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CASE REPORT

# Eosinophilic granuloma of the occipital bone in an adult: A case report

## 成人枕骨嗜伊紅性肉芽腫 — 病例報告

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**Abstract** Eosinophilic granuloma (EG) refers to the most common and benign form of the disorder known as Langerhans' cell histiocytosis. The disease is typically found in children and adolescents and rarely affects adults. We present a case of EG in the occipital bone in a 36-year-old man, who visited our hospital with the chief complaint of left occipital palpable tumor mass with local tenderness and pain for one month. An X-ray of the skull revealed a rounded osteolytic lesion. A computed tomography scan revealed a shadow of soft tissues in the left occipital site involving the entire thickness of the calvaria, which was indicative of marked destruction of the bone. The soft mass was successfully removed. The margins of the skull lesion were excised, and cranioplasty was performed simultaneously with bone cement. A definitive diagnosis of EG was made by histopathology and immunohistochemical detection of S-100 antigen in the tissue samples. With respect to management, we believe surgery is the best option for most accessible cranial lesions of EG. A cranioplasty with bone cement or autologous bone can be performed in the same session to repair the cranial defect.

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**摘要** 嗜伊紅性肉芽腫是蘭格罕細胞組織球增生症(Langerhans cell histocytosis)中最常見，且為良性的型態。此疾病常發生在兒童及青少年，成人的案例較少見。我們報告一個成人枕骨肉芽腫的案例。此一36歲男性主訴枕骨左側有局部壓痛的腫塊約月餘而來本院求診。頭骨X光顯示一圓形蝕骨病灶，頭部電腦斷層掃描則發現一軟組織病灶在枕骨左側且枕骨整層明顯受到破壞。手術將此軟組織病灶完整切除，病灶邊緣的頭骨亦將之切除，並施與顱骨整形術。術後的組織病理切片及免疫染色(S-100抗體)確診為嗜伊紅性肉芽腫。關於治療，我們認為，對於手術能夠切除的顱骨嗜伊紅性肉芽腫，手術是最佳的選擇。同次手術中，並施與以人工骨水泥或自體骨為材料的顱骨整形術來修補顱骨的缺損。


## Introduction

Eosinophilic granuloma (EG) represents the focal form of Langerhans' cell histiocytosis (histiocytosis X) [1]. Langerhans' cell histiocytosis refers to a group of rare syndromes, including EG, Hand-Schüller-Christian disease, and Letterer-Siwe disease, which vary greatly in their clinical courses. These three diseases share a common histopathology, an abnormal proliferation of dendritic antigen presenting in histiocytes, known as infiltrate of lymphocytes, eosinophils, and neutrophils, which results in the destruction of a variety of tissues [2]. Lesions may occur either locally or as widespread systemic disease. Typically, EG involves either unifocal or multifocal lesions; Hand-Schüller-Christian disease is a subset of multifocal EG with a triad of signs (lytic skull lesions, exophthalmos, and diabetes insipidus); and Letterer-Siwe disease is a systemic histiocytosis, which primarily involves soft tissue sites, such as malignant lymphoma of infancy [3]. Although some authors have recently demonstrated clonal proliferation in all forms of Langerhans' cell histiocytosis, suggesting a neoplastic disorder, the precise etiopathogenesis of the disease is still unknown and the clinical course unpredictable [4,5]. EG is uncommon, representing less than 1% of tumor-like lesions of the bone. The estimated incidence of EG is three to four per million, occurring predominantly in children and adolescents [6]. We report an unusual case of EG in a 36-year-old male presenting with a solitary lesion at the occipital skull bone.

## Case presentation

A 36-year-old male was admitted to our hospital with the chief complaint of left occipital palpable tumor mass with local tenderness and pain for one month. An X-ray of the skull revealed a rounded osteolytic lesion (Fig. 1). A computed tomography (CT) scan of the brain performed with specific windows for bony structures showed a circumscribed osteolytic lesion located in a left occipital site, involving the entire thickness of the calvaria (Fig. 2A). Furthermore, a CT scan of the brain revealed tissue of parenchymal intensity in a left occipital site, involving the subcutaneous soft tissues and partially extending internally. Administration of a contrast medium resulted in marked enhancement (Fig. 2B and C). Given the soft tender mass of 3 cm in diameter in his left occipital region, it was decided to operate on the patient.

The patient underwent craniectomy and removal of the soft tissue mass. The dura mater was intact. The margins of the skull lesion were excised, and the osseous gap was filled with bone cement.

