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Ewing sarcoma and peripheral primitive neuroectodermal tumor of middle: External ear: A rare case report and review of literature

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Abstract
Ewing sarcoma family of tumors (EFTs) of the head and neck are rare and account for only 1 – 4% of all EFTs. Here, we describe an extremely rare case of Ewing's sarcoma/PNET arising from middle-external ear in a 32-year old female, who presented with growth from external auditory canal. As per my knowledge, this is the first case reported in India. This patient is taken on EFT 2001 protocol and is doing well. Aggressive multimodality treatment including chemotherapy and surgery/radiation can improve the survival of these patients.

Keywords: Ewing's sarcoma, extra osseous Ewing's sarcoma, PNET, Peripheral primitive neuroectodermal tumor, middle-external ear carcinoma

Introduction
The Ewing sarcoma family tumors (ESFT) and peripheral primitive neuroectodermal tumor (pPNET) represent different manifestations of the same entity. Immunohistochemical and cytogenetic studies suggest that these tumors have a common origin. Ewing sarcoma is more common in bone, while pPNET is more common in soft tissues. Extraosseous Ewing sarcoma (EoES) involving head and neck region is rare and involvement of middle-external ear is extremely rare. Here, we present a case of a 32-year female presented with growth from left ear. The mass was later diagnosed as Ewing's sarcoma/PNET upon tissue biopsy and immunohistochemistry. The patient is on EFT 2001 protocol and doing very well.

Case Report
32-year-old non-diabetic, normotensive lady presented to Mahavir Cancer Sansthan, Patna on 06th March 2018 with complain of gradually developing growth from left ear for last more than 6 months. There was no history of any trauma, pain or fever. No any relevant past history. No history of any addiction. Regular menstrual history. On physical examination, there was a painless, smooth, firm to hard mass coming from the left external auditory canal. CT Scan face and neck (Fig: - 1) revealed 52 x 39 mm size ill-defined heterogeneously enhancing lesion seen involving left middle and external ear with prominent exophytic component with involvement of pre- post auricular region, left TM joint, proximal sternocleidomastoid muscle and adjacent parotid gland; a large peripherally enhancing collection measuring 52 x 21 mm in the subcutaneous plane of supra auricular region.There is diffuse thickening and enhancement of pinna and complete opacification of left mastoid air cells. A few prominent enhancing left intra-parotid and left upper cervical lymph nodes noted, largest measuring 15 x 12 mm in left upper cervical region.

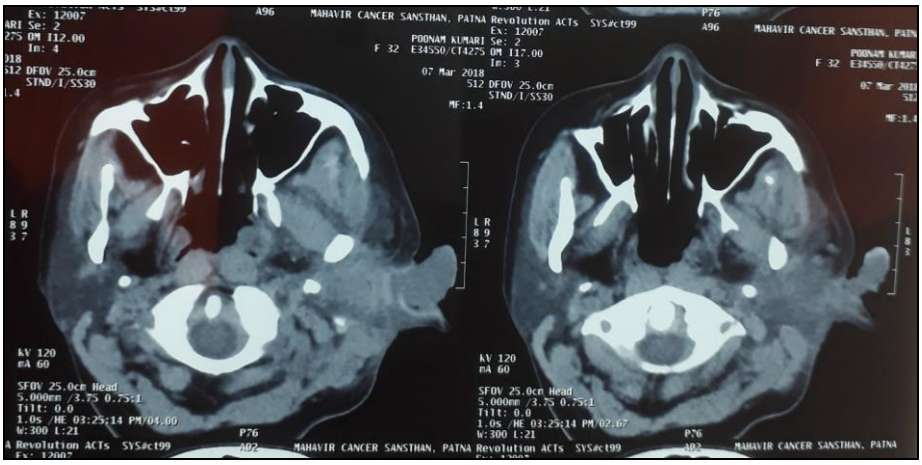

Fig 1: CT scan showing lesion involving left middle-external ear

Incision biopsy from left external auditory canal growth section shows small nests as well as scattered small round

cells having round hyperchromatic nuclei with scanty cytoplasm lying in myxoid stroma with prominent blood vessels, suggestive of round cell tumor (Fig: - 2 A-C). On further, immunohistochemistry examination the tumor cells show membranous expression of Mic-2, weak expression of EMA and focal and weak expression of Cytokeratin. The tumor does not express Synaptophysin, Desmin & LCA. The overall feature confirms the diagnosis of Ewing's sarcoma/Peripheral neuroectodermal tumor (PNET) (Fig: - 3 D-H).

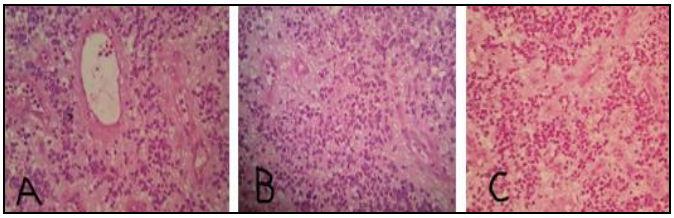

Fig 2: (A-C): - Section showing small nests as well as scattered small round cells having round hyperchromatic nuclei with scanty cytoplasm lying in myxoid stroma with prominent blood vessels. Suggestive of Round cell tumors.

