CASE REPORT

Multidetector CT findings of calvarial eosinophilic granuloma

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SUMMARY

Eosinophilic granuloma (EG) is a well-recognised benign form of Langerhans-cell histiocytosis. The estimated incidence of EG is 3–4 per million of the population. More patients are children and adolescents between the ages of 1 and 15 years. The value of plain films in the diagnosis of the EG is limited and the extent of the disease is better defined by multidetector CT (MDCT) scanning. MDCT scan with multiplanar and three-dimensional reconstruction image of the skull and brain clearly demonstrates bone destruction and soft-tissue involvement of the tumour. In this article we present the MDCT imaging characteristics of EG of the skull and discuss the differential diagnosis of this rare lesion with reference to recent literatures.

BACKGROUND

Eosinophilic granuloma (EG), also called histiocytosis X, is a disorder of unknown aetiology characterised by the presence of destructive granulomas containing numerous Langerhans cells (LCs).^{1–3} EG is known as a granulomatous lesion that often shows an aggressive course. However, it remains unclear whether the massive accumulation of LCs at the site of the lesion results from the abnormal proliferation of these cells or from the inflammatory responses initiated by activated LCs.³ EG is characterised by a single or multiple skeletal lesions, and predominantly affects children, adolescents and young adults. EG of the skull is very rare and most frequently seen in children or adolescents. Although imaging appearances of EG have been described, articles on the multidetector CT (MDCT) appearances of calvarial EG are sparse.^{4–5}

In this article we present the MDCT imaging characteristics of EG of the skull and discuss the differential diagnosis of this rare lesion with the recent literature.

CASE PRESENTATION

A 5-year-old male patient was referred to our clinic with headache and soft tissue swelling in the right parietal region that was present for 3 months. On physical examination, there was a solitary mass lesion in the right posterior parietal region of the head with soft tissue swelling. The parent had no idea when they first felt the lesion but they had the impression it was slowly increasing over time. There was no history of trauma or vomiting associated with the headache. Neurological examination was normal. Skeletal radiographic survey, bone scan, chest radiographs, complete blood cell count, liver function tests, coagulation studies and electroencephalography were normal.

INVESTIGATIONS

Plain radiograph of the skull, anterior–posterior projection revealed a single punched out area of bone destruction with sharp margins in the parietal region. Cranial MDCT scan with multiplanar reconstruction showed an osteolytic lesion on the right parietal skull with a large epidural and subcutaneous soft tissue density mass (figure 1). Three-dimensional reconstruction images were performed using a MDCT scanner, a significant protuberance on the scalp and a large, well-defined, irregular-marginated, osteolytic lesion involving the right parietal bone was detected (figure 2). A preliminary diagnosis was compatible with calvarial EG based on the characteristic clinical and radiological findings.

TREATMENT

Surgical removal was performed in the supine position under general anaesthesia, which found the dark red, soft mass adhering to the dura matter and to the galea. The lesion was totally removed together with the surrounding skull by a margin of 2 cm. Surgery was completed by primary skin closure after the cranioplasty with titanium plate. Histological examination confirmed an EG characterised by the proliferation of the histiocytes intermingled with multinuclear giant cells.

OUTCOME AND FOLLOW-UP

Postoperative course was uneventful, and the follow-up CT showed complete disappearance of the lesion. The patient successfully passed the entrance examination to the primary school and follow-up examination 1 year after the first presentation showed no recurrence of calvarial EG.

DISCUSSION

Calvarial EG is a disorder of benign and unknown aetiology characterised by the presence of destructive granulomas.⁶ The exact mechanisms responsible for the aggressive course such as rapid growth and invasion are unclear, although it is often assumed that the accumulation of LCs in the disease results from abnormal proliferation of these cells.⁷ Some recent studies, based on molecular analyses, have demonstrated that Langerhans-cell populations within Langerhans-cell histiocytosis (LCH) lesions are clonal in origin. These data strongly suggest that somatic genetic changes can contribute to the aetiology and pathogenesis of LCH, as documented in neoplastic disorders.^{8–9} EG affects children and young adults, particularly males. Solitary EG accounts for the majority of LCH cases, usually involving bone and less commonly the lymph nodes, lung or skin.¹⁰ The solitary bone lesion may be asymptomatic, or it may

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Figure 1 Axial (A), sagittal (B), coronal (C) section cranial multidetector CT scan shows an osteolytic lesion on the right parietal skull with a large epidural and subcutaneous soft tissue density mass (arrows).

cause bone pain because of the expansion of the medullary bone; pathological fractures may ensue.^{11 12} In a recently published case presentation, it was reported that a patient with solitary calvarial EG was presented with epileptic seizures.⁹

