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Case Report

Extraosseous Ewing sarcoma in the fossa jugular: A rare case report

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Received: 30 July 2023
Accepted: 09 October 2023
Published: 27 October 2023

DOI
10.25259/SNI_638_2023

Quick Response Code:

ABSTRACT

Background: Extraosseous Ewing sarcoma (EES) is a rare case that accounts for 20% of Ewing sarcoma cases. EES is the second most prevalent pediatric malignancy after peripheral primitive neuroectodermal tumors. EES mostly arise from soft tissue and extra-skeletal. Computed tomography (CT) and magnetic resonance imaging (MRI) are primary modalities for determining tumor location, characteristics, type, and extent of tumors. In addition, for presurgical management, radio intervention with arterial embolization is needed as a preoperative.

Case Description: We present a case of a 15-year-old boy diagnosed with EES. He had a "horn-like" tumor that grew progressively on his right ear over 5 months. Head CT scan and MRI were conducted to assess the extent. Embolization was performed before surgery. The surgery was conducted to excise the tumor radically. The histology pathology examination showed EES.

Conclusion: EES rarely occurs in the head and neck. This may manifest as a solid mass with bleeding components that destroy the nearby bones, with exophytic mass. Imaging is important for early finding and detecting complications of EES.

Keywords: Computed tomography scan, Ewing sarcoma, Extraosseous Ewing sarcoma, Magnetic resonance imaging

INTRODUCTION

Extraosseous Ewing sarcoma (EES) is a rare variant of Ewing sarcoma. The EES prevalence is 10 times lower than bone Ewing Sarcoma. EES is highly malignant, particularly in young adults.[1] This neoplasm is classified as a round well-differentiated cell tumor.[3] This tumor has a rapid growth often accompanied by localized pain within the soft tissue where the tumor grows. This neoplasm has some predilections such as the hands, shoulders, head, and neck. The clinical manifestations vary depending on their location.

A systematic review of 29 studies showed that the most commonly involved thoracic region is the most common site (33%), followed by extremities (31%) and then head and neck (14%).[6] These results are similar to previous literature that showed only 14% of cases were reported in the head and neck.

Another study (18 EES cases) performed a computed tomography (CT) scan in 13 patients and a magnetic resonance imaging (MRI) in five patients.[10] All of them (CT scan and MRI) showed soft-tissue masses with poorly defined borders and compressed surrounding structures due to tumor invasion. Hence, CT scans and MRIs were important to diagnose EES specifically. CT scans and MRIs also may determine its location, extent, and characteristics.[4,9]

EES rarely occurs in the head and neck. Diagnosing uncommon tumors with nonspecific clinical manifestations was challenging. An accurate diagnosis can significantly improve prognosis with a better survival rate.[5,8] Here, we present the case of a 15-year-old boy with EES in the head and neck.

CASE DESCRIPTION

A 15-year-old Asian boy came to the ear, nose, and throat–head and neck department with the chief complaint of a rapid lump behind the right ear, which had developed fast at 5 months. This "horn-like" tumor comes from the external meatus of the right ear. The tumor tends to bleed easily [Figure 1]. He also reported localized pain and loss of hearing. He also felt a decrease in body weight. There was no family history.

The physical examination revealed facial right nerve VII paralyzed, conductive hearing loss of the right ear, retroauricular erythema, and a bleeding mass from the external auditory externus. The tympanic membrane cannot be evaluated. The laboratory examination showed slight leukocytosis (11.210/μL), normal hemoglobin level (10 g/dL), and normal neutrophil count (81.7).

Head CT scan revealed soft-tissue solid mass enhancement (7.1 × 6.6 × 8 cm) in the right jugular fossa with an irregular border. The mass is exophytic into the middle ear, mesotympanum, and hypotympanum. It reaches the right external meatus. CT scan showed contrast enhancement (72HU) with evidence of mass-induced destruction in the right mastoid and right temporal bone. The feeding vessels come from the segment temporalis superficial and posterior segment auricular artery from the right external carotid artery (ECA) [Figures 2 and 3].

