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Beyond the Bone: A Case Report and Review of Literature of Extraskeletal Chondroblastic Osteosarcoma of the Scalp

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Abstract

Extraskeletal osteosarcoma (EOS) of the scalp is an extremely rare malignant mesenchymal tumor that accounts for approximately 1 to 2% of all soft tissue sarcoma and 2 to 4% of all osteosarcoma. A 40-year-old man presented with painless swelling on the left side of the scalp for 3 months and right-side limb weakness for 10 days. The patient underwent surgery and the postoperative histopathology report was suggestive of chondroblastic osteosarcoma of the scalp. The patient received five cycles of Adriamycin-based systemic therapy. A response assessment imaging was done, which was suggestive of partial response. Osteosarcoma is chemotherapy- and radiotherapy-resistant tumor; hence, it requires a combined modality approach. Complete surgical excision with adequate surgical margin followed by adjuvant chemotherapy and radiation therapy proved to be one of the optimal treatment modalities.

Keywords

- extraskeletal osteosarcoma
- chondroblastic osteosarcoma
- soft tissue sarcoma

Introduction

Extraskeletal osteosarcoma (EOS) of the scalp is an extremely rare malignant mesenchymal tumor that accounts for approximately 1 to 2% of all soft tissue sarcomas and 2 to 4% of all osteosarcomas.[1,2] EOS usually occurs in soft tissue of the lower extremity of adults, with the thigh being the most common site, followed by the upper extremity, retroperitoneum, and trunk.[3,4] EOS mostly occurs after 40 years, with the median age at presentation of 61 years and a male predominance, with the male-to-female ratio being 1.9:1.[5,6] Previous history of radiation or trauma has been associated with EOS in 10% cases.[3,7,8] EOS has six pathological subtypes: osteoblastic, chondroblastic, fibroblastic, telangiectatic, small cell, and well differentiated.[3] EOS is an aggressive sarcoma entity and has poor prognosis as 50 to 70% present with upfront metastasis or develop metastasis within 3 years after diagnosis.[4,9] The most common site for distant metastasis is the lung, followed by bone, lymph node, liver, or peritoneum.[5]

Case Presentation

A 40-year-old man presented with complaints of painless swelling on the left side of the scalp for 3 months and right-sided limb weakness for 10 days. The swelling was rapidly progressive in nature and associated with mild headache and occasional vomiting. There was no family history of malignancy or raised intracranial tension features. On examination, the Eastern Cooperative Oncology Group performance score (ECOG PS) was 3. His motor function on the right lower limb was 2/5 and that on the left lower limb was 2/5.

Preoperative T2-weighted contrast-enhanced magnetic resonance imaging (CEMRI) brain showed a $6.6 \times 6 \times 4$ cm

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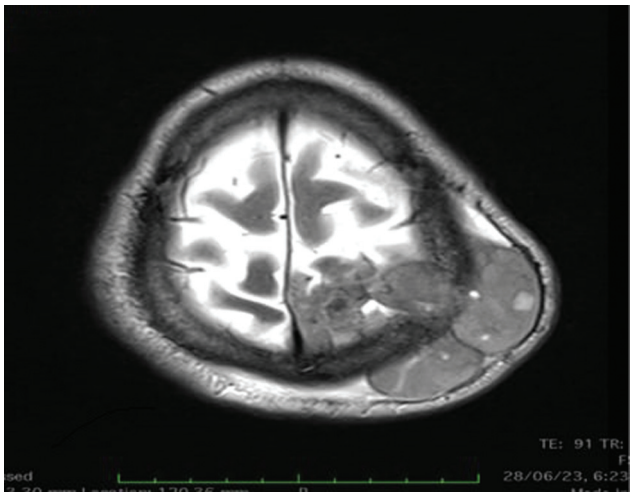

Fig. 1 Preoperative magnetic resonance imaging of the brain with contrast showing a hypointense lesion in axial view.

ill-defined lesion in the left high parietal region involving the scalp and underlying bone with dural tail enhancement (►**Fig. 1**).

