Case Report

Epithelioid angiosarcoma of the middle ear: A case report of a new location

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Abstract

Epithelioid angiosarcoma (EA) is a rare, highly malignant tumor that affects mainly the deep soft tissues of the extremities and has a poor prognosis. This angiosarcoma occurs rarely in the middle ear. Herein, we describe an unusual case of 36-year-old man who presented with an 8-year history of progressive left-sided hearing loss and unbearable otalgia for 1-month. Otoscopic examination showed that the left external auditory canal was dry and the tympanic membrane was intacted. An audiogram demonstrated a moderate mixed hearing loss in the left ear. High-resolution computed tomography of the temporal bone demonstrated a huge destructive lesion with significant bone erosion, a round soft tissue mass located mainly in the left mastoid cavity measuring 3.0 cm × 3.4 cm × 3.1 cm, and some soft tissue among the rest of the mastoid air cells and tympanic cavity. The appearance of the lesion did not change with contrast enhancement. Magnetic resonance imaging showed that the soft mass had mixed signal intensity with heterogeneous enhancement on T1- and T2-weighted images and well-defined margins. Histopathology and immunohistochemistry revealed an EA. To our knowledge, this is the first reported case of an EA with clear boundaries located mainly in the mastoid cavity.

Key words: Epithelioid angiosarcoma, immunohistochemistry, pathology, primary prognosis

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INTRODUCTION

Primary tumors of the middle ear are rare clinical entities and include squamous cell carcinoma, adenocarcinoma, lymphoepithelial carcinoma, rhabdomyosarcoma, multiple myeloma, and plasmacytoma. To our knowledge, no reports of epithelioid angiosarcoma (EA) in the middle ear have been published in the English literature, and only 1 case of EA arising from the middle ear has been reported in the Chinese literature. In the latter case, the EA had invaded the external auditory canal, middle ear, and mastoid, and the boundary was unclear.[1] Herein, we present a unique case of EA with clear boundaries that were located mainly in the mastoid cavity.

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CASE REPORT

A 36-year-old man presented with an 8-year history of progressive left-sided hearing loss. He did not attend a hospital for treatment until he had experienced unbearable otalgia in his left ear lasting 1-month. He denied any history of ear infections, ototoxic medication exposure, otorrhea, tinnitus, vertigo, head trauma, or family history of hearing loss. No facial nerve palsy symptoms were observed. Otoscopic examination showed that the left external auditory canal was dry and the tympanic membrane was intacted. An audiogram demonstrated normal hearing in the right ear and a moderate mixed hearing loss in the left ear. High-resolution computed tomography (HRCT) of the temporal bone demonstrated a huge destructive lesion with significant bone erosion, a round soft tissue mass located in the left mastoid cavity measuring 3.0 cm × 3.4 cm × 3.1 cm, and some soft tissue among the rest of the mastoid air cells and tympanic cavity. The left malleus showed suspicious bone absorption, and the appearance of the lesion did not change with contrast enhancement [Figure 1a-c]. Magnetic resonance imaging (MRI) showed that the mass had mixed signal

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intensity with heterogeneous enhancement on T1- and T2-weighted images, had well-defined margins, and was in contact with the sigmoid sinus [Figure 2a and b].

Cholesteatoma was suspected, and a mastoidectomy was performed through the retroauricular approach under general anesthesia. Intraoperatively, a round, soft, yellow-white tumor-like mass that filled the left mastoid cavity and middle ear [Figure 3] and slight adhesions to the sigmoid sinus and meninges were observed. During the operation, one biopsy was taken for frozen-tissue analysis, and a malignant tumor was indicated. Histology showed that the heteromorphic epithelioid cells were arranged mainly in nests or sheets and contained abundant cytoplasm and prominent nucleoli [Figure 4a]. Immunohistochemistry (IHC) showed positive staining for the vascular markers CD 31 and CD 34 [Figure 4b], which confirmed the diagnosis, and negative staining for smooth muscle actin, pan-cytokeratin, S-100 protein, and HMB45 (En-Vision). Staining for Ki-67 was about 20%, which confirmed the highly proliferative nature of the tumor. These findings were consistent with a diagnosis of EA. The patient received radiation therapy beginning 6 weeks after surgery. He received a total of 66 gray (Gy) in 33 fractions over 49 days, and there was no evidence of tumor recurrence 4 months after treatment.

