2A). It was also noted that the pedicle and transverse process of T2 were missing on the right (Fig. 2A). CT myelography demonstrated that a low density mass extending between the T1–T3 vertebrae caused shifting of the spinal cord to the left (Fig. 2B and C). The cord was markedly compressed at the level of T2 by the tumor, which lay posterior to the vertebral body (Fig. 2C). Furthermore, the posterior elements of the T2 vertebra were missing or destroyed on the right,