MRI demonstrated isointense to hyperintensity on the T1-weighted image, with prominent flow voids forming a "salt and pepper image" on the T2-weighted image and unrestricted diffusion in the diffusion-weighted imaging area, which lacks contrast enhancement. The mass destroyed the right mastoid, right temporal bone, and part of the sphenoid bone. There was an intracranial extradural extension. The mass compressed the right posterior fossa, right cerebellum, and right temporal lobe. The mass size was 7.8 × 6.7 × 6.6 cm

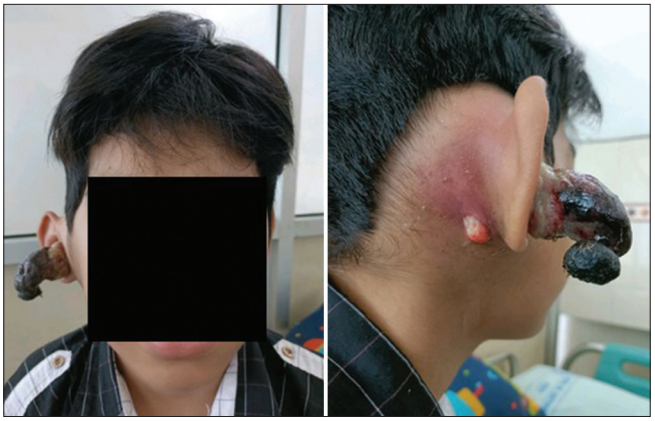

Figure 1: A 15-year-old boy, with a mass like "horn" came from acoustic externus.

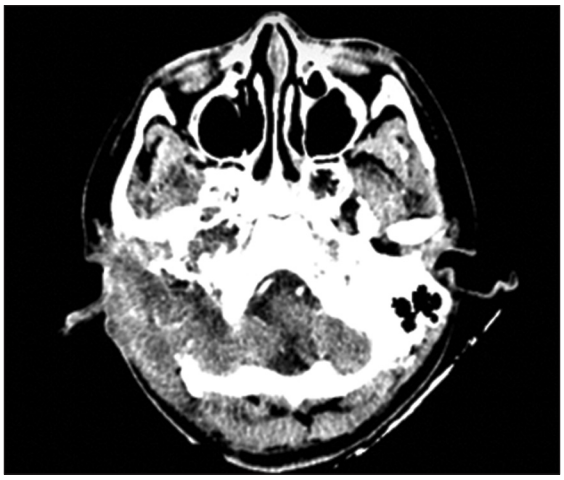

Figure 2: Head computed tomography with contrast in axial view soft-tissue solid mass enhancement, irregular border, in the right jugular fossa, and exophytic.

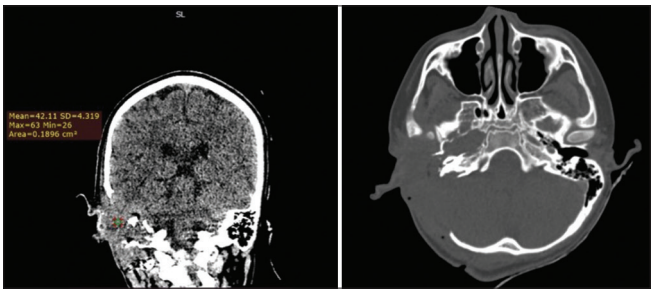

Figure 3: Head computed tomography with contrast in coronal view, soft-tissue solid mass enhancement, irregular border, like in the right jugular fossa that exophytic to the middle ear, meso, and hypotympanum until right external meatus, with destruction in the right mastoid and right temporal.

extending to the right fossa jugular, with exophytic to the middle ear, mesotympanum, and hypotympanum until

the right external meatus with feeding arteries from the superficial temporal segment and the posterior auricular artery segment of the right ECA branch [Figures 4-6].

The definitive management approach for this patient involved radical surgery, preceded by digital subtraction angiography (DSA) abolition before surgery. The DSA angiography revealed a hypervascular lesion supplied by the right ECA, with branches from temporalis superficialis and right occipital arteries. We performed embolization using PVa 355-500 mm in the ECA branch to the occipital and superficial right temporalis arteries [Figure 7]. After DSA, a craniotomy was conducted to excise the tumors. The tumor was completely removed and released detachment from healthy dura. The specimen was sent to the pathology anatomy department. This specimen was examined with an immunohistochemistry stain (i.e., CK antibody, vimentin, S100, SMA, synaptophysin, desmin, CD-34, and CD-99). The pathology examination showed EES characterized by small uniform primitive cells with round nuclei, scanty cytoplasm appearance, with positive in vimentin immunohistochemistry examination [Figure 8].

