Case Report

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Ewing's Sarcoma Multifocal Metastases to Temporal and Occipital Bone: A Rare Presentation

Ear Nose and Throat Section

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ABSTRACT

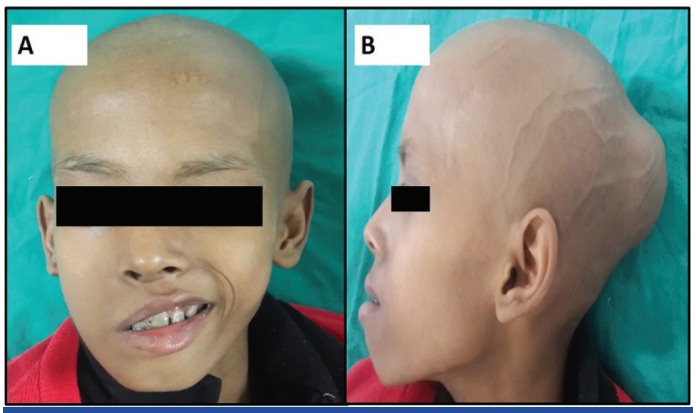
Ewing's sarcoma (ES) is a common malignant bone tumour seen to involve long bones, flat pelvic bones and ribs and vertebrae in majority of cases. Here, we present a rare case of aggressive primary ES of pelvic bones with multifocal metastases to temporal bone and occipital bone. The patient presented with facial palsy and an occipital swelling, and was referred for chemotherapy.

Keywords: Metastasis, Multiple, Skull

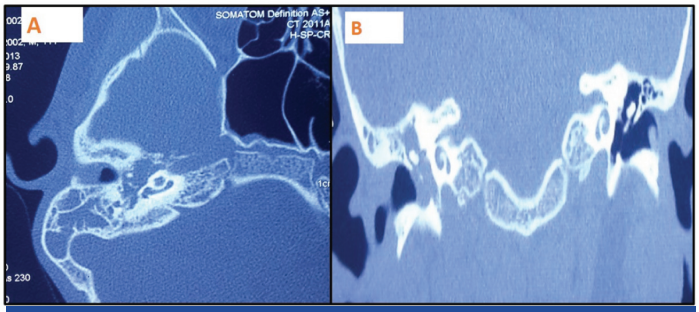
CASE REPORT

An 11-year-old male patient was referred to the ENT department at Lok Nayak Hospital with the complaints of right ear discharge, facial weakness, inability to close right eye, and a gradually progressive occipital swelling for one month. In addition, he complained of decreased hearing from right ear, as well as headache. The patient was a known case of Ewing's sarcoma of the left pelvic bones, with involvement of iliac bone, ischium and sacroiliac joint. He had presented initially with the complaints of pain in the left hip and a swelling in the groin and was started upon five cycles of multidrug chemotherapy that included ifosfamide, etoposide, vincristine, actinomycin- D and cyclophosphamide. This was followed by radiotherapy, with a total body dose of 55 Gy over 30 cycles. In view of persistent disease, patient further received 3 cycles of chemotherapy (CT) with ifosfamide, carboplatin and etoposide. It was during one of the follow up visits that patient complained of right sided ear discharge, facial weakness, and an occipital swelling, and was referred to ENT clinic for evaluation.

On examination, patient had a red polypoidal mass in the right bony external auditory canal (EAC) with some overlying sero-sanguinous discharge on the floor. The mass was found to be extending into the middle ear, with disruption of lower part of tympanic membrane. It was friable and bled on probing. He also had right sided facial weakness with history of deviation of angle of mouth towards left, suggestive of a right sided lower motor neuron (LMN) type facial nerve palsy [Table/Fig-1a]. However, patient's right eye was close at the time of examination due to conjuctival chemosis and oedema. The patient had an associated 6 x 6 cm swelling present over the occipital bone [Table/Fig-1b]. The swelling was non tender, immobile, and hard in consistency. The overlying skin appeared stretched, and there were multiple dilated scalp veins over the swelling and the adjoining area.



[Table/Fig-1a,b]: (a) Clinical photograph of the patient showing deviation of angle of mouth towards left side and no movement on right side of face, suggestive of right sided lower motor neuron type facial nerve palsy. (b) Clinical photograph of the patient showing an occipital swelling with stretched overlying skin and multiple dilated veins

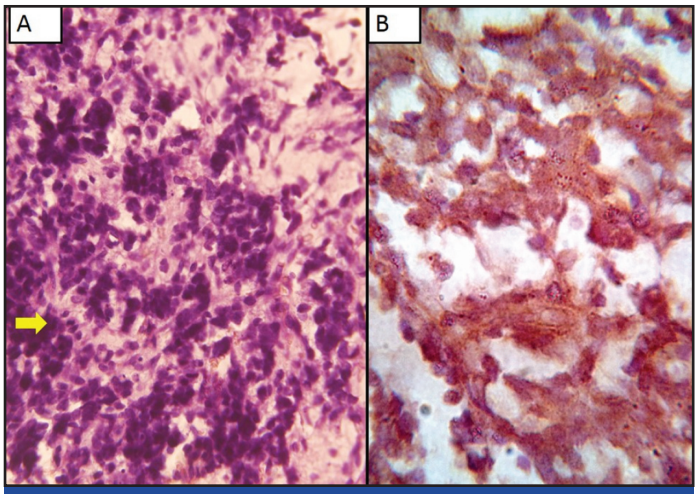


[Table/Fig-2a,b]: CECT Temporal bone showing soft tissue attenuation contents in the right mastoid air cells and middle ear, with erosion of right ear ossicles, tegmen tympani and scutum. (a) Axial section, and (b) Coronal section

