ORIGINAL RESEARCH PAPER

Neurosurgery

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A RARE PRESENTATION OF PRIMARY EWINGS SARCOMA OF SKULL

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INTRODUCTION: Ewing's sarcomas are the second most common bone tumors in children and primary involvement of the cranium is uncommon with less than 3% cases reported worldwide. Most of the times, this condition has a fatal outcome, although the prognosis of Ewing's sarcoma is improving with radiotherapy and chemotherapy following surgery. We present a case of primary ewings sarcoma of the cranium in a 9 year old child seen in our hospital.

CASE PRESENTATION: A 9-year-old female patient was admitted with complaints of swelling in the right side of scalp, above the ear for duration of 2 months. The mass was localized under the left temporal muscle and attached to the surface of the temporal bone. A physical examination showed a firm, unmovable, mildly tender, and slightly elastic subcutaneous mass measuring 7 × 5 cm in size in the temporal region with normal overlying skin. No cervical lymph node swelling.

INVESTIGATIONS: General, systemic, and neurological statuses were normal, as well as her blood examination results. C.T revealed a mass with bony spicule formation on the temporal bone, however, it did not show bone destruction or intracranial invasion (Figure 1)

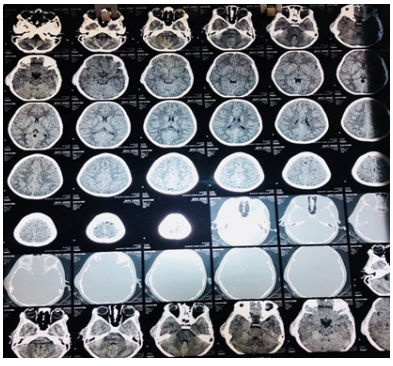

FIGURE 1

MRI of the brain revealed a right temporal extra cranial lesion. The lesion was hypointense in T1-weighted image (T1WI) with a strong heterogeneous contrast enhancement and of mixed intensity in T2WI. The lesion was associated with increased vascularity and drainage as noted in MRA/MRV. (FIGURE 2,3)

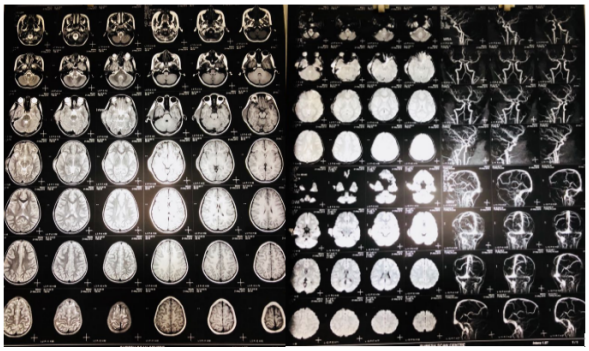

FIGURE 2 **FIGURE 3**

DIFFERENTIAL DIAGNOSIS

Eosinophilic granuloma, pleomorphic undifferentiated sarcoma.

TREATMENT

A temperoparietal craniotomy and gross total removal of the tumour with duroplasty was performed. The overlying scalp was found to be free, the bone was eroded and the tumour was found to be mostly extradural.(figure 4).

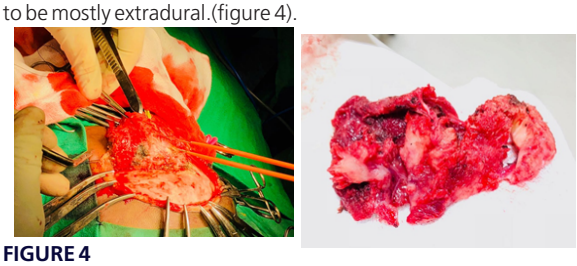

FIGURE 4

OUTCOME AND FOLLOW UP:

The patient underwent a gross total excision of tumour and HPE was reported as Ewings sarcoma. Post excision chemoradiation was given and patient is having a symptom free 1 year follow-up.