Any bone can be involved. The skull, long bones of the upper extremities and the flat bones are affected in descending order of frequency.¹⁰ In a study conducted by Okamoto *et al*¹³ it was revealed that the skull is most frequently involved, the calvaria more often than the base, especially the parietal region. Solitary lesions are more common than multiple ones. When the lesions are multiple, new osseous lesions occur within 1 or 2 years but the condition is still classified as EG. Radiologists need to be aware that additional EG of the bone occurring as long as 4 years after initial diagnosis should be interpreted as a localised form of LCH. This differentiation is important because the prognosis is more favourable with focal disease rather than with multifocal disseminated disease, which involves organs other than the skeletal system.¹⁴

The hallmark of LCH is the proliferation and accumulation of a specific histiocyte: the Langerhans cell. In bones this may cause pain and adjacent soft tissue swelling and might cause headache, as in our patient's case, but some lesions are asymptomatic. EG in the cranium usually presents as a fixed hemispherical mass.^{15 16}

The role of radiology in the assessment of EG of the bone was first described by Brisman *et al*.¹⁵ Imaging diagnosis of the disease in bones is first based on the plain radiographic appearance, which is usually a central destructive, aggressive-looking lesion. In the skull, the lesions develop in the diploic space, are lytic, and their edges may be bevelled, scalloped or confluent, or show a 'button sequestrum'.^{13 15 16} The value of plain films is limited and the extent of the disease is better defined by CT scanning. Punched-out radiolucent defect without reactive sclerosis or periosteal thickening is usually detected, which is similar to various osteolytic lesions.¹⁶ CT is better suited for demonstrating bone detail, MRI for bone marrow and soft-tissue involvement and to assist with planning a biopsy or a surgical excision. In our patient, CT scan with three-dimensional reconstruction image of skull clearly demonstrates bone destruction

and soft-tissue involvement of the tumour. Radionuclide bone scan can also be used to detect the bone involvement as well as to rule out recurrence or regrowth after surgery.^{12 13}

The differential diagnosis of a solitary lytic skull lesion in a child or young adult includes epidermoid or dermoid cysts, and other benign or malignant skull tumours such as osteoblastoma, haemangioma or osteogenic sarcoma.¹⁷⁻¹⁹ Epidermoids and osteoblastomas usually have a sclerotic rim,¹⁷ haemangiomas a characteristic honeycomb or sunburst pattern of bony spicules radiating from the centre of a radiolucent round or oval defect; their margins are usually not sclerotic or bevelled on plain films or CT.²⁰ Osteosarcomas have irregular, dense borders, or a poorly defined sclerotic area.²¹ Although our observations are compatible with previously described cases of EG,^{4 5} no single finding is specific to the disease.¹⁷⁻¹⁹ However, the constellation of these findings seems to be suggestive of EG.

Treatment for LCH is usually reserved for patients with symptomatic lesions. Single bone lesions are treated with curettage.^{12 15 22} However, in very rare instances, the spontaneous healing of EG has been reported in recent literature.^{2 13} Recurrent lesions are best managed with irradiation or short courses of systemic chemotherapy. More extensive disease involvement requires systemic chemotherapy and, in some cases, marrow transplantation.^{15 16} The prognosis for LCH is uncertain. Young age and signs of organ dysfunction predict a poor prognosis.⁶ The prognosis of unifocal EG is very good, with successful disease control being achieved in 95% of cases.²³

In conclusion, EG is a benign disorder that affects children and young adults, particularly males. The clinical and radiographic findings are often not specific enough to determine the diagnosis. CT plays an important role to demonstrate osseous perforation which may result in an extradural or extracranial soft-tissue mass with the advantages of the multiplanar and reformatted three-dimensional images. For symptomatic solitary calvarial EG, surgical excision is the main treatment option.

Figure 2 Three-dimensional reconstruction of multidetector CT scan shows a significant protuberance on the right parietal scalp (A) and a large, well-defined, irregularly margined, osteolytic lesion involving the right parietal bone was detected (B) (arrows).

Learning points

- ▶ Eosinophilic granuloma is a rare disorder of benign and unknown aetiology characterised by the presence of destructive granulomas and affects children and young adults, particularly males.
- ▶ Any bone can be involved. The skull, long bones of the upper extremities and the flat bones are affected in descending order of frequency.
- ▶ The clinical and radiographic findings are often not specific enough to determine the diagnosis. CT plays an important role to demonstrate osseous perforation which may result in an extradural or extracranial soft-tissue mass with the advantages of the multiplanar and reformatted three-dimensional images.

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