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Case Report

Primary Ewing's sarcoma of the temporal bone: a rare oncological enigma

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ABSTRACT

Ewing's sarcoma is an uncommon neuroectodermal malignancy that rarely develops in the temporal bone. The early clinical recognition and definitive management of this condition pose unique challenges. This report presents a case of a 4-year-old girl with right ear pain, right post-aural swelling and progressively increasing right-sided facial nerve palsy for the past 6 months. High-resolution computed tomography (HRCT) temporal bone revealed an ill-defined soft tissue lytic lesion epi centred in the right mastoid extending to the external auditory canal and intracranially. Microscopic sections revealed round blue cells which were strongly positive for CD-99. The diagnosis of primary Ewing's sarcoma of the temporal bone was made. Considering the intracranial extension of the tumour, chemotherapy as the first line of treatment was initiated and 9 cycles have been administered resulting in marked regression of the tumour. A range of nonspecific symptoms, such as otalgia, otorrhea, sensorineural hearing loss, post aural mass, and facial nerve palsy, are indicative of the clinical manifestation of primary Ewing's sarcoma in the temporal bone. Because of how subtle these symptoms are, more clinical knowledge is required, especially in light of the small number of documented cases. When children and adolescents present with similar symptoms, clinicians should rule out Ewing's sarcoma even though it is a rare entity.

Keywords: Neuroectodermal malignancy, Chemotherapy, Facial nerve palsy

INTRODUCTION

Neuroectodermal in origin, primary Ewings sarcoma is a small round cell tumor.[1] It is the second most common malignant bone tumour in young adults. The majority of Ewing's sarcoma are diagnosed in the second decade of life, but about 20-30 percent of cases are diagnosed in the first decade.[2] Among the paediatric population, Ewing's sarcoma accounts for 6-9% of the malignant neoplasms. In 1-6% of Ewing's sarcoma cases, this malignant tumour affects the skull.[3,4] It is extremely rare in the temporal bone of the skull.

To date, 43 cases in paediatric and adolescent age groups have been described in the literature.[5] The main components of the diagnosis are immunohistochemistry and histopathological analysis.

Owing to its clinical rarity, this report describes a case of primary Ewing's sarcoma of the temporal bone in a 4-year-old girl.

CASE REPORT

A 4-year-old girl presented to the outpatient department with right ear pain, right post-aural swelling and

progressively increasing right-sided facial nerve palsy for the past 6 months.

Physical examination revealed a right diffuse mastoid swelling which was 2×2 cm in size, hard in consistency, ill-defined, non–tender and not associated with skin changes (Figure 1).

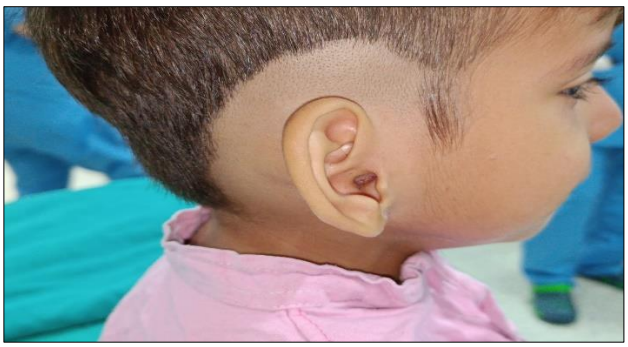

Figure 1: Clinical photograph showing right mastoid swelling and a soft fleshy mass in the external auditory canal.

Examination of the external auditory canal revealed a smooth mass, which on probing was non–tender, and did not bleed on touch. It was originating from the roof of the external auditory canal (Figure 1). The patient also had a facial nerve (House-Brackmann grade 3) palsy which on history was found to be progressing over 6 months (Figure 2).

