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MIDDLE EAR ADENOMA WITH AN ATYPICAL CLINICAL PRESENTATION

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ABSTRACT

In this study, we aim to report the case of a patient who had a middle ear adenoma with an atypical presentation. The patient complained of a cyclic (monthly) bleeding in the right ear for the past 10 years, which occurred concomitantly with her menstruation period. She also had tinnitus, hearing loss, and sporadic otorrhea in the same ear. Tomography and magnetic resonance revealed a contrast-enhancing mass occupying the middle ear cleft, without bone erosion or invasion of adjacent structures. We decided to remove the tumor "en bloc", after performing a canal wall-down mastoidectomy to better assess the limits of the lesion. Histopathologic evaluation confirmed the diagnosis of middle ear adenoma. The diagnosis, treatment options and prognosis of the middle ear adenoma are further discussed and critically reviewed.

General Terms: Diagnosis, treatment, middle ear tumors, middle ear adenoma

Keywords: Middle ear adenoma, Neurendocrine adenoma, middle ear, tumor, diagnosis, treatment, prognosis

1. INTRODUCTION

Middle ear adenoma (neuroendocrine adenoma) is a rare benign tumor that affects the temporal bone. These tumors may arise either from glandular and neuroendocrine epithelial cells from the middle ear mucosal or from the neuro-ectoderm of the neural crest.[1-3] Although benign, mechanical compression of the middle and inner ear structures caused by expansion of the tumor may lead to bone erosion and damage to adjacent structures.[4-7] The most common clinical symptoms resulting from this tumor are either caused by compression, obstruction, or local erosion/destruction, and they include ipsilateral hearing loss, otorrhea, local pain, facial nerve injury, and tinnitus.

The objective of this study is to report the case of middle ear adenoma with an atypical clinical presentation, and to critically discuss diagnostic methods, treatment, and prognosis of the disease.

2. CASE REPORT

A 57-year-old woman complains of a recurrent bleeding in the right ear, that occurred once a month for the past 10 years. She stated that the bleeding always occurred during her menstruation period. She also reported pulsatile tinnitus and ipsilateral hearing loss. She

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had sought medical attention for this problem several times in the past, and she had been subjected to many cycles of antibiotic therapy (either topic or systemic); nonetheless, her condition never improved.

Otoscopy of the right ear demonstrated the presence of a soft mass in the lower half of the external auditory canal (Figure 1); we could not identify the origin of the lesion. The rest of the otolaryngologic and physical examination was unremarkable. Audiometric evaluation showed moderate mixed hearing loss in the right ear; the left ear had normal auditory thresholds.

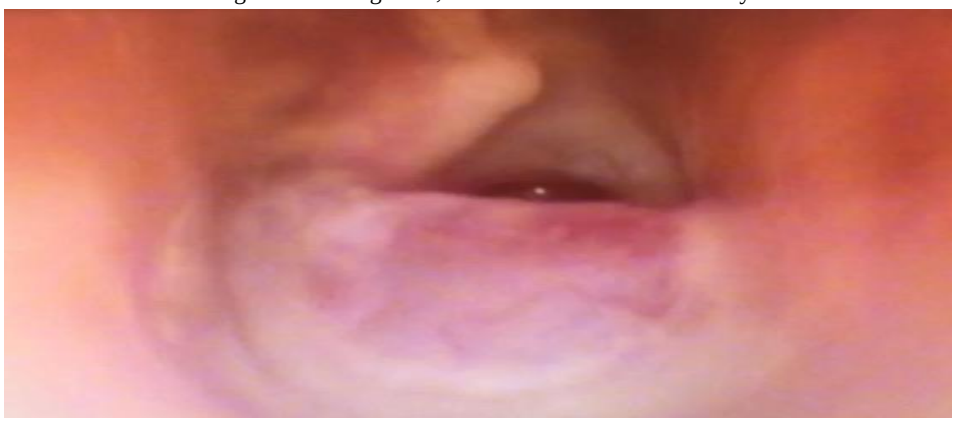

Figure 1. Otoscopy from right ear. It is possible to observe a mass occupying the lower half of the external auditory canal.

