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Case Report

Primary Ewing's sarcoma of the occipital bone: A case report and review of the literature

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ABSTRACT

Background: Primary Ewing's sarcoma (ES) arising from cranial bones is an extremely rare entity that accounts for only 1–4% of all ES cases.

Case Description: A 21-year-old woman presented with ES of the skull affecting the occipital region. The patient underwent surgical excision following radiotherapy and chemotherapy. No recurrence or metastasis occurred over a 10-month follow-up.

Conclusion: ES reaches adolescents. The mainstay of treatment includes surgical removal of the tumor, followed by radio- and chemotherapy.

Keywords: Ewing sarcoma, Occipital bone, Skull

INTRODUCTION

Ewing's sarcoma (ES) is a primitive malignant bone tumor that commonly occurs in the long bones of the lower limbs, pelvis, and ribs. Primary ES of the skull is extremely scarce, accounting for only 14% of all cases.[4,8] In the skull, this sarcoma typically affects the frontal, parietal, temporal, ethmoid, orbital, and maxillary bones. To the best of our knowledge, only nine cases of primary occipital ES have been reported in the literature.

Herein, we present the case of a 21-year-old woman harboring primary ES of the occipital region which was successfully treated with radical excision, followed by radiotherapy and chemotherapy.

CASE REPORT

Clinical presentation

A 21-year-old woman was admitted in June 2022 with a history of large progressive swelling over his occipital region, enlarging for a few months. There were no headaches, vomiting, seizures, or loss of vision.

On physical examination, the patient had a large solid, immobile mass over the occipital region, and no focal neurological deficits were observed.

Diagnosis assessment

Computed tomography (CT) of the head revealed a large well-circumscribed extracranial; isodense mass in the occipital region with bony erosion of the adjacent occipital bone [Figures 1-3].

Therapeutic intervention

After counseling the patient, surgical resection of the tumor was planned. The patient underwent gross total excision of the tumor. A spiculated gross periosteal reaction was observed and the calvarial occipital bone showed erosion. The mass had eroded through the underlying bone but without dura invasion. Methacrylate cranioplasty was performed thereafter [Figure 4]. Histopathological examination revealed that the tumor was an ES.

Follow-up

A postoperative CT scan confirmed the complete removal of the tumor [Figure 4]. Based on the diagnosis of ES, the tumor stage was T2, N0, and M0. The patient, then, received postoperative chemotherapy with multiple drugs: Vincristine (1.5 mg/m^2 intravenous weekly × 6), cyclophosphamide (1200 mg/m^2 intravenous weekly × 6), adriamycin (30 mg/m^2 intravenous daily × 2 every month × 6), and actinomycin-D (2 mg/m^2 intravenous once every month × 6). For

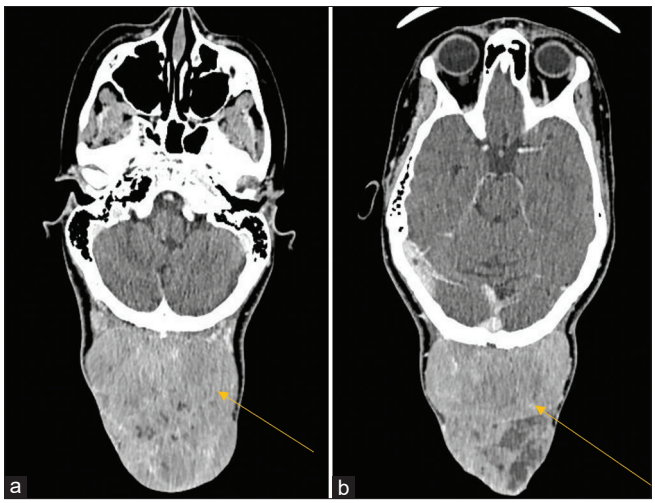

Figure 1: (a and b) Computed tomography of the head (axial): Showing an extracranial isodense mass lesion of the occipital region (arrow).

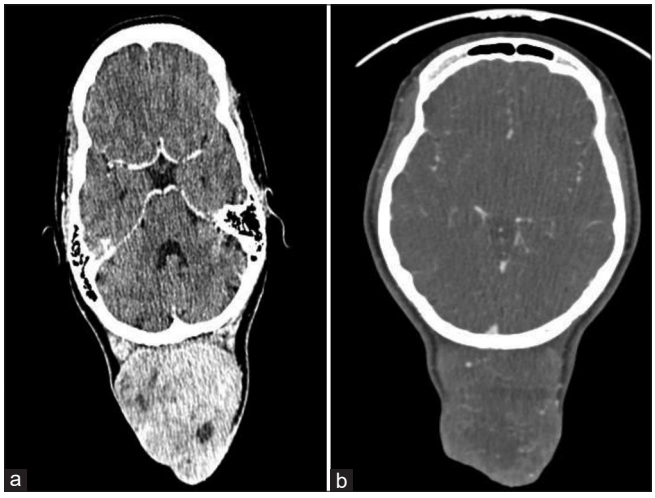

Figure 2: (a and b) Computed tomography of the head with contrast injection (axial): Showing an extracranial isodense mass lesion of the occipital region which was not enhanced after injection.

