ISSN : 2949 - 9100

International Journal of Health and Dentistry Care (IJHDC)

IJHDC : Health and Dental Publication
Available Online at: www.ijhdc.com
Volume – 4, Issue – 2, March – April – 2025, Page No. : 14 – 17

A Primary Evolution in Ewing's Sarcoma of Squamous Temporal Bone with Intracranial Extension: A Case Report

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Citation This Article: Soundest J D, Zerver A K, "A Primary Evolution in Ewing's Sarcoma of Squamous Temporal Bone with Intracranial Extension: A Case Report", IJHDC – March – April – 2025, Volume. – 4, Issue – 2, P. No. 14 – 17.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Ewing's sarcoma is a rare and aggressive malignant bone tumor that primarily affects children and young adults. While it commonly arises in the long bones, its occurrence in the temporal bone is extremely rare. We present a case of primary Ewing's sarcoma of the squamous temporal bone with intracranial extension, a highly unusual presentation that poses significant diagnostic and therapeutic challenges.

Keywords: Bones, Cranial Fossa, Histopathological MRI.

Introduction

Ewing's sarcoma (ES) is a malignant round cell tumor that originates from the bones, primarily affecting children and adolescents. It constitutes approximately 3% of all childhood malignancies. The occurrence of ES in the cranium is exceedingly rare, comprising only 1% of cases. When it does occur, it typically involves the frontal, parietal, and ethmoid bones. It is extremely rare for ES to affect the temporal bone of the skull. Given the unusual location and intracranial extension, we present a case of primary Ewing's sarcoma of the squamous temporal bone.

Case Report

A 25-year-old male patient presented with pain and swelling in the right periorbital and right temporal regions for 15 days, accompanied by drooping of the right eyelid for the past 10 days. Clinical examination revealed complete third nerve palsy on the right side. Magnetic Resonance Imaging (MRI) scan showed an extra-axial mass that is T2 hypointense and T1 hyperintense, measuring approximately 43x41x45 mm. It involves the squamous part of the right temporal bone and extends into the scalp region.

FDG PET/CT post pterional craniotomy findings indicate a diffuse FDG avid large ill-defined soft tissue

density lesion in the right infratemporal fossa. This lesion involves the right frontal and temporal lobes, the right eye causing proptosis, sphenoid sinus, right temporalis muscle, and erodes the zygomatic process as well as the greater and lesser wings of the sphenoid. The findings were consistent with a primary neoplastic etiology, with no evidence of distant metastasis.

Operative findings indicated that the lesion was extradural, reddish-grey in coloration, fleshy in consistency, suckable, highly vascular, with a well-defined plane of cleavage.

Histopathological examination identified a tumour composed of cells arranged in diffuse sheets. The tumour cells had round to oval nuclei with finely dispersed chromatin and scant cytoplasm. Rosette formation was observed. The bony fragments exhibited bony trabeculae infiltrated by tumour cells, and adjacent skeletal muscle fibres were also present.

Immunohistochemistry demonstrated that the tumour cells were positive for Vimentin, CD99, and FLI1, while CD34 positivity was restricted to blood vessels. The tumour cells were negative for Chromogranin, PanCK, STAT6, EMA, S100, and MyoD1.

A final histopathological diagnosis of ES was made.

The patient received seven cycles of chemotherapy followed by radiotherapy. A clinical examination on completion of treatment showed the disappearance of lesion in the right temporal region. The patient is recurrence free at 2 years on follow-up.

Discussion

ES of the temporal bone is a rare condition. Primary ES in the skull is reported to have a better prognosis compared to other locations. These tumors respect dural planes, often presenting with large lesions that extend intracranially or cause significant neurological deficits. Previous case reports primarily identify the petrous part as the site of origin, with only a few instances documented from the squamous part of the temporal bone as documented in the above case.

The index case in this study had a large swelling in the right periorbital and right temporal region.

A multimodality imaging approach is employed for the detection and follow-up of Ewing's Sarcoma (ES). Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) are essential for forming a differential diagnosis, determining the extent of the disease, planning treatment, and managing long-term care.

MRI is a highly effective modality for evaluating local staging of masses, especially in assessing focal extension and involvement of adjacent structures. In 96% of cases, Ewing Sarcoma (ES) is associated with a large soft-tissue mass that is often circumferential but asymmetric around the osseous involvement.

The differential diagnosis for a large, progressively increasing mass includes embryonal rhabdomyosarcoma, lymphoma, or metastatic neuroblastoma.

The diagnosis of ES is confirmed through histopathological examination of a biopsy specimen and immunohistochemistry (IHC) study. ES belongs to the small round cell tumor group and is identified by small cells arranged in sheets, featuring a high nuclear-to-cytoplasmic ratio and minimal cytoplasm.

Additional microscopic characteristics include round nuclei with finely dispersed chromatin and single or multiple small nucleoli. The cytoplasmic membrane of Ewing sarcoma (ES) demonstrates CD99 expression on immunohistochemistry (IHC). Based on the extent of neuroectodermal differentiation, tumour cells may also exhibit neuron-specific enolase, synaptophysin, and S-100 protein expression. Anti-FLI1 antibody is found in

the cell nucleus and is specific to Ewing's family tumors.