To better evaluate the origin and the characteristics of the lesion, we performed a computed tomography (CT) and a magnetic resonance imaging (MRI) of the temporal bones. The CT scan demonstrated the presence of a soft tissue attenuation material which filled the antrum of the mastoid, aditus-ad-antrum, and the middle ear. We did not observe erosion of the bony structures in this side. CT of the right temporal bone was unremarkable. (Figure 2).

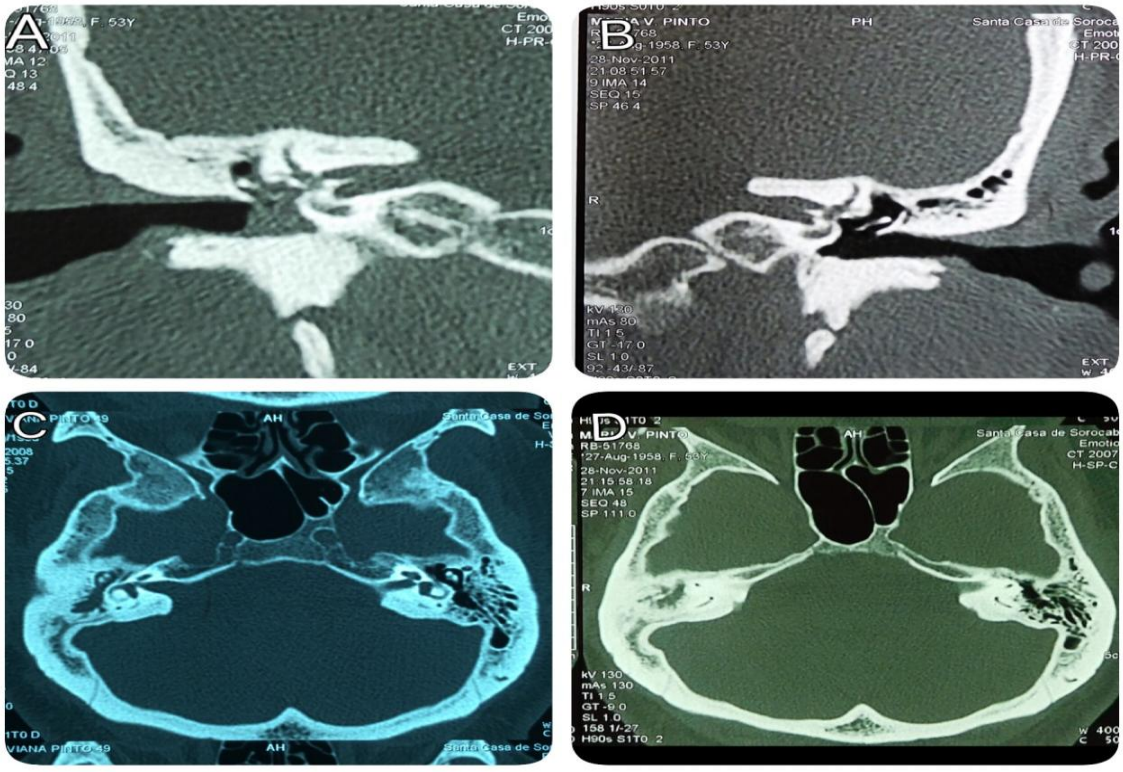

Figure 2. CT scan of the temporal bone without contrast enhancement, bone window. A: Coronal plane slice of the right temporal bone demonstrating the presence of a soft tissue attenuation material occupying the middle ear; the ossicles, bony walls and inner ear

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are intact. The mass projects to the external ear canal through the tympanic membrane. B: Coronal plane slice of the left temporal bone – unremarkable. C: Axial plane slice. Right: we observe a sclerotic mastoid, with a soft tissue attenuation material filling the epitympanum, aditus ad antrum, and the antrum of the mastoid; Left side: unremarkable. D: Axial plane slice. Right: Sclerotic mastoid; left: unremarkable.

MRI demonstrated an hypointense mass occupying the middle ear and mastoid on T1 ponderation, and an isodense material filling the same compartments in T2 ponderation. The T1 ponderation image under paramagnetic contrast revealed an enhancement of the material filling the right middle ear. The inner ear structures were normal. The left side was unremarkable (Figure 3).