HISTOPATHOLOGY

Report Type	Final Report
Test	Histopathological Examination
Clinical Data	Right temporal extra cranial SOL
Specimen	Excision biopsy.
Gross Pathology	Specimen consists of single irregular greybrown soft tissue mass measure 7x5x3cms. Cutsection is solid,greywhite, brownish , brownish and focal gritty areas. P/E- 3Blocks-ABC, R1(Futher bits)-Decal
Microscopy	ABC, R1-Sections show woven bone infiltrated by a tumour composed of nests and sheets of small round cells having vesicular nuclei and very scanty cytoplasm. Mitotic rate 80/10 HPF. Surrounding skeletal muscle is also infiltrated by the tumour. **Immunohistochemical study:** CD99- Positive in all the cells Synaptophysin- Weakly positive in all the cells. CD45, Myogenin-Negative (Antibody used: Path in situ, US, Staining kit & Instrumentation: Ventana, Roche)
Impression	ABC, R1- Ewing's sarcoma (ES/ PNET)

DISCUSSION:

Ewing's sarcoma, a small round cell tumour that was first described by James Ewing in 1921 and is most commonly diagnosed in the second decade of life. Although it is the second most common form of primary bone cancer in childhood, the primary involvement of the calvaria is rare. Ewing's sarcoma typically grows extradurally and often reaches a very large size before dural invasion or clinical detection, or both.

Symptoms tend to develop as a result of dural invasion, hydrocephalus or raised intracranial pressure. Headaches and scalp swelling are the most common symptoms, and papilloedema is the most common sign. The duration of symptoms before presentation ranges from 2 weeks to 2 years. Men are affected more than women by a ratio of 1.8 : 1.Approximately 90% of cases occur in the first two decades of life with the peak incidence being between 5 and 13 years.

Ewing's sarcoma can be considered as an undifferentiated form of peripheral primitive neuroectodermal tumour.

Histologically, these tumours are characterised by sheets of small round blue cells with an increased nucleus-to-cytoplasm ratio. Pseudorosettes may be present, but sheets of cells are more characteristic. Mitoses are common. Bony spicules may be present, and CD99 and vimentin may be expressed.

Plain radiographs of the skull may reveal layering of bone in an 'onion peel' arrangement, with layers of bone mottling and erosion, as well as new bone formation. This distinctive periosteal reaction and calcification may also be noted on CT. In some cases, the tumour will simply be manifested as a lytic lesion on plain radiographs and CT. Bone healing after chemotherapy is better demonstrated in CT scans. MRI may show heterogeneous signal characteristics and avid contrast enhancement of any associated soft tissue component. Ewing's sarcoma exhibits increased radioisotope uptake in nuclear bone scanning images suggesting an ossification process. Scintigraphy is particularly helpful in detecting the presence of any extracranial lesions.

Surgical resection plays an important role in the management of cranial Ewing's sarcoma. Excision of the tumour should be as radical as possible to minimise tumour mass and increase the effectiveness of adjuvant therapy. The dura should be inspected for tumour infiltration, and if infiltration is noted, the dura should be resected as well. Local recurrence after resection has been reported. We did a gross total resection of the tumour.

Adjuvant therapy after resection, including radiotherapy and chemotherapy, is essential. The recommended radiotherapeutic method is supervoltage radiation, because it is better tolerated and causes less destruction to normal tissue compared with other forms of radiotherapy.Adjunctive chemotherapy with a combination of vincristine, cyclophosphamide, cisplatin, etoposide, dactinomycin and doxorubicin has raised the overall 5-year survival rate from 5–10% to 50–60%.

Various factors indicate a good outcome for patients with cranial Ewing's sarcoma: duration of symptoms for a period of longer than 6 months; absence of fever or systemic symptoms; peripheral localisation of the tumour and absence of metastases; initial lactate dehydrogenase levels of <170 IU/l; leucocyte count of <7000/dl; and lymphocyte count of <2000/dl).

Recurrence of Ewing's sarcoma of bone in general is most common within 2 years of initial diagnosis (approximately 80%). Higher rates of local failure are seen in patients older than 14 years who have tumours more than 8 cm in length.

Time to recurrence has been considered the most important prognostic factor.