DISCUSSION

This patient was 15 years old. The previous studies showed that this condition commonly occurs in individuals below 20 years.[6] The diagnosis was established by comprehensive assessment, physical examination, laboratory test, radiology, and pathology examination. Head and neck CT scans and/or MRIs are important to identify the tumor location, the area of destruction, and the involved organs. These tumor characteristics were important to determine the appropriate clinical management.[2,4]

This patient had a solid mass (approximately $10 \times 7, 6 \times 81$ cm) in the right fossa jugular with a bleeding component. MRI showed that this tumor had vascularization from the temporalis superficialis segment artery and posterior segment auricular artery. MRI also showed an exophytic in the middle ear, involving epi, meso, and hypotympanic until the right meatus externus. Contrast-enhanced CT scan showed rim contrast enhancement and mass destruction of the right mastoid and right temporal bone, with evidence of intracranial extradural extension.

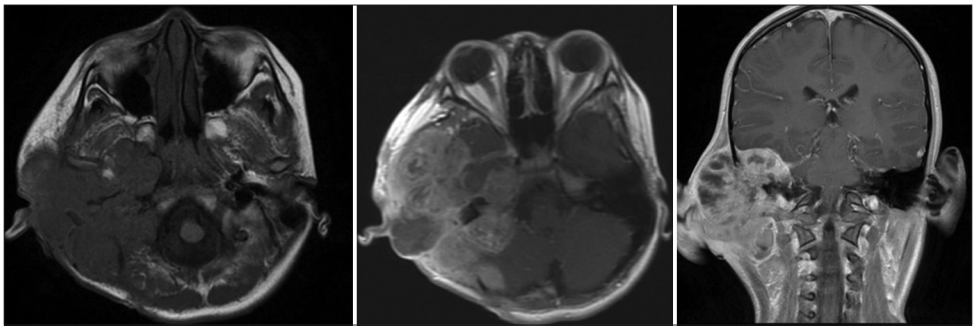

Figure 4: Head magnetic resonance imaging with contrast axial and coronal view, there is hyperintense on T1-weighted image, a mass was seen destroying the right mastoid, right temporal bone, and part of the sphenoid, visible intracranial extradural extension.

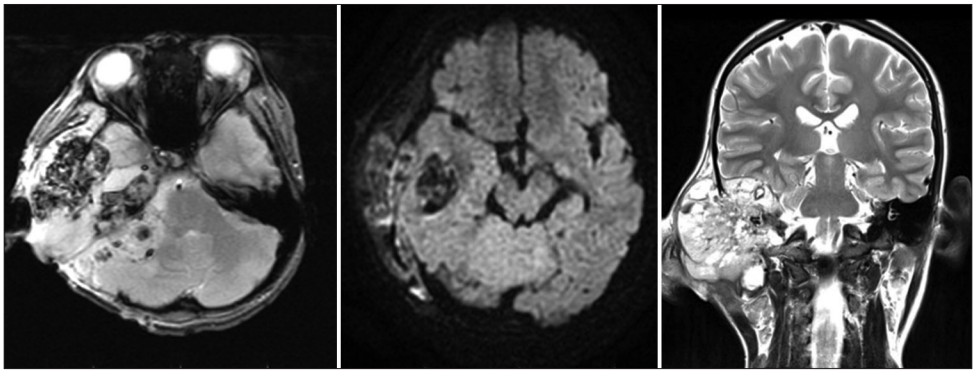

Figure 5: Head magnetic resonance imaging with contrast axial and coronal view, hyperintense with prominent flow voids forming a salt and pepper image on T2-weighted image, unrestricted diffusion in the diffusion-weighted imaging area, which did not show contrast enhancement, a mass was seen destroying the right mastoid, right temporal bone and part of the sphenoid looks intracranial extradural extension.

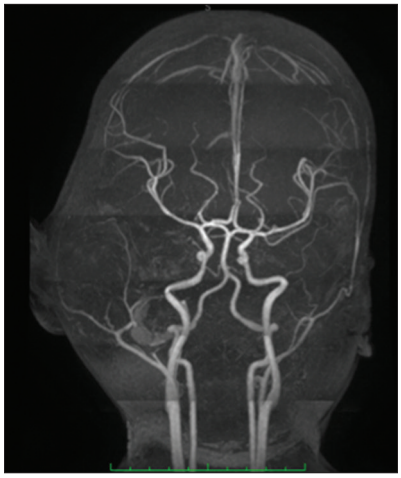

Figure 6: The mass is seen feeding from the superficial temporal segment and the posterior auricular artery segment of the right external carotid artery branch.