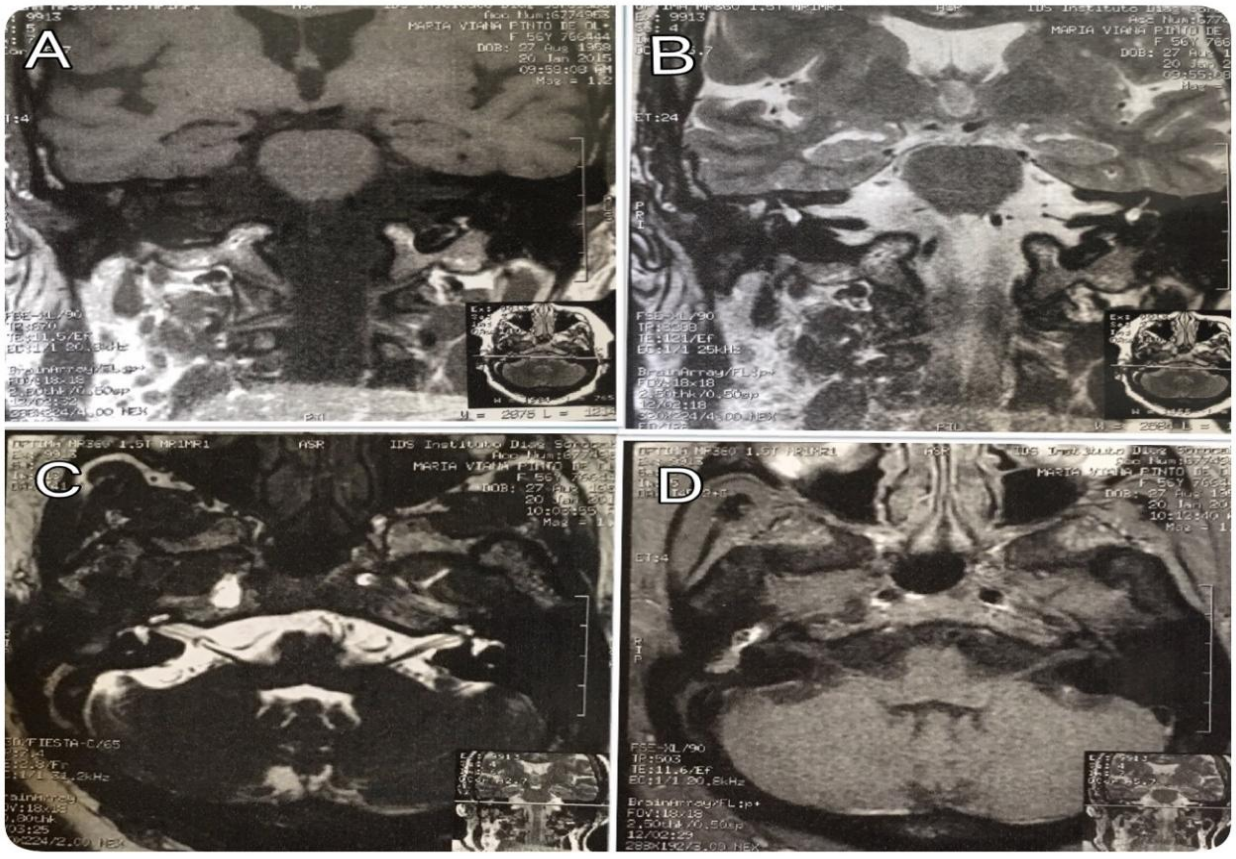

Figure 3. MRI of the temporal bones and internal auditory canals. A: Coronal, T1-pondered image, showing a hypointense mass occupying the middle ear on the right side. B: Coronal, T2-pondered image showing an isointense image in the right middle ear cleft. C: Axial, T2-pondered image showing integrity of the nerves in the internal ear canal, as well as the cochlea and vestibular structures. D: Axial, T1-pondered under paramagnetic contrast image, demonstrating that the material filling the middle ear and mastoid antrum was enhanced by the contrast.