A biopsy of the swelling was done, which was suggestive of a mesenchymal tumor with features of myofibroblastic differentiation. The patient underwent debulking and excision of tumor (►**Fig. 2**).

The postoperative histopathology report with microscopic examination showed tumor cells arranged in sheets and lobules with prominent osteoid matrix and osteoclastic giant cells, suggestive of chondroblastic osteosarcoma of the scalp with positivity for osteonectin and SATB2, and Ki-67 proliferation index of 80% (►**Fig. 3**).

Postoperative T2-weighted CEMRI of the brain showed a hypointense $6.5 \times 5 \times 4.8$ cm extra-axial lesion in the left high frontoparietal region with area of nodularity, infiltration of the superior sagittal sinus, moderate edema, and midline shift toward the right side. Contrast-enhanced computed tomography (CECT) of the chest did not show lung metastasis or any other distant metastasis (►**Fig. 4**). Ultrasound of the whole abdomen was done, which did not show any metastatic disease and was normal.

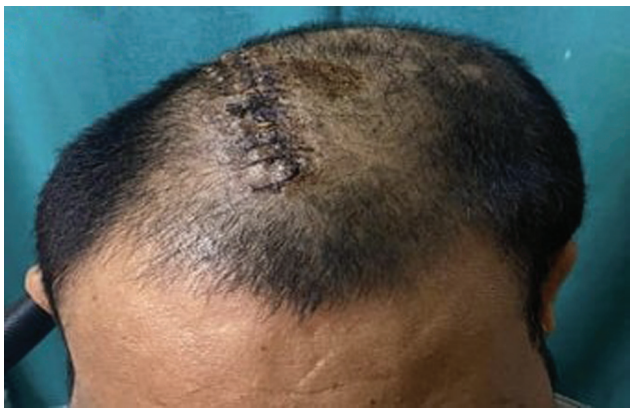

Fig. 2 Clinical picture showing postoperative swelling over the left parietal region of the scalp.

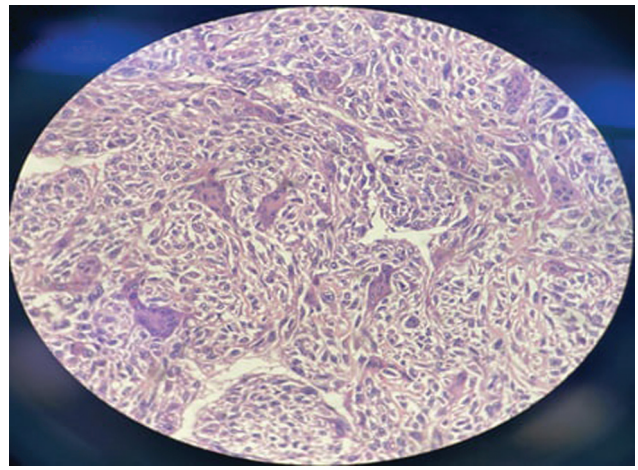

Fig. 3 Histopathology showing the tumor cells are arranged in sheets and lobules with a prominent osteoid matrix and osteoclastic giant cells present.

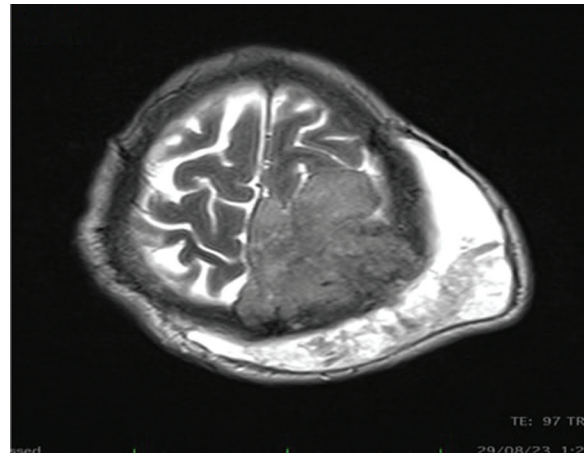

Fig. 4 Postoperative magnetic resonance imaging of the brain with contrast showing a hypointense lesion in axial view (T2-weighted [T2W]).

