CASE STUDY

EXTRASKELETAL EWING'S SARCOMA IN PEDIATRIC PATIENT RARE CASE: A CASE REPORT

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ABSTRACT

This study reports a 12-year-old male was referred to our institute with a mass in the right ear. The mass was rapidly growing thus needing an immediate surgical attention. MRI revealed an exophytic, hyper vascular mass extending to the right jugular fossa and, the middle ear, and to the right acoustic meatus, suggestive of a right glomus jugulare tumor. Angiography showed a hypervascular lesion with robust feeding from the superficial temporal artery and right occipital artery. After surgery, the patient rapidly recover and he was discharged home on the 5th day after surgery. Three months after surgery, the lump reappeared and grew rapidly as before.

KEY WORDS: Extraskeletal sarcoma, Ewing sarcoma, sarcoma, Intracranial invasion

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INTRODUCTION

Ewing sarcoma/peripheral primitive neuroectodermal tumor (ES/pPNET) is an undifferentiated malignant, small, round cell tumor which rarely occurs in the skull and meningeal tissue[1]. It accounts for 4-10% of all primary bone cancers affecting teenagers and young adults and rarely develops after age 30 years [2]. According to anatomical site of occurrence it is classified as : (a) intraosseous (most common) (b) extraskeletal (less common) and (c) periosteal (rare) type. Intracranial invasion of extraskeletal ES extremely rare [3].

Intracranial invasion of extraskeletal ES is often misdiagnosed as an atypical teratoid/rhabdoid tumor (AT/RT) or a primary leptomeningeal medulloblastoma [2]. In addition, because of the lack of clinical symptoms in the early stages of cranial ES, most patients are diagnosed in advanced stages, leading to worse outcomes [4]. In this case report we present a case of a 12-year-old male with intracranial invasion of extraskeletal ES.

CASE REPORT

A 12-year-old male was referred to our institute with a mass in the right ear for 5 months prior to admission. The progression of the mass size was rapid. He also had persistent headache and ear pain. Clinical examination revealed paresis of the 7th cranial nerve (House-Brackmann 2), bilateral papilledema and conductive hearing loss of the right ear. Motor strength, sensory and reflexes were normal.

Radiological workup with MRI revealed an exophytic, hyper vascular mass extending to the right jugular fossa and, the middle ear, and to the right acoustic meatus, suggestive of a right glomus jugulare tumor. The mass was supplied by a branch of external carotid artery.

Angiography (Fig 3.) showed a hypervascular lesion with robust feeding from the superficial temporal artery and right occipital artery. Laboratory examination was conducted, there is no abnormalities in the pre-operative lab result. Excision of the tumor was conducted, and the patient's recovery went well.

OPERATIVE MANAGEMENT

Intraoperatively, the tumor was found to have invaded the dura and the cranium. The involved bone (including the lateral sphenoid wing) and the large dural cuff were resected. The tumor was soft and hypervascular.

The patient's postoperative recovery went well and he was discharged home on the 5th day after surgery. Three months after surgery, the lump reappeared and grew rapidly.

HISTOPATHOLOGICAL EXAMINATION

Histopathological examination showed round to oval cells arranged in lobules, separated by a thin vascular