Based on the findings of the physical examination and CT and MRI scans, our main hypothesis was of a tumor affecting the middle ear cleft, probably benign due to its long evolution and lack of invasion of adjacent structures and great vessels. We decided to perform a complete excision of the lesion, based on a canal wall-down mastoidectomy, to assess all the limits of the lesion and to ensure its complete removal. Intra-operatively, we observed that the mass in the external ear canal was an extension of the middle ear tumor, extruding from the middle ear through the tympanic membrane. The lesion was successfully removed “en bloc” and sent for histopathologic analysis.

GLOBAL JOURNAL OF ADVANCED RESEARCH
(Scholarly Peer Review Publishing System)

Initial follow-up was eventful, and the patient reported that the ear bleeding stopped after the surgery. Histopathological analysis revealed a polypoid lesion with proliferation of epithelial coated microcysts without atypia. Immunohistochemical analysis confirmed the diagnosis of middle ear adenoma.

3. DISCUSSION

The middle ear adenoma is a very rare tumor. The symptoms resulting from such lesion are similar to several other diseases affecting the ear; thus, thorough physical examination, clinical history, and imaging evaluations are needed to perform a correct diagnosis. The classic symptoms of middle ear adenoma include conductive hearing loss, tinnitus, otorrhea, and local pain.[1-3] Our patient presented an atypical symptom: she had otorrhagia in her right ear only during her menstruation period. We found no parallel study in the literature to support a possible pathophysiologic mechanism that may explain that clinical feature. Nonetheless, the pro-thrombotic effects of the estrogen have been extensively described in the literature[8]; it is possible that areas of focal necrosis of the tumor secondary to mechanic compression caused by the growth of the tumor against the bony walls of the middle ear cleft could have led to bleeding, especially in the times of a decrease of estrogen in the blood stream during the menstruation period.

Among the main differential diagnoses of middle ear tumors, the tympanic glomus and the schwannoma of the tympanic segment from facial nerve are the most frequent lesions affecting that area of the temporal bone[9]. Other less frequent lesions affecting the middle ear cleft include adenomatous tumors (adenomas, adenocarcinomas, and adenoid cystic carcinomas), ceruminomas and paragangliomas[10]. Furthermore, middle ear hemangiomas and meningioma of the tegmen tympani may also present with similar characteristics in the CT and MRI.

Imaging tests such as computed tomography and nuclear magnetic resonance are important to assess the origin and extension of the lesion. These exams provide important information regarding the anatomical relationships of the lesion to the middle and inner ear structures, and they are considered important tools in both performing the diagnosis and surgical planning[3-5]. Nonetheless, the diagnostic confirmation is performed by histopathologic analysis of biopsy fragments under light microscopy, immunohistochemistry, and electron microscopy[5-7,11,12]. Characteristically, light microscopy analysis reveals regular, cuboidal and columnar cells, which form small, juxtaposed glands, or in solid, or trabecular, arrangements. Periodic acid-Schiff (PAS) staining may be positive for mucoprotein content in the lumen of some glandular structures[11].

The treatment of middle ear adenomas is surgical resection. In cases in which the surgical treatment is not possible due to poor clinical conditions, radiotherapy is considered a therapeutic option; however, to our knowledge, there is no evidence in the literature regarding the efficacy of the radiotherapy in these tumors[13]. There is consensus among authors that the middle ear adenoma does not require additional therapies other than surgical removal followed by mastoidectomy, given its benign behavior, low rate of recurrence and absence of metastatization[14-17].

Recurrence of middle ear adenoma is rare when the tumor is excised completely. However, some studies observed the possibility of long-term relapse after 10 years of the removal, and the main factor associated with recurrence is the size of the tumors – large tumors seem to have a higher incidence of recurrence when compared to smaller lesions[9,14]. Therefore, long-term follow-up is required for these patients, and it should be based on otoscopy findings, audiometric evaluation, and eventual computed tomography and/or magnetic resonance imaging in search of early signs of recurrence[9,14].

4. CONCLUSION

Middle ear adenoma is a benign, rare lesion that can cause destruction of adjacent structures. The clinical diagnosis is difficult, since the characteristic symptoms are similar to those resulting from other diseases affecting the middle ear and external ear canal. The diagnosis is confirmed by histopathologic analysis of material obtained by biopsy, associated with immunohistochemical analysis.