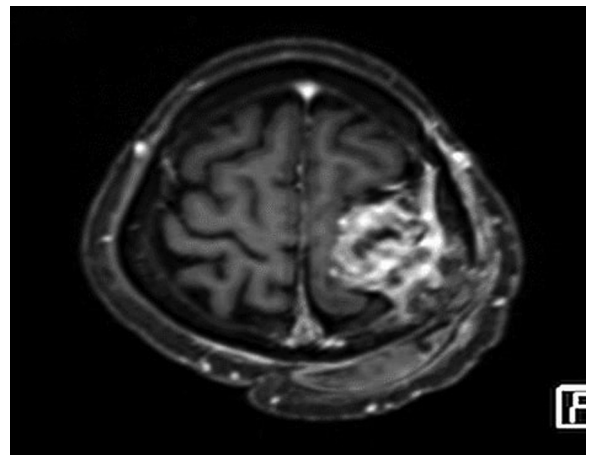

Fig. 5 Postchemotherapy magnetic resonance imaging of the brain with contrast showing a hyperintense lesion in axial view (T1 postcontrast).

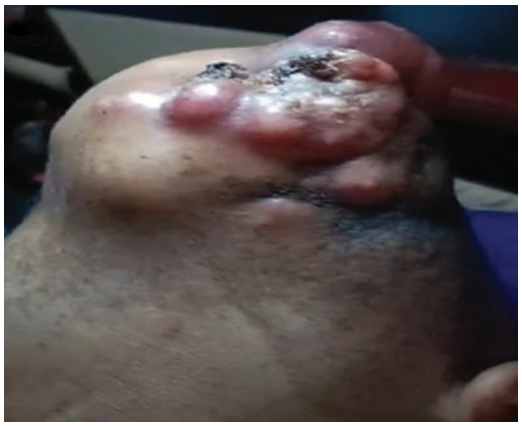

Fig. 6 Clinical picture showing disease progression in the patient.

The patient was planned for Adriamycin- and platinum-based chemotherapy. Adriamycin was given at a dose of 25 mg/m^2 (days 1–3) and cisplatin was given at a dose of 50 mg/m^2 (days 1–2) every 21 days. With subsequent chemotherapy, the patient's general condition significantly improved along with enhanced motor functions (ECOG PS of 2; motor function on the right lower limb of 3/5 and on the left lower limb of 5/5). After four cycles of chemotherapy, T1 postcontrast CEMRI of the brain showed a 4.5 × 5.3 × 4.3 cm mass lesion in the left parietal region involving the scalp, underlying brain parenchyma, and infiltrating superior sagittal sinus, suggestive of partial response to the treatment (►**Fig. 5**).

However, the patient defaulted for 3 months and later presented with an increase in the swelling size with involvement of the overlying skin with multiple nodules, suggestive of clinical progression (►**Fig. 6**).

Discussion

Osteosarcoma is mainly characterized by the production of an osteoid matrix by malignant cells, which primarily originates from long bones in adults.[8,10] EOS usually originates from deep soft tissue in extremities, but primary skull osteosarcoma is uncommon.[3,4] Previous exposure to radiation is the only known environmental risk factor