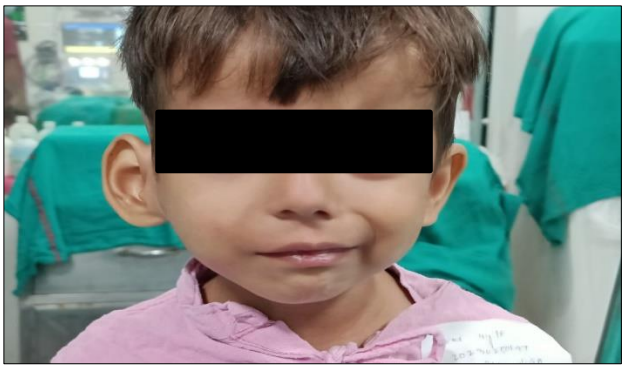

Figure 2: Clinical photograph showing right-sided facial nerve palsy (House-Brackmann grade 3).

A high resonance computed tomography (HRCT) of temporal bone revealed a large ill-defined soft tissue lytic lesion epi-centred in the right mastoid causing its destruction along with extension into the right external auditory canal causing its attenuation. The lesion can also be seen extending to the right middle ear including the mastoid antrum, epitympanum, mesotympanum and hypotympanum medially along with intracranial (extra-dural) extension along the right temporal lobe superiorly (Figure 3). A positron emission tomography (PET) scan was done which ruled out any other site of involvement or metastasis.

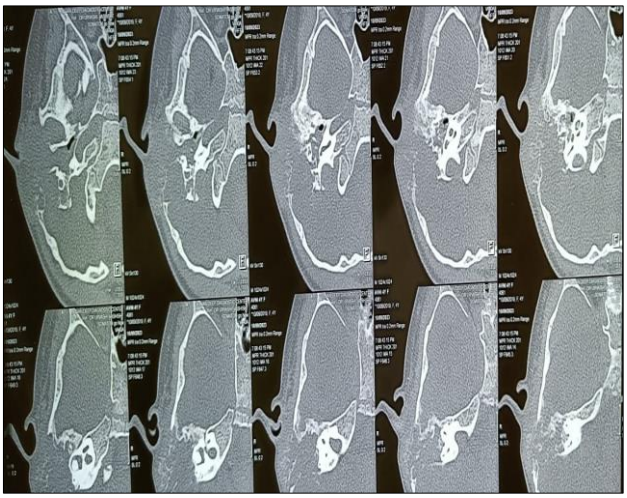

Figure 3: Large ill-defined soft tissue lytic lesion epi-centered in the right mastoid causing its destruction and also extending into the right external auditory canal causing attenuation of the right external auditory canal.

Histopathological examination of the external auditory canal mass revealed sheets of small, uniform round blue cells with scant cytoplasm, round nuclei, and small punctate nucleoli consistent with Ewing's sarcoma (Figure 4).

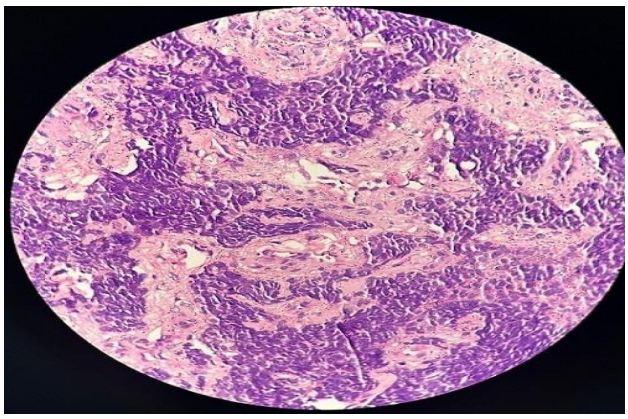

Figure 4: 400x magnification: Haematoxylin and Eosin staining sheets of small, uniform round blue cells with scant cytoplasm, round nuclei, and small punctate nucleoli.

On immunohistochemistry, it was strongly positive for CD99 but was negative for CD1a, myogenin, LCA, and desmin. This confirmed the diagnosis of primary Ewing's sarcoma of the temporal bone (Figure 5).

Considering the intracranial extension of the tumour, the patient was planned for chemotherapy and has received 9 cycles, consisting of Ifosfamide and Etoposide (IE) alternating with vincristine, Adriamycin, and cyclophosphamide (VAC) regimen. The patient has been on follow-up for 6 months and is doing well.