DISCUSSION

First documented by Fletcher *et al*.[2] in 1991, EA is characterized by aggressive biological behavior and accounts for <2% of all sarcomas.[3] It has a male predilection and a peak incidence in the seventh decade of life, and arises most often in the deep soft tissues of the extremities,[2] although other primary sites have been reported including the thyroid gland, adrenal glands, gallbladder, skin, esophagus, tonsil, bone, and middle ear.[1]

The clinical diagnosis of EA in the middle ear is difficult because of the nonspecific clinical course and otoscopic and radiological features. However, a pre-operative radiological evaluation is necessary to rule out pathological changes to important structures, and it is important to distinguish EA from a cholesteatoma. Cholesteatoma is a soft tissue mass that can cause bony erosion, which

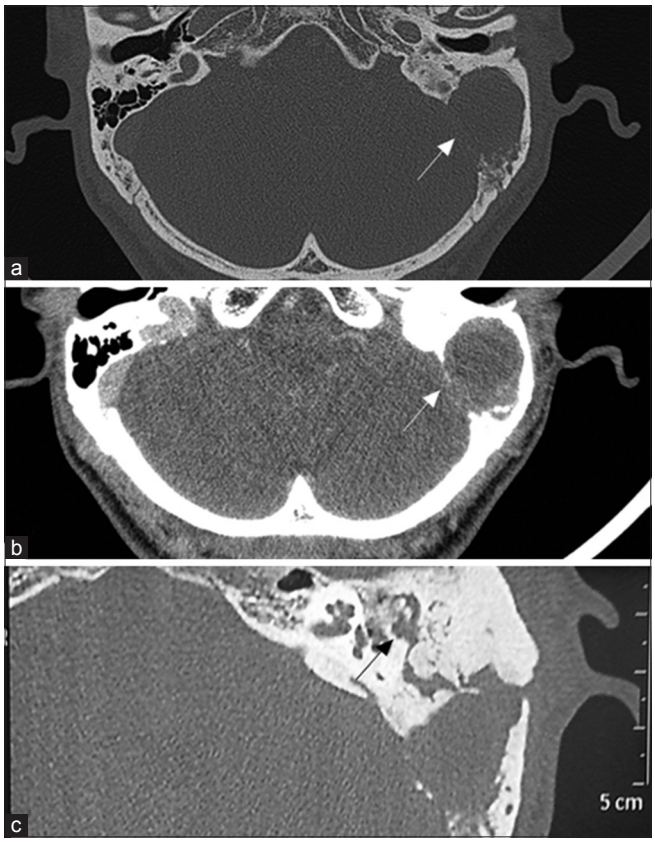

Figure 1: Axial computed tomography of the left temporal bone. (a) The temporal bone exhibited a huge destructive lesion and significant bone erosion, and a round soft tissue mass was located in the left mastoid cavity (arrow showed). (b) The appearance of the lesion did not change with contrast enhancement (arrow showed). (c) Some soft tissue was seen in the rest of the mastoid air cells and tympanic cavity, and the left malleus showed suspicious bone absorption (arrow showed)

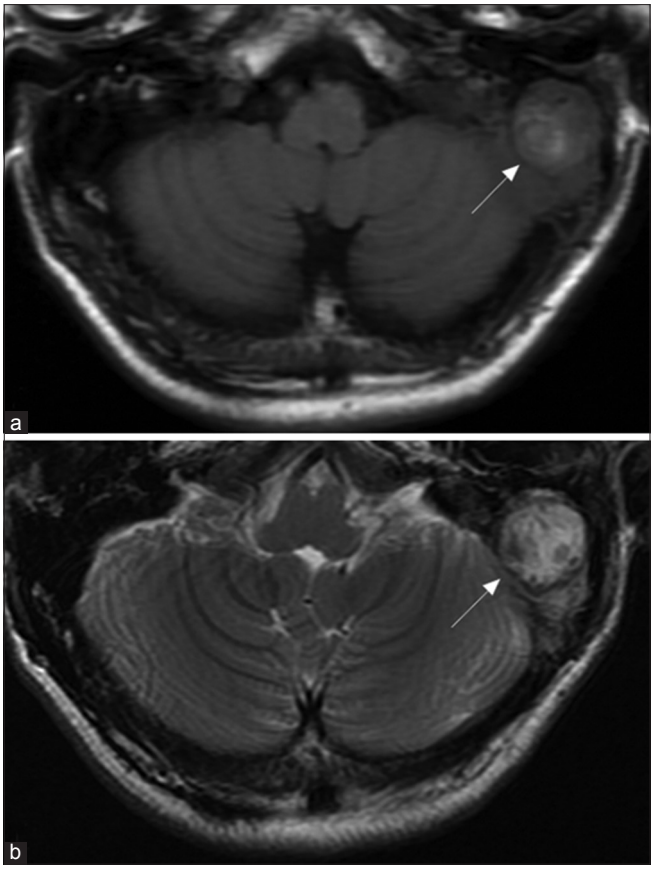

Figure 2: Axial magnetic resonance imaging of the left middle ear shows that the mass had mixed signal intensity with heterogeneous enhancement on T1- and T2-weighted images and well-defined margins, and was in contact with the sigmoid sinus (arrow showed). (a) T1-weighted images. (b) T2-weighted images

