Case Report

Malignant Hemangioendothelioma of Occipital Bone

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ABSTRACT

Epithelioid hemangioendothelioma is a rare vascular tumor of bone, and rarely these lesions can present as unique and extremely aggressive tumor. We report a case of highly aggressive epithelioid hemangioendothelioma and discuss the imaging findings. CT brain plain study revealed a poorly-defined, mixed density expansile and lytic lesion involving the occipital bone with extension to the left side with poorly defined trabecula formation. There was significant but irregular enhancement after intravenous administration of contrast material and also marked bone destruction. Microscopic examination of the fine needle aspiration cytology showed a tumor composed of vascular channels lined by plump endothelial cells, which had enlarged hyperchromatic nuclei. In view of the extensive infiltration the patient was submitted for the radiotherapy.

Key words: Hemangioendothelioma; Bone tumor; Occipital bone

INTRODUCTION

Epithelioid hemangioendothelioma (EHE) is a rare vascular tumor of bone that constitutes less than 1% of primary malignant skeletal neoplasm[1,2]. These lesions can behave like a benign or malignant tumor[3], and rarely these lesions can present as unique and extremely aggressive tumor[4]. We report a case of highly aggressive EHE and discuss the imaging findings.

CASE REPORT

A 23-year-old female was noted to have a small bony prominence over occipital region for 2–3 years. She noticed recent progressive increase in the size of the lesion over last 2 months associated with pain. She delivered a baby recently and the increase in the size of the swelling was associated with delivery. The lesion was firm and non-pulsatie (Figure 1). No similar lesions were present in other sites. No lymph nodes were palpable. CT brain plain study revealed a poorly-defined, mixed-density, expansile and lytic lesion involving the occipital bone with extension to the left side with poorly defined trabecula formation (Figure 2A). There was significant but irregular enhancement after intravenous administration of contrast material and also marked bone destruction (Figures 2B, 2C, and 3). Microscopic examination of the fine needle aspiration cytology showed a tumor composed of vascular channels lined by plump endothelial cells, which had enlarged hyperchromatic nuclei (Figure 4). On the basis of these combined findings, a diagnosis of grade II hemangioendothelioma was made. In view of the extensive infiltration, the patient was submitted for the radiotherapy.

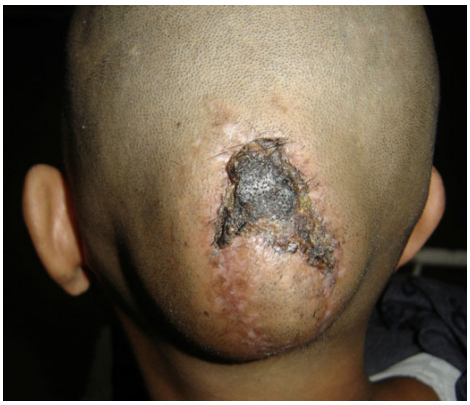

Figure 1. Clinical photograph showing large lesion with scab and previous incision.

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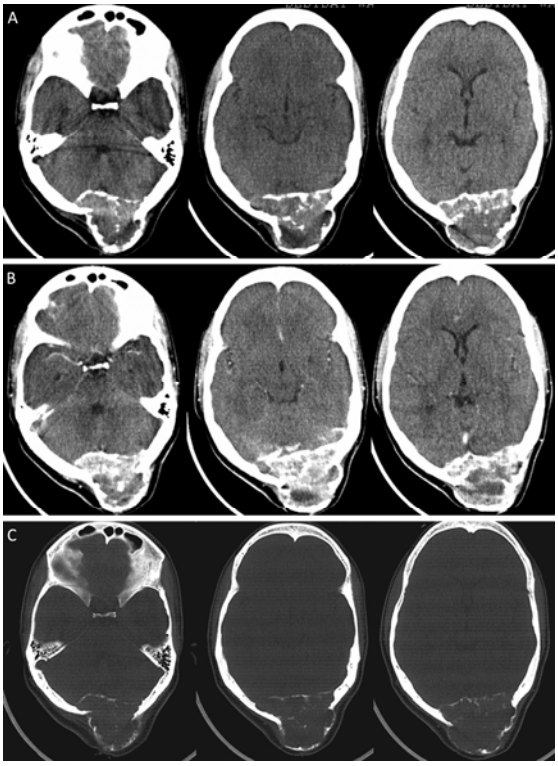