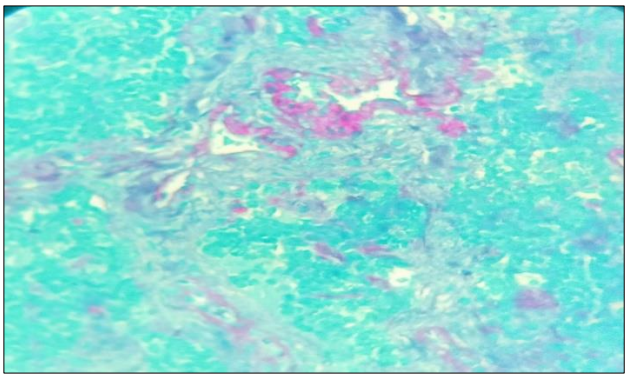

Figure 5: Immuno-histochemistry stain showing positivity for CD-99.

DISCUSSION

A rare and malignant tumour with an unfavourable prognosis, Ewing's sarcoma of the temporal bone is characterized by aggressive behaviour. It is extremely uncommon to occur in the temporal bone, accounting for only 1–6% of cases of Ewing's sarcoma. The disease predominantly affects paediatric and adolescent populations, with an equal predisposition in males and females.[6]

The second decade of life is typically when Ewing's sarcoma first manifests itself. Its cytogenetically defined reciprocal translocation (t11;22) (q24; q12) yields the EWS-FLI1 fusion gene.[7] It is derived from the primitive neuroectodermal tissue. Molecular diagnostics use this mutation as a target because it plays a crucial role in the tumorigenesis of Ewing's sarcoma. Since the symptoms of Ewing's sarcoma often resemble those of other illnesses, a misdiagnosis may occur when the tumour affects the temporal bone. In addition to otalgia and potential facial nerve paralysis if the tumour impinges on the facial nerve, resulting in facial droop, other possible symptoms include pain and swelling in the affected area, auditory symptoms such as hearing loss, which could be conductive or sensorineural, and discharge from the ear if the tumour causes erosion into the ear canal or middle ear.

Ewing's sarcoma of the temporal bone is diagnosed by a biopsy along with imaging tests. Computed tomography (CT) and magnetic resonance imaging (MRI) may be used as preliminary imaging tests to assess the tumour's extent and damage to surrounding structures. Definitive diagnosis of Ewing's sarcoma is based on, a tumour biopsy, histological examination, and immune-histochemistry to confirm the presence of the small, round, blue cells that are indicative of the tumour. The distinctive EWS-FLI1 fusion gene can be identified by molecular genetic testing.

The treatment protocol for Ewing's sarcoma of the temporal bone generally involves multimodal strategies which include neo-adjuvant chemotherapy, surgical resection, and adjuvant therapy.[8,9]

Initial systemic chemotherapy is essential to decrease tumour size and control micro-metastatic disease. Common regimens include combinations of vincristine, doxorubicin, cyclophosphamide, ifosfamide, and etoposide.[10]

Complete surgical excision with negative margins is challenging due to anatomical constraints, but it is necessary. A surgical strategy that balances oncological control with the preservation of neurological and auditory function is required. When surgical resection presents a significant risk of morbidity or when the surgical margins are positive, post-operative radiation therapy may be warranted. Important considerations for continuing chemotherapy include the initial response and the postoperative histopathological results.

When considering the other sites, the prognosis for temporal bone Ewing's sarcoma is typically worse because of the difficulty in accomplishing total surgical resection and the increased risk of local recurrence. However, recent advancements in multimodal therapy and early detection have improved the results.

CONCLUSION

In conclusion, temporal bone Ewing's sarcoma is an uncommon and aggressive entity that needs a high suspicion index to be detected promptly. To achieve optimal clinical outcomes, a multidisciplinary approach involving pathologists, otolaryngologists, radiologists, and oncologists is necessary for optimal management.

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