The Journal of International Advanced Otology

J Int Adv Otol 2019 • DOI: 10.5152/iao.2019.4743

Case Report

Leiomyoma of the External Auditory Canal

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Cite this article as: Tombolini M, Ralli M, Greco A, de Vincentiis M, Carletti R. Leiomyoma of the External Auditory Canal. J Int Adv Otol 2019; DOI: 10.5152/iao.2019.4743.

Angioleiomyomas (ALMAs) are benign tumors that arise from smooth muscle cells that form the tunica media of vascular structures. To our knowledge, only five cases of ALMA of the external auditory canal (EAC) have been reported. We report the case of a 52-year-old man with left otalgia, otorrhea, auricular fullness, and hearing loss. On otomicroscopic examination, a reddish and smooth mass completely occluding the EAC was seen. Computed tomography scan showed a soft tissue mass with a complete opacification of the left EAC. A possible diagnosis of ALMA should be considered when finding a tumor inside the external ear canal. An accurate pathological examination should be performed to differentiate benign vascular ALMA from malignant leiomyosarcomas. Complete surgical excision is the treatment of choice in benign tumors.

KEYWORDS: Leiomyoma, angiomyoma, ear canal, head and neck neoplasms

INTRODUCTION

Leiomyomas (LMAs) are benign tumors of smooth muscle phenotype. Angioleiomyomas (ALMAs) emerge from the smooth muscle cells that form the tunica media of vascular structures. LMAs are common in the uterus and in the skin according to different studies. LMAs of the head and neck regions are uncommon and usually affect the esophagus, skin, turbinates, oral cavity, paranasal sinuses, larynx, and pharynx [1].

Leiomyomas represent only 1.3% of the soft tissue tumors in the head and neck and 0.16% of them occur in the auricle [2]. In the head and neck, there is a peak in the sixth decade of life and a female predilection, with a ratio of 3.5:1 [3].

The occurrence of LMA in the external auditory canal (EAC) is extremely rare. To the best of our knowledge, only five previous cases have been described in the literature [4-8]. In this case report, we describe a case of the EAC and review the current literature.

CASE PRESENTATION

A 52-year-old man was transferred to our university hospital from an otorhinolaryngology ward of a smaller hospital with chief complaints of left otalgia and otorrhea since 1 week and a 3-month history of ipsilateral hearing loss. After admission, the patient underwent intravenous antibiotic therapy and endoauricular antibiotic drops with ready regression of ear pain and discharge. Written informed consent was obtained from the patient for all the diagnostic and therapeutic procedures performed.

On otomicroscopic examination, a reddish and smooth non-bleeding mass completely occluding the left EAC was seen. Cervical lymph nodes were not palpable. A complete blood cell count revealed no abnormalities.

Weber tuning fork test was lateralized to the left ear, whereas ipsilateral Rinne test was negative. Pure tone audiometry (PTA) was recorded (Audiometer AA222; Interacoustics, Middelfart, Denmark) according to the principles of the American Academy of Ophthalmology and Otolaryngology and showed a mild conductive hearing loss in the left ear mainly affecting low and middle frequencies with an average threshold of 26.40 dB HL in the 125-8000 Hz frequency range and of 31.25 dB HL in the 250-1000 Hz range. Hearing was normal on the right ear (average PTA threshold: 18.20 dB HL).

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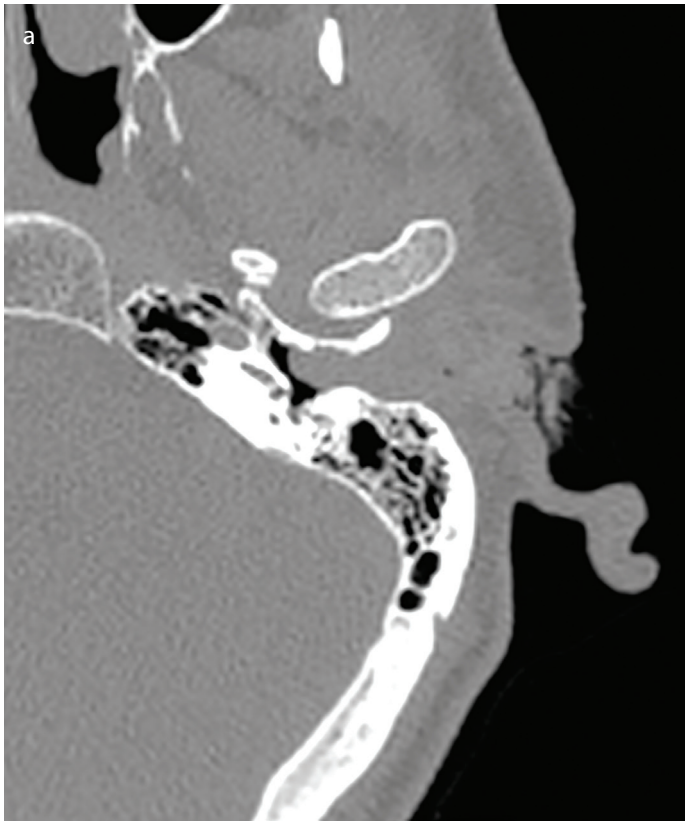
Submitted: 27.10.2017 • **Revision Received:** 02.02.2018 • **Accepted:** 29.10.2018 • **Available Online Date**: 15.03.2019