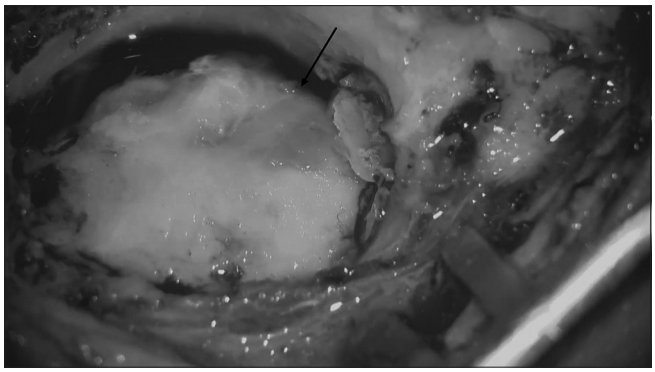

Figure 3: A round, soft, yellow-white mass, which filled the left mastoid cavity and middle ear, was observed

usually exposes surrounding structures. On HRCT, a cholesteatoma appears as hypointense or isointense on T1-weighted and hyperintense on T2-weighted images. The mass does not enhance after gadolinium infusion or shows only some rim enhancement on MRI.[4]

Hwang and Lim[5] reported that fluorine-18 fluorodeoxyglucose positron emission tomography/CT is helpful in the diagnosis of EA. Confirmation of the diagnosis of EA relies on pathology examination, but it is difficult to diagnose using hematoxylin and eosin-stained sections. EA comprises neoplastic cells with abundant amphophilic or eosinophilic cytoplasm and large vesicular nuclei. IHC is an important adjunctive procedure in the diagnosis, and the neoplastic cells stain positively for CD31, CD34, factor VIII-related antigen, and vimentin.[6] Among them, CD31 is the most sensitive and specific endothelial cell marker and is expressed by about 90% of angiosarcomas.[7] CD34 positivity has been reported to range from 40% to 100% but is much less specific and this marker is expressed by several other types of soft tissue tumor.[7] In our case, there was strong and diffuse expression of CD31 and CD34, which led us to diagnose EA.

Because of its rarity, early diagnosis is usually not possible. EA may be misdiagnosed as an epithelial tumor, such as epithelioid hemangioendothelioma or malignant peripheral nerve sheath tumor. The differential diagnosis of primary EA of the middle ear includes cholesteatoma, squamous cell carcinoma, and other poorly differentiated sarcomas, which can be distinguished by IHC. In the cases of EA reported, the treatment has included wide resection, chemotherapy, and radiotherapy. EA has an aggressive course and tends to recur locally, spread widely, and have a high rate of metastasis to lymph nodes, lungs, bone, soft tissue, and skin. The prognosis for EA is very poor, and death usually occurring within 2–3 years of the diagnosis.[8] In the current patient, the treatment included surgery and radiation, and after 4 months following treatment, the tumor remains under control and the patient is asymptomatic.

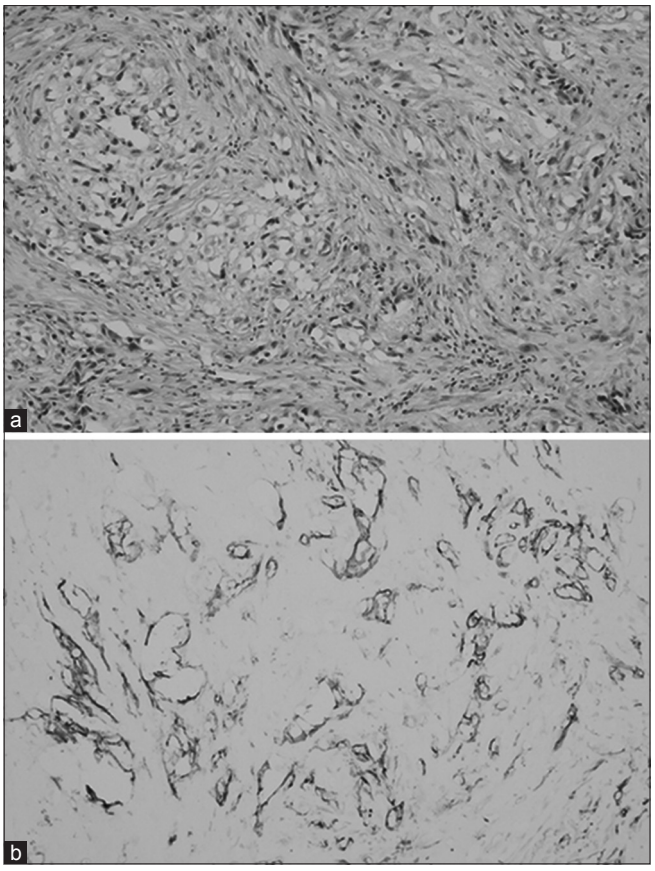

Figure 4: (a) Histopathological examination of the lesion showed heteromorphic epithelioid cells arranged mainly in nests or sheets and that these cells contained abundant cytoplasm and prominent nucleoli (H and E, original magnification ×200). (b) Immunohistochemistry showed positive staining for CD 34 (En-Vision, original magnification ×200)

In summary, EA originating from the middle ear is an extremely rare, aggressive malignant vascular tumor. The symptoms and otoscopic and radiological findings are not specific, and IHC is helpful for making a correct diagnosis. EA remains a difficult disease to treat and has an overall poor prognosis. Surgical resection together with adjuvant radiation or chemotherapy is the best treatment modality.

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