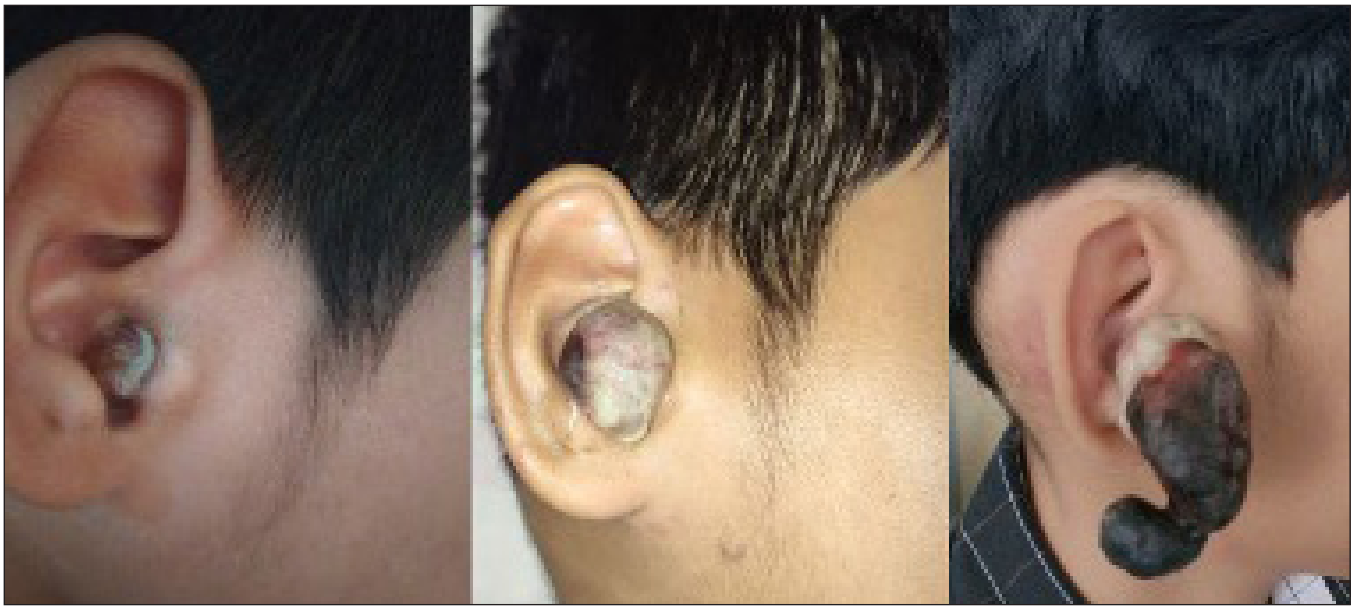

Fig. 1. The appearance of the mass at the first visit in the ENT outpatient clinic (a), 1 month after the first visit, the lump was increasingly visible from outside of the ear canal (b), two months after the first visit the lump was visibly protruding from the ear canal with necrotic parts (c)