The patient underwent computed tomography scan of temporal bone and head revealing soft tissue attenuation contents in the right mastoid air cells and middle ear, with erosion of right ear ossicles, tegmen tympani and scutum [Table/Fig-2a&b]. The facial canal was eroded at the genu and tympanic segment. Scans through the head revealed an isodense lesion involving occipital bone, heterogeneously enhancing after contrast administration. There was no brain involvement. The patient was taken up for biopsy of the mass which revealed fibro collagenous tissue infiltrated by small round cells, having high nucleus: cytoplasm ratio and few mitoses. The tumour cells were positive for vimentin, neuron specific enolase (NSE), and CD99, and negative for desmin and leukocyte common antigen (LCA) [Table/Fig-3]. Hence, a diagnosis of primary ES of pelvic bones with multifocal distant metastasis to temporal bone (middle ear and mastoid) and occipital bone was established. In view of the aggressive disease and poor prognosis, the patient was considered for palliative chemotherapy with vincristine, actinomycin D and cyclophosphamide, with high dose morphine for pain relief. Patient took 3 cycles of palliative CT, however, he died within 2 months due to the ongoing disease process.

DISCUSSION

Ewing's sarcoma (ES) was first described in 1921 by James Ewing as a "diffuse haemangioendothelioma of bone" [1]. It is an aggressive malignant tumour of childhood, and the second most common primary bone tumour. It can involve almost any bone in the body, however, trunk and long bones are more commonly affected [2]. In 90% cases, it is seen in patients less than 20 years of age, the highest incidence being observed in 5-13 year old patients [3]. It



[Table/Fig-3a,b]: (a) Histopathology showing diffuse arrangement of small round uniform cells with scant cytoplasm, round nucleus with small nucleoli showing mitosis (arrow) (H&E, 200X); (b) Strong membranous expression of CD99 by the tumor cells on immunohistochemistry (400X)

is associated with male predominance, the male: female ratio being 1.6: 1 [4]. It can have multicentric manifestations, and distant metastases may present as early as within 2 years of presentation of primary disease. Distant metastasis usually involves the lung (38%), bone (including the spine; 31%), and the bone marrow (11%) [5]. Metastases to skull bones is rare and is seen in approximately 9% of cases, only few cases being reported in literature till date [6]. Primary involvement of the skull bones is seen more commonly than metastasis to the skull. Skull metastases to petrous bone, cuneiform bone, parietal bone, frontal bone, and clivus associated with scalp involvement have been described in the literature [4,7-10]. Dural metastases, though rare has also been described with ES by various authors [11,12]. Since metastases to skull bones is rarely seen with ES, it can be confused with other lesions like olfactory neuroblastoma as described by Gaba et al., [13].

The clinical presentation depends on the location and spread of the tumour, with no significant difference seen in the presentation of primary and metastatic tumours. Headache, skull swelling and signs of raised intracranial pressure are usually the main symptoms. Temporal bone involvement mainly presents with facial paralysis, hearing loss and ear discharge. Diagnosis of ES mainly relies on histopathology and immunohistochemistry. Main histological features include round cells arranged in the form of solid strata, scanty cytoplasm, protruding nuclei, mitoses, and the presence of bony structures. Immunohistochemistry reveals vimentin and CD99 expression. On contrast enhanced computed tomography (CECT), the tumour enhances heterogeneously and extensive bone destruction with a moth eaten appearance can be appreciated; the classical "onion skin" sign is usually not seen in skull lesions.

The treatment of ES is multimodal, including surgery, chemotherapy and radiotherapy. Surgical intervention mainly aims at reducing the tumour bulk and shows good results for peripheral tumours. Multi-drug chemotherapy as per the round cell therapy (RCT) II includes 3 weekly cycles of ifosfamide, etoposide, vincristine, cyclophosphamide, doxorubicin and/or actinomycin D. Radiotherapy with a total body dose of 40-50 Gy is recommended [6]. A recent study by Casey et al., studied the effect of radiation in bony metastases in ES, and concluded that doses in the biologic range prescribed for the definitive treatment of primary disease should be used for metastatic sites of disease for effective control [14].

We lost our patient within 3 years of the primary pelvic tumour due to the presence of multifocal skull metastases. Hattori T et al described three patients of skull metastasis of ES [7]. One of the patients had a primary ES of distal femur, and developed multiple cranial metastasis involving frontal and parietal region within months of the surgery. He received palliative treatment for the metastasis, but died due to intracranial hypertension. The other two patients had solitary, well defined metastatic lesions, which were completely excised and postoperative chemotherapy was given. They both showed favourable treatment outcomes without any recurrence. Fatal outcomes were also seen in a study by Cherekaev et al., where three out of four patients of metastatic ES to skull base expired within 3 years of treatment with surgery and chemotherapy [6].

CONCLUSION

Despite the multimodal treatment, the outcome of ES remains complex, mainly due to its aggressive nature and the propensity for distant metastases. Primary ES of skull base has a more favourable course as compared to a metastatic tumour. Early diagnosis of the primary tumour and aggressive therapy are the key points for an overall favourable outcome.

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