According to this finding, this case was decided to surgical intervention. Presurgical DSA to minimize intraoperative bleeding was conducted. The histopathology examination showed EES rather than a rhabdomyosarcoma though both belong to the same in the round cell family tumor. It could be a differential diagnosis, it was positive in CD-99, S-100, and synaptophysin.[1,9,8]

The patient had improved general condition after tumor resection but he passed away after chemotherapy. It is essential to note that the standard therapy of EES is radical resection combined with chemotherapy. Radiotherapy may also play an important role. Nevertheless, it is crucial to be aware that secondary metastases can develop.[7] Indeed, the EES had a poor prognosis with a survival rate of around 70% in 5 years. The prognosis will be worse in advanced EES (tumor volume more than 200 cc and the age is more than 14 years).[3]

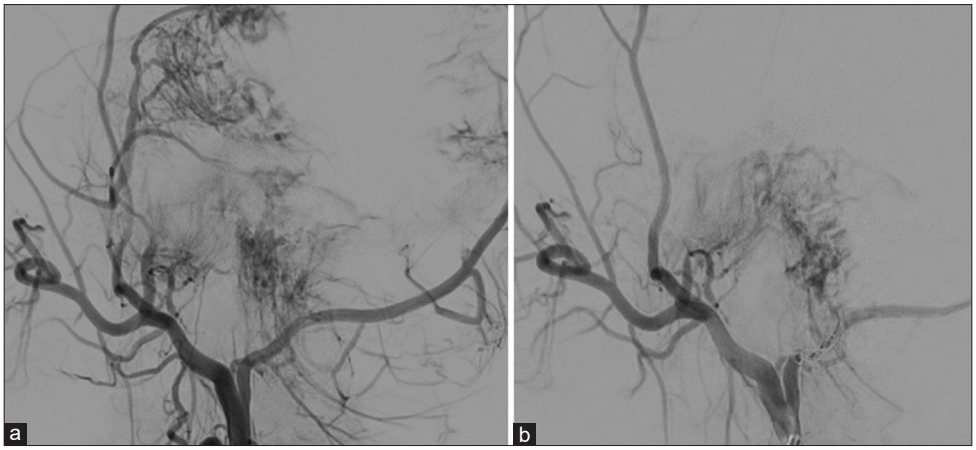

Figure 7: (a) Pre Embolization and (b) Post embolization intra-arterial embolization were done at the maxillary artery, middle deep temporal branch, meningeal accessories artery, and occipital artery use 250 micron–355 microparticle.

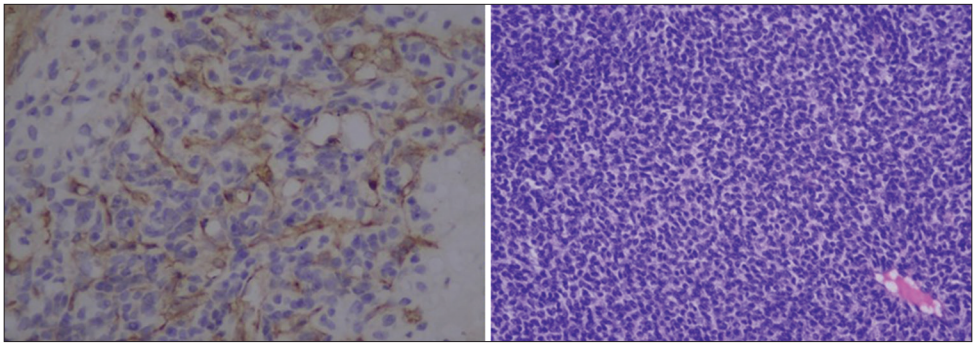

Figure 8: A small uniform primitive cell with round nuclei and scanty cytoplasm appearance.

CONCLUSION

Here, we present a case of a 15-year-old boy diagnosed with EES characterized by a giant mass that caused bone destruction and exophytic growth toward the right ear. Prompt diagnosis and early diagnosis must be established in this patient because EES is the second most common malignancy among teenagers. Early intervention has better outcomes. Biopsy is a gold standard. CT and MRI are also important for tumor evaluation. Presurgical DSA is important to minimize intraoperative bleeding.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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How to cite this article: Marthawati S, Ferriastuti W, Mukherji SK, Artono A. Extraosseous Ewing sarcoma in the fossa jugular: A rare case report. Surg Neurol Int 2023;14:382.