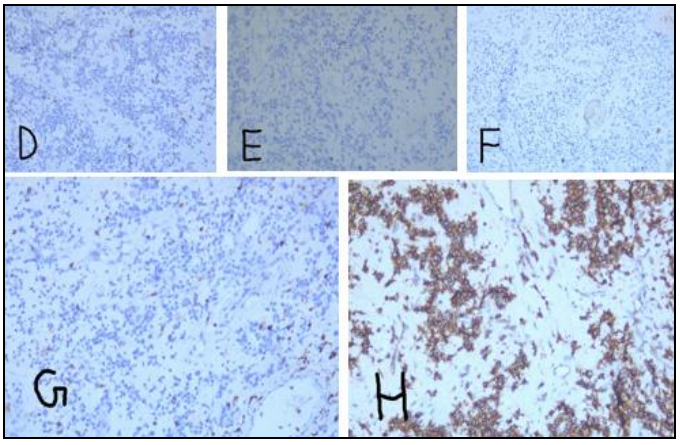

Fig 3: (D-H): - Immunohistochemistry examination of the tumor cells showing weak expression of Cytokeratin (D – ck 20x), not express Desmin (E – desmin 20x), weak expression of EMA (F – EMA 20x), not express LCA (G – lca 20x) and showing membranous expression of Mic-2 (H – mic-2 20x).

Discussion

Ewing sarcoma (ES) is a highly malignant tumor composed of small round cells. They are the second most common malignant tumor in children after osteosarcoma. Most cases arise in the long bones of the limbs or the pelvis. Primary Ewing's sarcoma arising from the bones of the head and neck region is extremely rare representing only 1 – 4% of all Ewing's sarcoma cases [1].

The most commonly affected bones in the head and neck region are the skull [2], temporal bone [3] and the maxilla [4]. There have also been case reports of localized Ewing's sarcoma affecting the mandible [5], orbital roof [6], the retropharynx [7]. Ewing's sarcoma/PNET of middle-external ear is extremely rare. Only one case "Ewing's sarcoma of the external ear canal" is reported in English literature [8].

Its clinical and radiological features are non-specific, and a histological diagnosis is essential to confirm the diagnosis. Our patient presented with a growth coming from the external auditory canal. CT scan was suggestive of neoplastic etiology involving the left middle-external ear although infectious etiology like malignant otitis externa was other possibility. Histopathologic examination of specimen from external auditory canal growth reveals round cell tumor and on further Immunohistochemical examination Ewing sarcoma/peripheral primitive neuroectodemal tumor is confirmed.

Multidisciplinary care is indispensable for these patients. When localized, this is treated by surgical excision with adjuvant or neoadjuvant chemotherapy. Recent studies have shown that neoadjuvant and adjuvant chemotherapies produce comparable results in patients with localized disease [9]. EES is quite radiosensitive, but improvements in surgical technique and the risks associated with radiation (secondary malignancies) have reduced the reliance upon radiation [10]. Radiotherapy is used to treat localized disease which is considered inoperable, in patients with inadequate surgical margins and in those with a poor response to chemotherapy [11].

It is responsive to a number of chemotherapeutic drugs, including vincristine, doxorubicin, actinomycin D and cyclophosphamide. The VAC/IE regimen [12] and EFT-2001 protocol both are considered standard treatment for EES. We are using EFT 2001 protocol for treating this type of tumor. The patient has tolerated the neoadjuvant chemotherapy very well, the tumor size has regressed very much. At present patent is undergoing radical radiotherapy and doing very well.

The prognosis for extraosseous ES appears more favourable than that for ES in bone [13], although prognostic factors of EES seem to be similar to primary bone ES [14]. The outcome for localized extraosseous ES tumours was similar to that reported for all patients with ES treated on protocols at the St. Jude's Children Research Hospital [15]. Patients with localized EES have estimated 5-year overall survival rates of about 70 % [13]. Patients with metastatic or recurrent disease have a worse outcome; 5-year overall survival remains about 25 % [16]. ES-HN does not appear to be a separate clinical entity compared to ES-other; rather, its associated improved prognosis is likely secondary to its smaller size and lower metastatic rate compared to ES-other [17].

Conclusion

Ewing sarcoma/PNET has a wide histological spectrum and varied in location. Middle-external ear is an extremely rare location. Due to its rarity of the disease, there is no standard guideline regarding its management. Aggressive multimodality treatment including chemotherapy and surgery/ radiation can improve the survival of these patients in a tertiary centre where all the facility and expertise are available.

Conflicts of interest - None

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