Figure 2. Plain axial CT scan images. **A:** Mixed-density, expansile and lytic lesion of the occipital bone; **B:** irregularly enhanced lesion after contrast administration; **C:** extensive destruction of the occipital bone with a poorly defined, coarse trabecular pattern and irregular border.

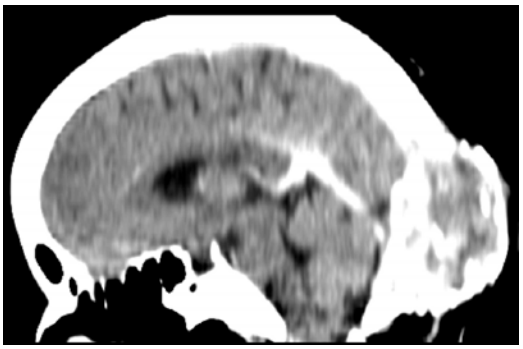

Figure 3. Contrast enhanced sagittal reconstruction CT scan images showing the expansile and lytic occipital bone lesion.

DISCUSSION

On CT and magnetic resonance imaging (MRI), EHE is characterized by a well-demarcated, osteolytic and expansile lesion involving the skull bone, which has the sclerotic edges specks of calcification[4-7] giving rise to a honeycomb configuration with trabeculation[7,8]. Although the imaging pattern of malignant EHE may be similar to that of a benign lesion[9], Unni, et al. have described a positive correlation between the radiographic picture and the histological grade[10]. They found that low-grade tumors will show sharply demarcated margins and some bony trabeculae, whereas high-grade tumors will have indistinct and irregular margins[10]. Depending on the biologic behavior and microscopic features, EHE can be categorized as between a hemangioma and a conventional angiosarcoma[1,4,11]. Lack of cytological atypia and sparse mitosis is associated with favourable prognosis and the lesion can be cured by complete wide-resection[6]. The histopathological features considered suggestive of more aggressive clinical behavior include a mitotic rate more than one per 10 high-power fields, cellular atypia, focal necrosis, and increase in proportion of the spindle cells[1,4,11]. The treatment choice for hemangioendothelioma has been surgical resection, combined with radiation, especially in high-grade lesions[8,12]. Because of their extensive vascularity, EHE lesions can lead to substantial intraoperative bleeding with resultant mortality[13]. Radiation therapy has been used alone when surgery was not feasible[14]. Chemotherapy currently has no significant role in the treatment[14]. The prognosis of EHE has not been well defined despite of the favorable outcome in the majority of cases[5,6,11,15]. It has been described that malignant hemangioendotheliomas are capable of metastasis and can lead to death [9,16].

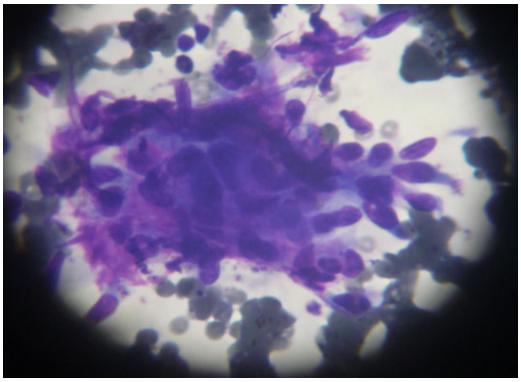

Figure 4. Photomicrograph shows proliferation of epithelioid-like endothelial cells having large hyperchromatic nuclei (HE staining, ×40).

Disclosure of Potential Conflicts of Interest

No Potential conflicts of interest were disclosed.

REFERENCES

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