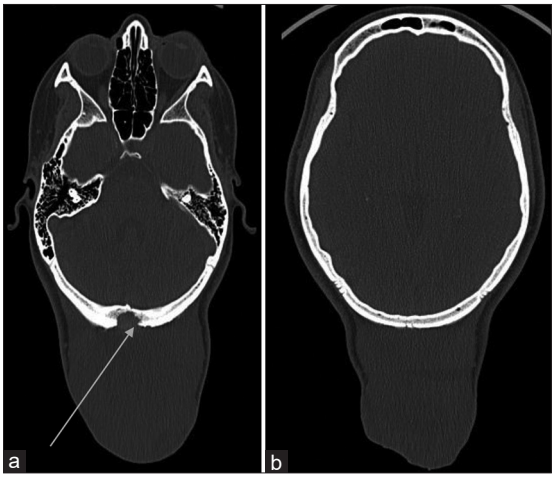

Figure 3: (a and b) Computed tomography of the head with contrast injection (axial bone cuts): Showing a bony erosion (arrow).

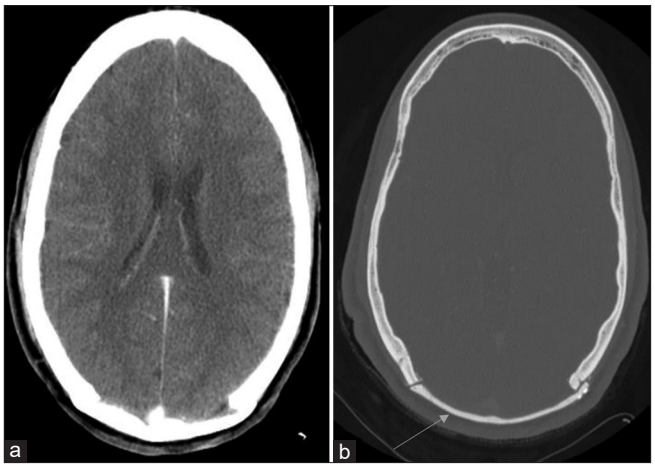

Figure 4: (a and b) Postoperative computed tomography of the head: Showing completely excised tumor with cranioplasty (arrow).

Table 1: A literature review of primary Ewing's sarcoma of the occipital bone from 1988 to date.

Studies	Age (year)/Sex	Symptoms	Type of surgery	Adjuvant treatments	Follow-up period (months)	Survival
Freeman *et al.* (1988)[2]	17/M	Headache, vomiting	Gross total excision	RT/CT	11	Alive
Hollody *et al.* (1992)[4]	7/F	Headache	Gross total excision	RT/CT	24	Alive, metastasis
Tournut *et al.* (1994)[5]	11/F	Headache, diplopia	Gross total excision	CT	18	Died due to metastasis
Zenke *et al.* (1994)[8]	12/M	Headache, vomiting, fever	Gross total excision	RT/CT	18	Alive, no recurrence
Yamashita *et al.* (1997)[6]	19/F	Headache	Gross total excision	RT	72	Alive, no recurrence
Desai *et al.* (2000)[1]	14/M	Headache, vomiting	Gross total excision	RT/CT	26	Alive, no recurrence
Desai *et al.* (2000)[1]	18/M	Headache, vomiting	Gross total excision	RT/CT	64	Alive, no recurrence
Yasuda *et al.* (2003)[7]	1/M	Bad moods, crying, gait disturbance, scalp swelling	Gross total excision	RT/CT	84	Died due to metastasis
Garg *et al.* (2007)[3]	7/F	Scalp swelling, headache, gait ataxia, blindness in the left eye	Gross total excision	NA	NA	Alive, no recurrence
Present case (2023)	19/F	Scalp swelling	Gross total excision	RT/CT	10	Alive, no recurrence

M: Male, F: Female, RT: Radiotherapy, CT: Chemotherapy

radiotherapy, the patient received 2 Gy/day, 5 days/week for 5 weeks. Ten months after the surgery, the patient remained asymptomatic and tumor-free.

DISCUSSION

In the literature, we have identified nine cases of primary ES of the occipital bone from 1988 to date [Table 1].[1-8] Typically, it occurs in the first two decades of life; however, the youngest patient has been reported to be 16 months old.[8] Interestingly, our study involved the oldest patient in the literature.

Radiologically, CT scans and magnetic resonance imaging (MRI) provide information about the size, extent, and brain/dura involvement. In general, a CT scan has documented ES as an iso-hyperdense mass that enhances after contrast injection, delineating the bone involvement. On MRI, the mass is hypo-to isointense on T1W and iso-or hyperintense on T2W, with heterogeneous enhancement following contrast injection. Therapeutic options include surgical removal of the tumor, radiotherapy, and multi-drug chemotherapy.

Regarding surgical treatment, tumor resection should be as radical as possible. The previous reports showed that long-term survival can be expected in ES, provided that the tumor is radically removed followed by radio- and chemotherapy.

CONCLUSION

ES of the occipital region of the skull is an extremely rare location. It reaches adolescents and radical surgical resection is the mainstay of treatment. Adjuvant radiotherapy and chemotherapy has been shown to improve patient survival.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The author(s) confirms that there was no use of Artificial Intelligence (AI)-Assisted Technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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