The radiological findings in the index case suggested an aggressive meningioma, which was excluded based on tumour morphology and immunohistochemical analysis.

After a patient is diagnosed with ES, it is crucial to identify any metastatic lesions, as these will influence treatment and prognosis. Most ES cases initially have subclinical micrometastases that will become evident without systemic treatment.

The metastasis evaluation encompasses 18F-FDG-PET/CT and/or whole-body MRI to identify skeletal metastases, as well as chest CT for staging pulmonary metastases.

In this case, the patient underwent FDG-PET-CT, and no significant metastatic deposits were observed in the body.

The treatment options for Ewing sarcoma include surgical resection, chemotherapy, and radiotherapy. Due to the rarity of skull base Ewing sarcoma, there are currently no established management guidelines.

Research indicates that combining surgery with chemotherapy results in better 5-year survival rates and lower local recurrence rates compared to the combination of radiotherapy and chemotherapy.

Conclusion

In this case report highlights the rare but aggressive nature of Ewing's sarcoma of the temporal bone, emphasizing the need for prompt recognition and multidisciplinary treatment. Early diagnosis is crucial due to its proximity to critical neurovascular structures, helping prevent severe complications like cranial nerve involvement and intracranial extension.

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Legend Figures

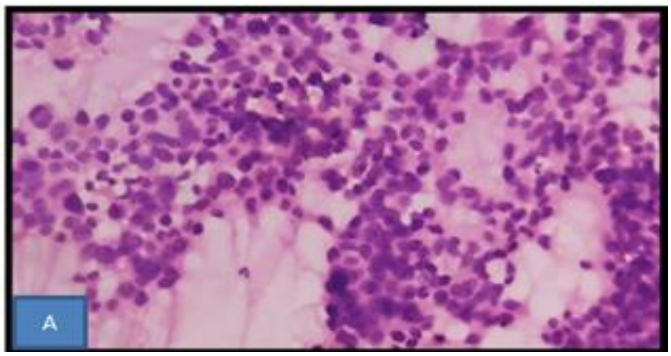

Figure 1 A: Squash cytology revealed monomorphic small round blue cells.

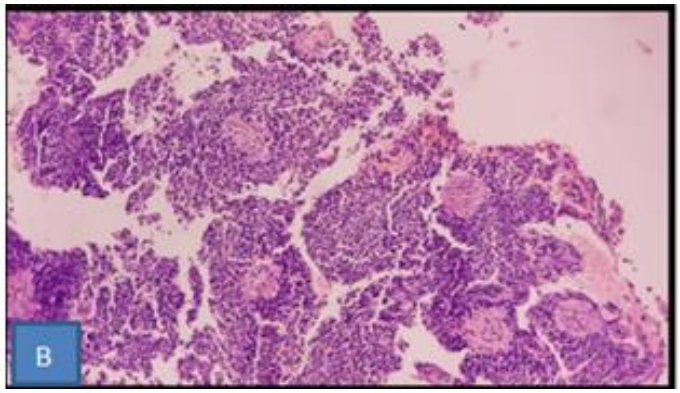

Figure 1 B: (Haematoxylin & Eosin – 100X) – displays a tumour composed of diffuse sheets of small round cells.

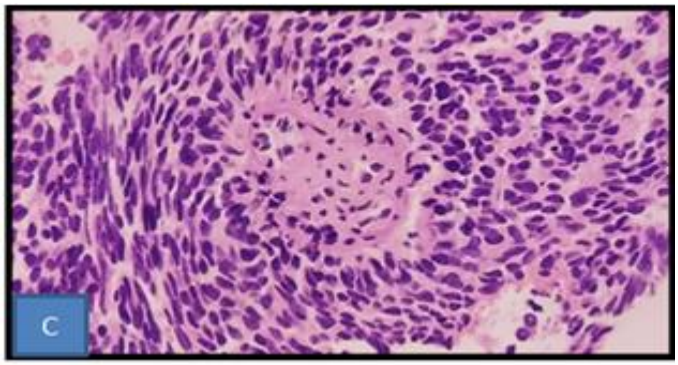

Figure 1 C: (Haematoxylin & Eosin – 400X) shows rosette formation.

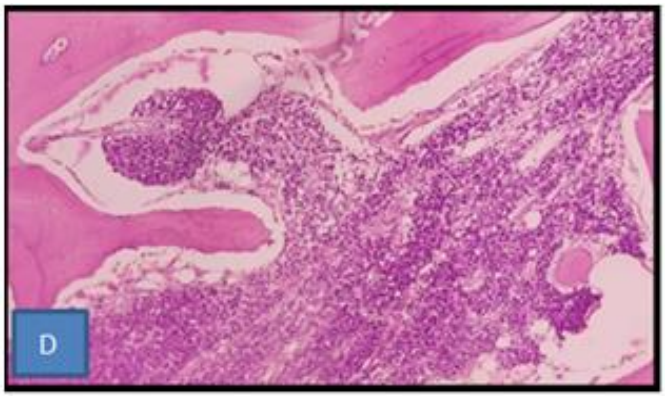

Figure 1 D: (Haematoxylin & Eosin – 100X) shows infiltration of adjacent bone by tumour cells.