Histopathology revealed many folded nuclei of histiocytes (Langerhans' cells) with smaller and dispersed eosinophilic and multinucleated cells (Fig. 3A). Immunohistochemical stain for S-100 showed strong positivity of tumor cells, confirming the diagnosis of Langerhans' cell histiocytosis (Fig. 3B).

At the 36-month follow-up, the patient was asymptomatic and no recurrence was noted.

## Discussion

EG is uncommon, representing less than 1% of tumor-like lesions of the bone. Slater and Swarm [2] have reported that the skull is the most common site of EG (43%), whereas the femur is the next frequent site. Arseni et al. [7] reported that 80% of patients with EG presented with a solitary skull lesion. The lesion in our case was monostotic and solitary; no other site of involvement was demonstrated on a skeletal survey.

The most common presenting symptom of EG of the skull is a tender, gradually enlarging mass frequently seen in the

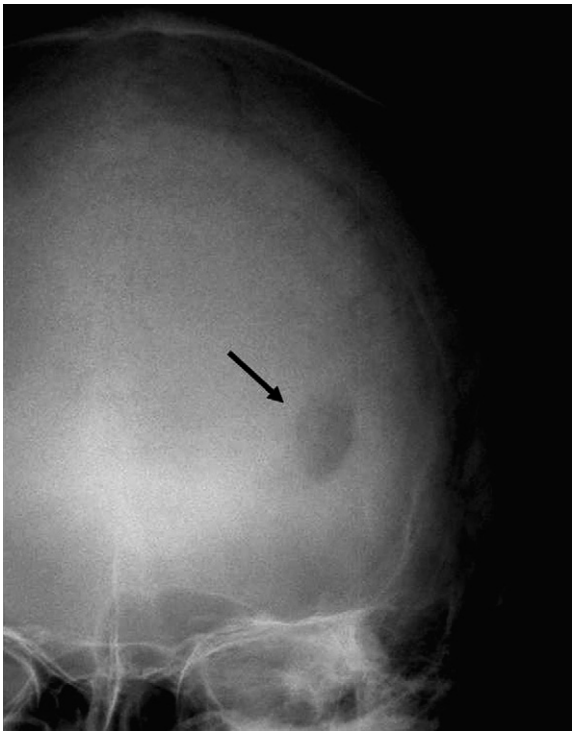

**Figure 1.** An X-ray of the skull revealed a rounded osteolytic lesion (black arrow).

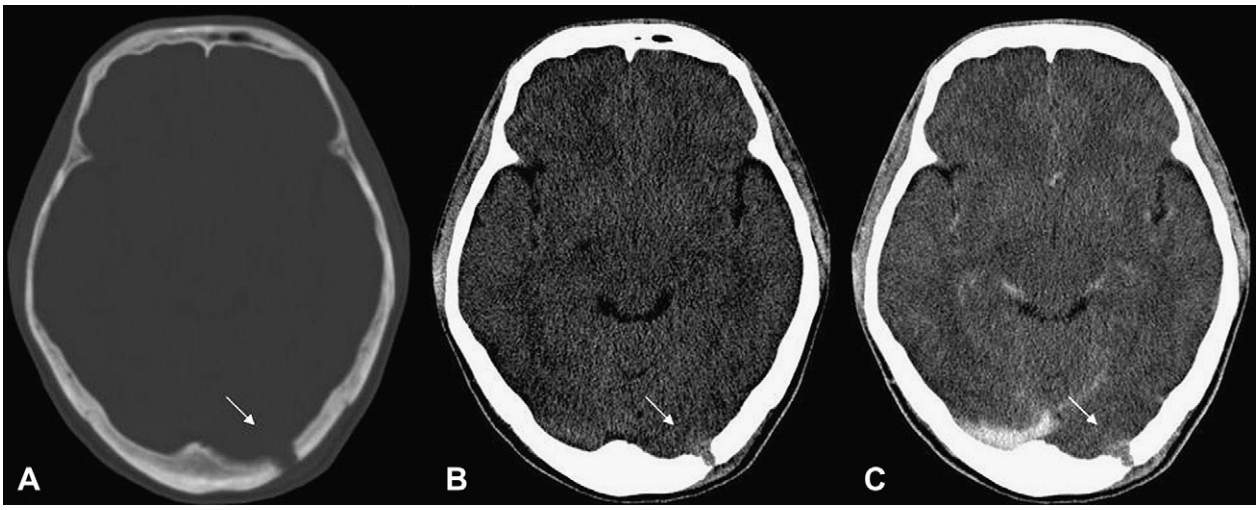


**Figure 2.** (A) Computed tomography scan with bone windows showing a circumscribed osteolytic lesion located in a left occipital site involving the entire thickness of the calvaria (white arrow); (B) Computed tomography scan of the brain showing tissue of parenchymal intensity in a left occipital site involving the subcutaneous soft tissues and partially extending internally (white arrow); (C) Administration of contrast medium resulted in marked enhancement (white arrow).

parietal and frontal bones. At the time of diagnosis, more than 90% of patients have a skull mass [8]. Ardekian et al. [9] reviewed 25 patients with 41 EGs and found pain to be the most common presenting symptom (92% of patients), often accompanied by swelling.