Although improved by treatment regimens, the prognosis for many patients with Ewing's sarcoma continues to be poorbecause of early metastasis to the lungs and to other bones. This early metastasis is less common in cases of primary Ewing's sarcoma, and thus primary Ewing's skull tumours are considered to carry a better prognosis.

CONCLUSION:

The treatment of primary Ewing's sarcoma of the cranium still remains to be radical surgery, aggressive multidrug chemotherapy, and radiotherapy. The outcome is usually good if there is no early recurrence or metastasis.

REFERENCES

1 Agrawal A, Dulani R, Mahadevan A, et al. Primary Ewing's sarcoma of the frontal bone with intracranial extension. J Cancer Res Ther (Case Rep) 2009;5:208–9.
2 Bhatoe HS, Deshpande GU. Primary cranial Ewing's sarcoma. Br J Neurosurg (Case Rep Rev) 1998;12:165–9.
3 Kobayashi H, Terasaka S, Yamaguchi S, et al. Primary Ewing's sarcoma: peripheral primitive neuroectodermal tumour of the jugular foramen. Acta Neurochir (Case Reports) 2008;150:817–21.
4 Pfeiffer J, Boedeker CC, Ridder GJ. Primary Ewing sarcoma of the petrous temporal bone: an exceptional cause of facial palsy and deafness in a nursling. Head Neck (Case Rep) 2006;28:955–9.
5 Garg A, Ahmad FU, Suri A, et al. Primary Ewing's sarcoma of the occipital bone presenting as hydrocephalus and blindness. Pediatr Neurosurg (Case Rep) 2007;43:170–3.
6 Desai KI, Nadkarni TD, Goel A, et al. Primary Ewing's sarcoma of the cranium. Neurosurgery (Review) 2000;46:62–8; discussion 8–9.
7 Li WY, Brock P, Saunders DE. Imaging characteristics of primary cranial Ewing sarcoma. Pediatr Radiol (Case Reports) 2005;35:612–18.
8 Balasubramaniam S, Nadkarni T, Menon R, et al. Primary Ewing's sarcoma of the petroclival bone. J Clin Neurosci (Case Rep) 008;15:712–14.
9 Desai K, Goel A, Nadkarni TD. Primary petrous bone Ewing's sarcoma. Br J Neurosurg (Case Reports Review) 2000;14:143–5.
10 Erol FS, Ozveren MF, Ozercan IH, et al. Primary Ewing's sarcoma of the occipital bone—case report. Neurol Med Chir (Case Rep) 2001;41:206–9.
11 Salunke PS, Gupta K, Malik V, et al. Primary Ewing's sarcoma of cranial bones: analysis of ten patients. Acta Neurochir (Case Rep) 2011;153:1477–85.
12 Carlotti CG Jr, Drake JM, Hladky JP, et al. Primary Ewing's sarcoma of the skull in children. Utility of molecular diagnostics, surgery and adjuvant therapies. Pediatr Neurosurg (Case Rep) 1999;31:307–15.
13 Yasuda T, Inagaki T, Yamanouchi Y, et al. A case of primary Ewing's sarcoma of the occipital bone presenting with obstructive hydrocephalus. Childs Nerv Syst (Case Rep Comp Study Rev) 2003;19:792–9.
14 Mishra HB, Haran RP, Joseph T, et al. Primary Ewing's sarcoma of the skull. A report of two cases. Br J Neurosurg (Case Rep) 1993;7:683–6.
15 Kuzeyli K, Akturk F, Reis A, et al. Primary Ewing's sarcoma of the temporal bone with intracranial, extracranial and intraorbital extension. Case report. Neurosurg Rev (Case Rep) 1997;20:132–4.
16 Sharma RR, Netalkar A, Lad SD. Primary Ewing's sarcoma of the greater wing of the sphenoid bone. Br J Neurosurg (Case Rep Rev) 2000;14:53–6.
17 Varan A, Caner H, Saglam S, et al. Primary Ewing's sarcoma of the sphenoid bone: a rare presentation. Pediatr Radiol (Case Rep) 1998;28:311.