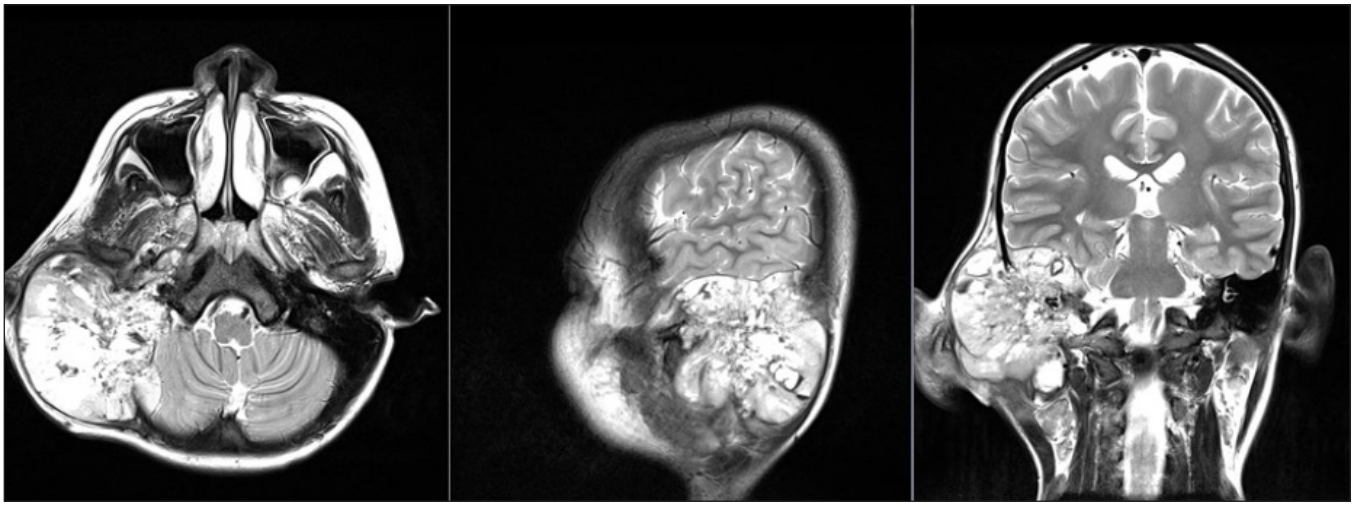

Fig. 2. MRI of the patient. T2W1 with contrast.

channel, with vesicular nuclei. Mitotic figures were observed with foci of necrosis. The immunohistochemistry positive for CD99 but negative for CD20, CD3, myosin and glial fibrin acid proteins. The overall features are consistent with a primary ectodermal neuroendocrine tumor (PNET), suggesting an Ewing sarcoma in the temporal region.

ADJUVANT TREATMENT

After the result of pathological anatomy with IHC CD99 was reported, chemoterapy immediately conducted. The chemotherapy regimen consisted of vincristine-doxorubicin-cyclophosphamide. However, before the chemotherapy regimen is finished, the clinical condition of the patient is deteriorating. The patient unfortunately passed away before receiving any radiotherapy treatment.

Cases of the central nervous system extraosseous Ewing's sarcoma (ES) are extremely rare. Only few that have been reported literature [5]. Jay might be the first to describe a patient with an isolated posterior fossa mass that histologically resembles a medulloblastoma [6].

Paulus mentioned in their study in 1991 that out of 2500 cases of brain tumor, only 9 were sarcomas, among which only one was reportes as ES [7]. Similarly, Krishnamani et al. reported only 7 cases were primary ES of the skull out of 332 cases of ES of any sites [8]. These tumors clinically present with signs of increased intracranial pressure. Motor deficit or visual disturbances were uncommon and endocrine abnormalities can occur depending on the tumor location.

