

Primary Ewing sarcoma of squamous temporal bone with intracranial extension: A case report

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Abstract: Primary Cranial Ewing sarcoma (ES) is rare and that of temporal bone is even rarer entity. Only a few sporadic cases have been reported and no such case has been reported from Eastern India

In this report we describe a case of 18 yrs old male with primary cranial ES of squamous temporal bone involving intracranial and extracranial compartment. The patient presented with swelling in zygomatic fossa and imaging studies showed a mass originating in the right temporal bone. Gross total resection of the tumor was done and sent for histopathological study. The case was referred for radio-chemotherapy and had no recurrence up to eight months of follow up.

Key words: Primary Ewing sarcoma, squamous part of temporal bone, MRI, Histopathology

Introduction

Ewing sarcoma (ES), also called peripheral primitive neuroectodermal tumor (PNET) commonly originates from bone and soft tissue of extremities and pelvis [1]. The skull is involved in less than two percent of cases [2,3]. Till now only 14 such cases involving temporal bone have been reported in the literature .One such case is being reported.

Case History

A 18 year old male child presented with history of swelling in his right temporal region since one year. The swelling was increasing in size and associated with pain since last one

month. There was no preceding history of trauma and fever. Physical examination suggested a palpable mass in right temporal region of 8 x 6 cm in size. The mass was firm and fixed to underlying bone. Overlying skin was free and distended veins were seen over the swelling (Figure 1). His general as well as systemic examination and neurological status was within normal limits.

His routine haematological and biochemical investigations were normal. Ultrasonographic evaluation of abdomen and thorax were normal. Skeletal radiographs revealed no other extracranial lesion. CT scan of brain showed increased attenuating

enhancing lesion in right temporal area with involvement of inner and outer table of right squamous temporal bone with both intra and extracranial extension. MRI study of Brain revealed extra-axial mass in right temporal region with involvement of bone and extracalvarial soft tissue swelling showing marked enhancement on contrast study (Figure 2).

Plan for excision of mass was decided. A Right Fronto-Temporo-Parietal craniotomy was performed. Intraoperatively a firm tumor was noted which originated from the squamous part of temporal bone, extending extradurally under temporal lobe and involved dura. Gross total excision of tumor and involved dura was done and sent for histopathological study.

Hematoxylin & Eosin (H&E) stained sections revealed tumor mass composed of uniform small round cells with round nuclei containing fine chromatin, scanty cytoplasm and indistinct cytoplasmic membrane. Immunohistochemistry for CD 99 was positive and negative for GFAP and vimentin (Figure 3). Histopathological diagnosis was Ewing sarcoma.

The patient subsequently received induction chemotherapy with cyclophosphamide, vincristine and adriamycin alternating with ifophospamide and etoposide administered at 3 weeks. The local area was irradiated with 40–50 Gy eight weeks later. This was followed by eight cycles of chemotherapy same as induction at 3-week intervals. Patient was followed for eight months after radiochemotherapy where he developed no recurrence and later lost in follow up.



Figure 1 - Preoperative -Right temporal palpable swelling extending to frontal and parietal region

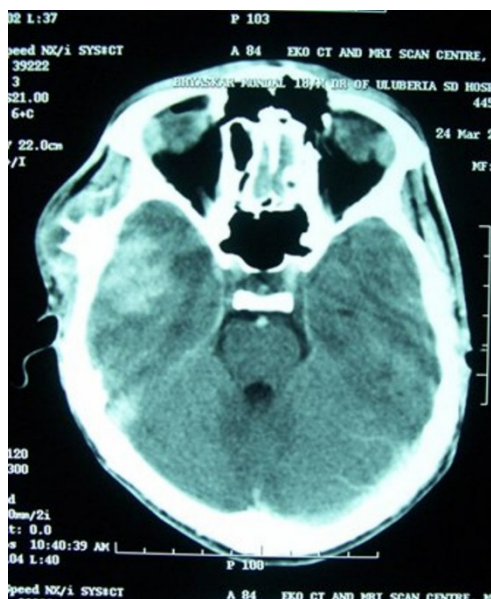


Figure 2 - Axial contrast enhanced CT images show enhancing lesion in right temporal region with involvement of right squamous temporal bone and overlying scalp tissue

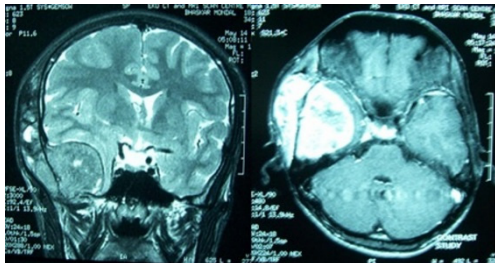


Figure 3 - T2W coronal and T1W axial contrast MRI image reveals well defined extra-parenchymal and extra calvarial lesion maintaining distinct border with brain and shows marked contrast enhancement

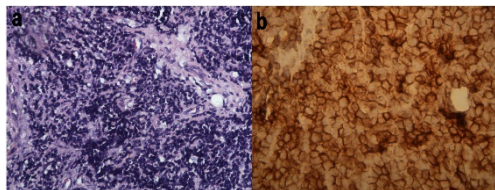


Figure 4 - a. Tumor section showing small round cells (H&E, x400). b. Tumor cells showing strong membranous staining for CD99 (immunoperoxidase, x400)

Discussion

ES is a highly malignant bone tumor arising from the pluripotent cells in bone marrow or primordial bone marrow derived mesenchymal stem cells [4,5]. The tumor is now classified under Ewing Sarcoma Family of Tumor (ESFT) which includes Classic Ewing sarcoma, PNET, Askin tumor and Extra osseous Ewing sarcoma [6]. Median age of patient with ESFT is 15 years and more than 50% are adolescent. The Tumor primarily involves lower extremity (41%), pelvis (26%), chest wall (16%) & upper extremity (9%) [7]. Common presentations are localised bone pain and swelling, local tenderness, palpable mass.

In the present case, the presenting symptom was a large swelling in the temporal

region with no evidence of symptoms of elevated intracranial pressure suggesting extracranial extension was larger part of tumor. The predominant presenting feature in other such reported cases was scalp swelling, headache and findings of raised intracranial pressure.

Involvement of cranium by ES is very rare. Only 29 cases of primary ES of cranium have been reported [8] and 14 cases pertaining to involvement of temporal bone.

CT of Ewing's sarcoma has often shown a diffusely enhancing hyperdense extra axial mass and extensive bone destruction involving both inner and outer table. The present case also had similar findings on the CT scan of brain [2,9,10,11].

Differential diagnosis of such large and progressively increasing mass can be embryonal rhabdomyosarcoma, lymphoma or metastatic neuroblastoma. ES as diagnosis was confirmed by CD 99 positivity.

Management of such tumor is radical excision and radio-chemotherapy [12]. Current standard chemotherapy includes vincristine, doxorubicin and cyclophosphamide alternating with ifosfamide and etoposide [13,14].

The prognosis for patient with ES has improved considerably with current treatment protocol. However outcome for patient with metastasis or early relapse is poor.

Though primary cranial ES of temporal bone is rare, it should be kept as differential diagnosis in cases of temporal scalp swelling. The present case is exceptional as it originates from the squamous part of temporal bone and no such tumor has been described from eastern India.

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