

## Introduction

Leiomyoma (LMA) of the external auditory canal (EAC) is a benign tumor of the smooth muscle first described by Petschenik AJ and Linstrom CJ in 1996[1]. The involvement of head and neck regions is rare, while LMAs are common in the uterus and alimentary tract [2]. In the head and neck regions they are more common in females (3.5:1). We present the case of a patient affected by a **vascular LMA/angioleiomyoma** of the external auditory canal. Clinical and **histopathological findings are described and discussed. As far as we know, only nine cases of LMA of the EAC** have been reported in the literature to date. [3].

## Case report

A 51-year-old Caucasian female presented with a one-month history of right auricular fullness and hearing loss without other symptoms or history of trauma. In our hospital she was submitted to otomicroscopic exam that revealed a smooth non-bleeding mass occluding the right external auditory canal. Pure-tone audiometry showed right mild mixed hearing loss.

Computed tomography showed the presence of hypodense material that occupied the external auditory meatus without mastoid involvement. MRI demonstrated a well-defined, oval mass. It was isointense to muscle on T1-weighted spin echo images and with multiple linear or branching areas of hyperintensity on T2-weighted ones. The enhancement after gadolinium was heterogeneous.

Endoaural surgery with a total resection was performed and the skin of the EAC was reconstructed using a Thiersch flap.

Histological examination revealed a proliferation of closely packed oval/spindle cells associated with a florid vasculature (Fig. 1A and 1B). Adipocytes were not detected within the lesion. The oval/spindle cells were **diffusely** immunoreactive for Smooth Muscle Actin (Fig. 1C) **and focally for Muscle Specific Actin and h-Caldesmon** and negative for HMB45, **a marker for melanocytic and**

myomelanocytic tumors [4]. The vasculature was highlighted by the immunostain for the endothelial markers CD31 and CD34 (Fig. 1D) and consisted of predominantly slit-like channels. Necrosis and mitosis were absent. The proliferation index (Ki67) was less than 5%. These findings were consistent with a diagnosis of vascular LMA/angioleiomyoma.

At 6-month follow-up, the patient was disease-free with normal hearing.

## Discussion

LMA is a benign tumor rarely arising in the head and neck region. Its origin in the EAC is even less common. As far as we know, only nine cases of LMA of EAC have been previously reported in literature, the minority of which consistent with vascular LMA/angioleiomyoma.

In our case, the patient presented auricular fullness and hearing loss without pain. However, pain not responsive to medical treatment remains the most common ear symptomatology [1]. Macroscopically LMA of EAC appears as an oval lesion that obliterates the entire cavity and that usually does not allow visualization of the implant base.

Three main histological subtypes of vascular LMA/angioleiomyoma have been distinguished [5]: solid, cavernous and venous ones. Solid-type tumors are composed of fascicles of smooth muscle cells intercalated between slit-like vascular channels. Cavernous-type tumors are composed of dilated vascular channels with less smooth muscle bundles than the other types. Venous-type tumors have vascular channels with dense muscular walls and non-compact smooth muscle bundles. By immunohistochemistry, neoplastic cells are immunoreactive for smooth muscle cell markers and negative for endothelial markers and for HMB45, a marker for melanocytic and myomelanocytic tumors [4]. Imaging techniques (CT and/or MR) give information only regarding the site and the extent of the disease, but are not able to provide a diagnosis of nature due to non-specific imaging characteristics. Surgery is the treatment of choice and the lesion must be excised totally to avoid recurrences that, however, occur very rarely [5].

## References

[1] Petschenik AJ, Linstrom CJ, McCormick SA. Leiomyoma of the external auditory canal. *Am J Otol* 1996; 17: 133-6.

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[3] Tombolini M, Ralli M, Carletti R et al. Leiomyoma of the External Auditory Canal. *J Int Adv Otol*. 2019 Apr; 15(1): 169–172.

[4] Goldblum JR, Folpe AL, Weiss SW. *Enzinger and Weiss's Soft Tissue Tumors*. 7th ed. Philadelphia, Elsevier 2020, p 154.

[5] Hachisuga T, Hashimoto H, Enjoji M. Angioleiomyoma. A clinicopathologic reappraisal of 562 cases. *Cancer* 1984; 54: 126-130.

### **Legend of figure**

*Figure 1* Low and high power magnifications of the excised lesion are shown in A and B respectively. The immunoreactivity of the oval/spindle cells for Smooth Muscle Actin is illustrated in C. The florid vasculature of the lesion is highlighted by CD34 immunostain (D). A and B: haematoxylin and eosin; bar in A: 200  $\mu\text{m}$ ; bar in B, C and D: 100  $\mu\text{m}$ ; *ep*: epidermis of the external auditory canal.