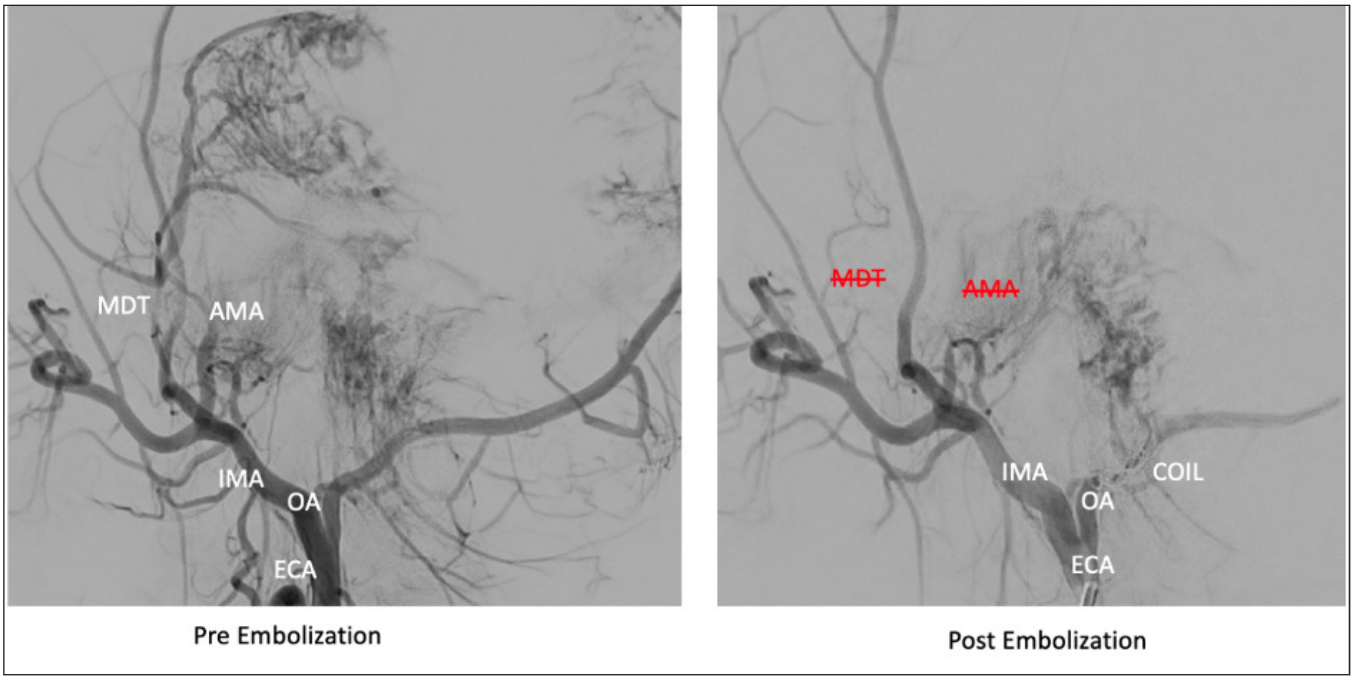

Fig. 3. Angiography study of pre-embolization and post embolization.
MDT: Middle deep temporal; AMA: Accessory Meningeal Artery; IMA: Inferior Meningeal Artery; OA: Occipital Artery; ECA: External Carotid Artery.

Diagnosis of ES requires a histopathological examination, immunohistochemistry, and cytogenetics. The differential diagnosis of this intracranial round cell tumors is primitive neuroectodermal tumor (neuroblastoma), lymphoma, rhabdomyosarcoma, and Ewing's sarcoma. Macroscopically, intracranial invasion of extraskeletal ES might mimic meningioma due to its dural base and bone-eroding manner, as presented in our case.

Radiographs may show a nonspecific soft tissue mass. Computed tomography (CT) usually shows a soft tissue mass with attenuation similar to that of skeletal muscle [9]. In our patients, head CT Scan showed a rounded, poorly-defined, heterogeneously enhancing lesion in the right temporal region, with a mass effect and destruction of the right temporal bone, extending into the scalp. Head MRI revealed a solid mass from right jugular fossa to right external acoustic meatus, which appears isointense on T1-weighted images, hyperintense with prominent flow voids on T2-weighted-images. In patient with ES, MRI imaging usually revealed a mass with signal intensity similar to that of skeletal muscle on T1-weighted imaging. On T2-weighted images, the mass often demonstrates a heterogeneous intermediate to hyperintense signal. Hyperintense areas representing foci of cystic or necrotic changes can be found. High-flow vascular channels or flow voids may also be seen which is extremely common, although not unique to ES [9].

A report by Deshpande et al., described 8 similar case with the age range of 1 to 33 years, with the median age of 9 years. Clinical manifestation in this report also varied greatly ranging from seizure, ptosis, weakness in extremity, but headache, and vomiting were the most reported complaint [10]. This is similar to our case which described a 12 year old male with solid mass at right jugular fossa which also complained of headache and showed facial palsy as neurological deficit.

Huang et al., reported a case that is similar to our case. In this report, a 19 year old patient is admitted with a complaint of headache, vomiting, and behavioural changes. Neurological examination is remarkable with an exception in right pronator drift. MRI showed a large extra-axial mass which is heterogeneously enhanced on the left frontal convexity [10].

Another report by Jiang et al, described a 55 year old female with a chief complaint of memory decline over 1 month with hemiparesis. In this case, MRI revealed a large irregular mass located in the left frontal lobe with mixed isointense-to-hypointense signals on T1-weighted imaging, heterogenous hypointense-to-hyperintense signals on T2-weighted imaging, and T2 dark-fluid [7].