5. REFERENCES

[1] Davies JE, Semeraro D, Knight LC, Griffiths GJ. Middle ear neoplasms showing adenomatous and neuroendocine components. J Laryngol Oto 1989; 103(4):404-7.
[2] Ketabchi S, Massi D, Franchi A, Vannuchi P, Santucci M. Middle ear adenoma is na amphicrine tumor: why call it adenoma? Ultrastruct Pathol 2001; 25(1):73-8.
[3] Gunduz M, Yamanaka N, Saito T, Kuki K, Yokoyama M, Nakamine H. Middle ear adenoma with neuroendocrine differentiation. Auris Nasus Larynx 2000; 27(1):73-6.
[4] Namyslowski, G,Scierski, and W, Lange D. 2001. Middle ear adenoma: a case report. Ultrastruct Pathol (2001); 25(1):73-8

GLOBAL JOURNAL OF ADVANCED RESEARCH
(Scholarly Peer Review Publishing System)

[5] Bailey QR, and Weiner JM. 1986. Middle ear adenoma: a case report with ultrastructural findings. J Laryngol Otol (1986); 100(4):467-70.
[6] Kundig H, Kos I, Kurt AM, and Guyot JP. 2000. Adenoma of the middle ear. Ann Otolaryngol Chir Cervicofac (2000); 117(5): 274-80.
[7] Hale RJ, McMahon RF, and Whittaker JS. 1991. Middle ear adenoma: tumour of mixed mucinous and neuroendocrine differentiation. J Clin Pathol (1991); 44(8):652-4.
[8] Scarabin PY, Alhenc-Gelas M, Plu-Bureau G, Taisne P, Agher R, and Aiach M. 1997. Effects of oral and transdermal estrogen/progesterone regimens on blood coagulation and fibrinolysis in postmenopausal women. A randomized controlled trial. Arterioscler Thromb Vasc Biol. (Nov 1997);17(11):3071-8
[9] Hu H, Lim WY, Tan TY, and Yuen HW. 2016. Neuroendocrine adenoma of middle ear with new bone formation and review of literature. Am J Otolaryngol. (2016);37:108-111
[10] Ribe A, Fernandez PL, Ostertarg H, Claros P, Bombi JA, Palacin A et al. 1996. Middle-ear adenoma (MEA): a report of two cases, one with predominant "plasmacytoid" features. Ann Pathol (1996); 16(4):271-5.
[11] Sahan M, Yildirim N, Arslanoglu A, Karslioglu Y, and Kazikdass KC. 2008. Carcinoid tumor of the middle ear: report of a case. Am J Otolaryngol. (2008);29:352-6.
[12] Ferlito A, Devaney KO, and Rinaldo A. 2006. Primary carcinoid tumor of the middle ear: A potentially metastasizing tumor. Acta Otolaryngol. (2006);126:228-31.
[13] Hasan, Z, McGinness S, Gunaratne DA, Coleman H, Varikatt W, and da Cruz M. 2016. Neuroendocrine Adenoma of the Middle Ear: A Rare Histopathological Diagnosis. Case Rep Otolaryngol. (2016) 2016.
[14] Bakhos D, Lescanne E, Fetissof F, Robier A, and Morinière S. 2007. Neuro-endocrine adenoma of the middle ear: a case study. Eur Arch Otorhinolaryngol (2007) 264:1525–1528
[15] Wassef M, Kanavaros P, Nemeth J, Adjamagbo H, and Ba Huy PT. 1993.Amphicrine adenoma of the midddle ear. Histological, immunohistochemical and ultrastructural study of a case. Ann Pathol 1993; 13(3):170-5.
[16] Torske KR, and Thompson LD. 2002. Adenoma versus Carcinoid Tumor of the Middle Ear: a Study of 48 Cases and Review of the Literature. Mod Pathol (2002); 15(5):543-55.
[17] Paraskevakou H, Lazaris AC, Kandiloros DC, Papadimitriou K, Adamapoulos G, and Davaris PS. 1999. Middle ear adenomatous tumor with a predominant neuroendocrine component. Patlhology 1999; 31(3):284-7.