Although most bone lesions can be accurately detected and differentiated using radiography, CT is the preferred imaging technique for studying cranial vault lesions. CT is favored mainly because of its superior ability to depict cortical bone and to allow analysis of internal characteristics. Radiological and clinical differentiation between neoplastic and nonneoplastic lesions of the calvaria is one of the neurosurgeon's most difficult tasks [10]. Of the destructive neoplastic and nonneoplastic lesions of the calvaria, the most important in the differential diagnosis with EG are osteomas (benign tumors), epidermoids, dermoid cysts, dermal sinus, vascular tumors, osteogenic sarcoma (malignant sarcoma), metastatic disease, meningiomas, and infectious pathological conditions [10,11].

Treatment of EG depends on the extent to which the disease has progressed. Potential therapies include surgery, radiotherapy, and chemotherapy, alone or in combination. Surgery is generally indicated for isolated lesions in which an appropriate curettage may lead to complete removal of the lesion. Wide resection with tumor-free margins provides the best chance for cure of EG. Cranioplasty is necessary when the EG exceeds 3 cm in diameter or if it is located in an exposed region of the skull.

Use of radiotherapy is generally limited to lesions that are not resectable or to cases of subtotal resection. Rawlings and Wilkins [7] did not find any local recurrences in a series of patients with EG of the calvaria treated with postoperative radiation therapy. Moreover, in cases of unifocal disease, the complete response rate to radiation therapy is quite good; Gramatovici and D'Angio [12] observed a radiation response of 33% in 12 patients with bone lesions. In previous reports, the dose of radiation delivered ranged from 600 to 1,200 rad.

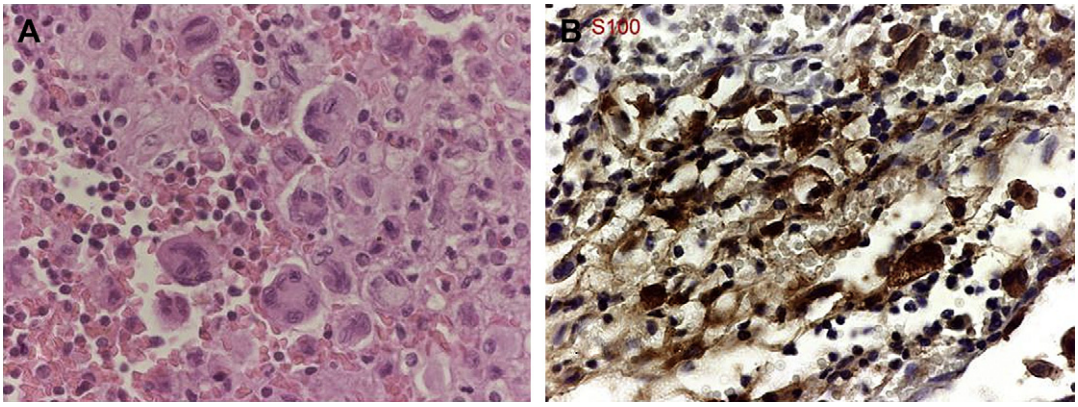


**Figure 3.** (A) Photomicrography showed many folded nuclei of histiocytes (Langerhans' cells) with smaller and dispersed eosinophilic and multinucleated cells; (B) Immunohistochemical stain for S-100 showed strong positivity of tumor cells, confirming the diagnosis of Langerhans' cell histiocytosis.

With respect to management, we believe surgery is the best option for most cases of accessible cranial lesions of EG [1]. A cranioplasty with bone cement or autologous bone can be performed in the same session to repair the cranial defect [13].

In conclusion, because EG can occur in adults, differential diagnosis of this disease must be included in adult osteolytic skull bone lesions.

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 [1,8,14]. There are frequent local and distant recurrences in series with longer follow-up periods. Therefore, follow-up evaluations for at least 10 years have been recommended in these cases [1,8,14].

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