The exophytic part of the tumor protruding to the outside is the most interesting point in our finding case. The presentation extra skeletal ES which protruding from intracranially to the outside to the ear canal has not been reported. It differs in presentation from other previously reported case by Huang et al., that revealed as a well-defined mass with dural or bony involvement, mimicking the appearance of a meningioma [11]. An-

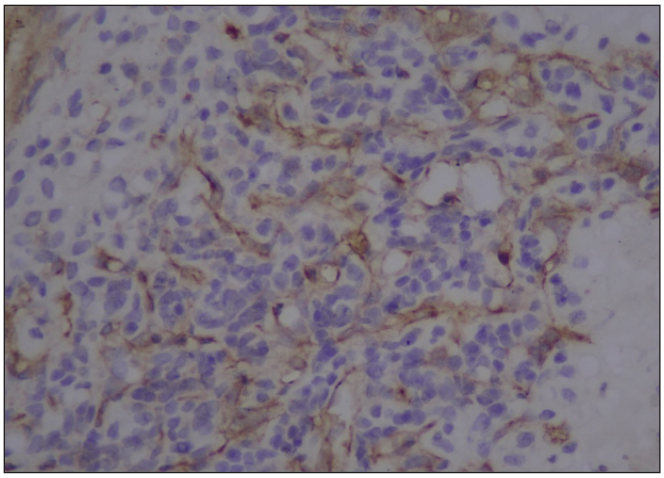

Fig. 4. EWS / PNET's are characterized Immunohistochemistry revealing strong positivity of tumour cells to the surface antigen CD99.

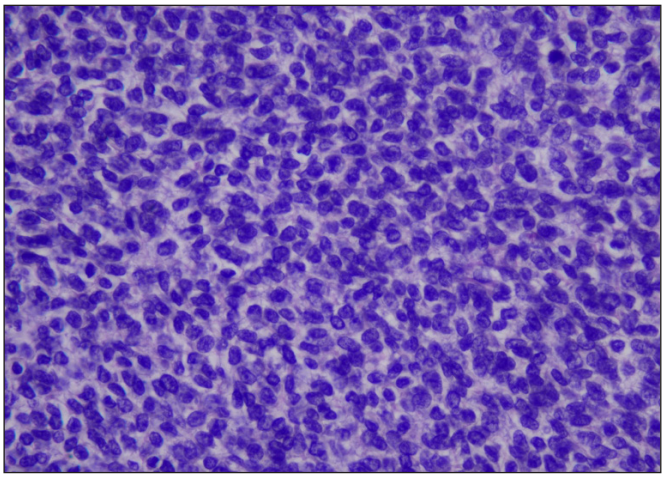

Fig. 5. Hematoxylin and eosin stain of typical EWS/PNET's at 20× magnification. Notes: Uniform small cells with round nuclei and fine chromatin are seen.

other study by Alan et al., reported the radiographic finding of their patient, demonstrating a circumscribed homogenous subcutaneous mass with no erosion of the calvarium [9]. The presentation of cranial ES is heterogenous, and awareness of extra skeletal ES as one of the differential diagnosis may aid in better management and outcome for patient.

This case is extremely rare, describing a patient with a rapidly progressive intracranial invasion of extraskeletal ES. Diagnosis of glomus jugulare was assumed, and excision of tumor was conducted with excellent early post-operative condition. However, a rapid progression of regrowth should alert a clinician that a malignant mass might be the cause of this condition. Pathological examination revealed ES, an aggressive tumor of bones and soft tissue. Radiotherapy was then planned for this patient. Unfortunately, since the waiting list for radiotherapy in our institution is quite long, the patient passed away before receiving further treatment. An early diagnosis and management in the suspicion of central nervous system ES should be done.

CONCLUSIONS

Extraskeletal ES is rare tumor with nonspecific clinical presentation and radiological features. It is locally aggressive and requires multimodality treatment with surgery and adjuvant chemoradiation therapy. Early diagnosis and treatment of intracranial invasion of extraskeletal ES may aid in better management for patient.

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Conflict of interest:
The Authors declare no conflict of interest.

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