Table 1 Review of literature

Study	*n*	Histopathology (HPE)	Treatment	Follow-up	Response
Al-Janabi et al[1]	1	Osteosarcoma	Mohs microscopic surgery	38 mo	Widespread biopsy-proven squamous cell carcinoma (SCC) metastasis after 38 mo on imaging
Liao et al[2]	22	Extraskeletal osteosarcoma	Surgery followed by adjuvant chemotherapy (Methotraxate (MTX) + Ifosfamide (IFO) + Cisplatin (DDP) + Adriamycim (ADM) for 3 cycles)	3-y Overall survival (OS): 69% 5-y OS: 58%	Combination chemotherapy does not improve OS and Progression Free Survival (PFS)
Massi et al[3]	1	Osteoblastic osteosarcoma	Wide local excision	6 mo	No recurrence
Pillay et al[4]	1	Synovial sarcoma	Surgery (incomplete excision)→ Adriamycin + cisplatin × 1 cycle→ defaulted	6 mo	Recurrence in the neck
Lee et al[9]	40	Osteosarcoma	Surgery (100%) Pre-op RT: 5% Post-op RT: 22.5% Adjuvant chemotherapy: 5%	3 y	Local recurrence: 45% Of these local recurrences, distant metastasis in 61%
Paludo et al[12]	43	Osteosarcoma	Surgery f/b chemotherapy (Adriamycin ± platinum)	5 y	Platinum-based chemotherapy improves 5-year OS by 17% and PFS by 48%
Wu et al[11]	1	Osteosarcoma of frontal bone	Surgery (gross total resection)→ adjuvant chemotherapy (pirarubicin + ifosfamide) × 1 cycle→ radiation therapy (30 Gy)→ 3 cycle chemotherapy	4 y	Recurrence within 2 y→ resurgery + adjuvant chemotherapy After 4 y, the patient was free of disease
Chennupati et al[10]	1	Osteosarcoma of the skull base with leptomeningeal enhancement	Surgery→ adjuvant chemotherapy (cisplatin, doxorubicin, methotrexate, etoposide, ifosfamide)→ adjuvant radiation (40 Gy to craniospinal irradiation and 70 Gy to skull base)	–	–

and other risk factors include trauma, Paget's disease, fibrous dysplasia, enchondromatosis, and Li–Fraumeni syndrome.[3,8,11]

EOS is an aggressive sarcoma with poor prognosis characterized by local recurrence and distant metastasis. Tumor diameter more than 5.5 cm and high-grade pathology are poor prognostic factors.[2] A study by Liao et al retrospectively analyzed 22 patients after surgery and showed that the 5-year overall survival (OS) was 62% and the 5-year progression-free survival (PFS) rate was 33%.[2] EOS has 45 to 50% local site recurrence within 3 years of surgery.[2,9] Complete surgical excision with adequate surgical margin is known to improve survival. Adjuvant chemotherapy improves the 5-year OS from 20 to 60 to 70%.[11,12] In our study, the patient received anthracycline- and platinum-based chemotherapy and showed partial response to treatment. A study by Paludo et al also showed decreased relapse or recurrence rate in patients who received anthracycline- and platinum-based chemotherapy.[12] As per previous studies, Adriamycin, cisplatin, ifosfamide, methotrexate, and gemcitabine are known chemotherapeutic agents used for EOS.[9,12]

Choi et al analyzed outcomes in 53 cases with EOS, of which patients with localized disease primarily underwent surgery. After surgery, 19 patients (45%) underwent observation, 10 (24%) received adjuvant radiation, 5 (12%) underwent adjuvant chemotherapy, and 8 (19%) received both radiation and chemotherapy. They concluded that there was no significant association of disease-specific and event-free survival with the addition of radiation, chemotherapy, or both.[13] Hence, early recognition of tumor, correct diagnosis, and optimal treatment are important for better outcome.

Osteosarcoma management is done by a multidisciplinary team. Osteosarcoma is an aggressive tumor; hence, it requires a combined modality approach. Complete surgical excision with adequate surgical margin followed by adjuvant chemotherapy or radiation therapy proved to be the optimal treatment approach. Various studies reported in a review of the literature are shown in ►**Table 1**.

Conclusion

Primary EOS of the scalp is a very rare entity and can be managed in the lines of sarcoma of extremities. For patients presenting with localized extraosseous osteosarcoma, surgery is the primary treatment approach. Chemotherapeutic systemic therapy and radiation therapy can be considered as adjuvant and palliative therapies wherever feasible.

Patient Consent

Written informed consent was given by the attendant for treatment and the prognosis was explained.

Authors' Contributions

J.K. and V.Y. conceived the idea of treatment. All data acquisition was done by A.J.D., N.R., and A.A. Drafting was done by A.J.D. and N.R. All the authors read and approved the final version of the manuscript. *Being a novel case, all the authors have contributed in the decision-making, treatment, and drafting of the report.

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None.

Conflict of Interest

None declared.

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