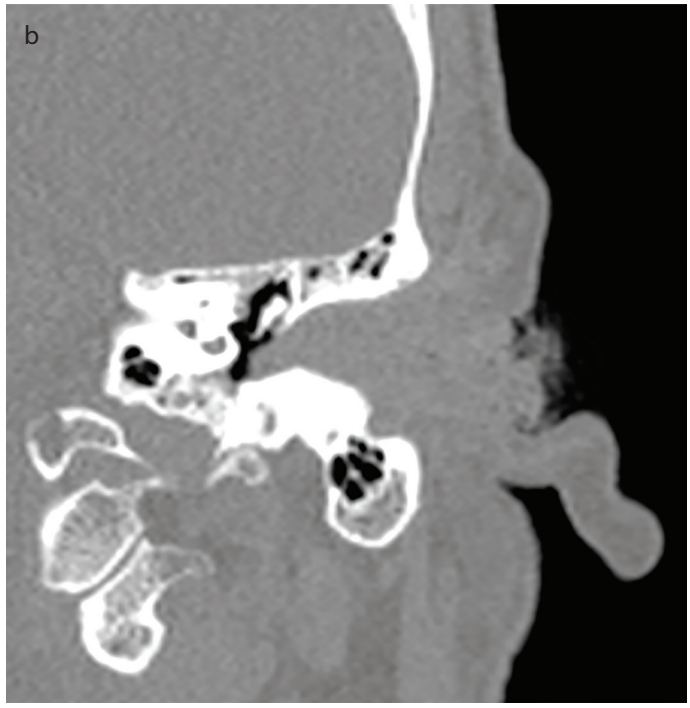


Figure 1. a, b. Non-enhanced preoperative computed tomography scan in the axial (a) and coronal (b) planes showing a soft tissue mass with a complete opacification of the left external auditory canal, without erosion of the bone or abnormalities of the middle or inner ear.

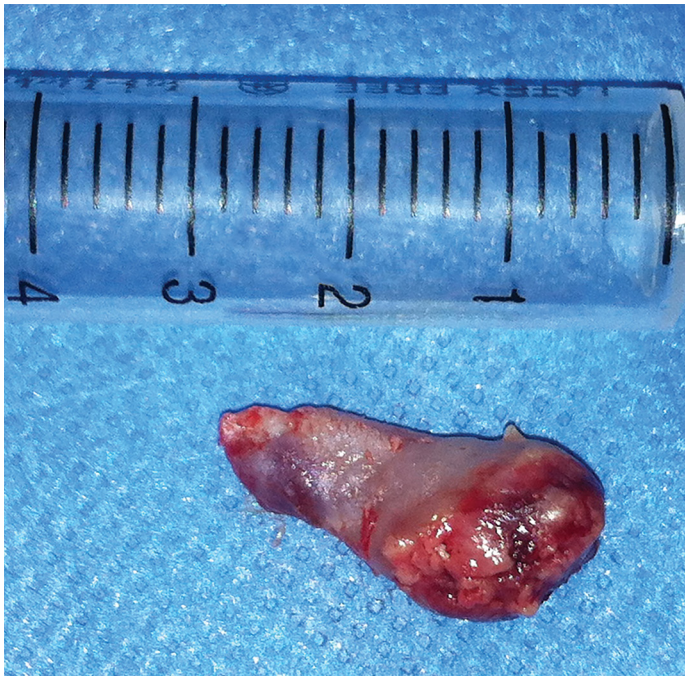


Figure 2. The tumor removed from the left external auditory canal showing its polypoid conformation.

Computed tomography scan showed a soft tissue mass with a complete opacification of the left EAC without erosion of the bone or abnormalities of the middle or inner ear. The left mastoid was well developed and without evidence of bony erosion or opacification (Figure 1).

The mass was surgically removed en bloc under microscopic control through an endaural approach under general anesthesia. Skin graft was not required. The specimen measured 1.6×0.9×0.6 cm and showed a polypoid conformation (Figure 2).

Histological examination of the mass showed fibrous tissue lined by the malpighian epithelium with predominantly orthokeratotic hyperkeratosis. In this context, there was a proliferation of cells, mostly spindle-shaped cells (positive for smooth muscle actin), organized in vortical bundles, in combination with many vascular structures (Figure 3). These structures were coated with the endothelium only focally activated and did not show significant alterations. There was no mitosis or necrosis observed.

The surface of the tumor showed focally intense neutrophilic exocytosis of the epithelium with necrosis associated with granulation and inflammatory tissue; necrosis was not seen elsewhere within the neoplasm. These findings were consistent with a diagnosis of ALMA.

There were no complications in the postoperative course, and the patient's hearing fully recovered after removal of the packing from the auditory canal. The postoperative otomicroscopy confirmed an intact eardrum with a well-healed EAC. A PTA recorded after the surgery showed an average hearing threshold in the left ear of 19.55 dB HL in the 125-8000 Hz frequency range and 15 dB HL in the 250-1000 Hz range with no residual interaural asymmetry. Preoperative PTA and postoperative PTA are shown in Figure 4. There was no sign of local recurrence after 18 months of follow-up.

DISCUSSION

Primitive tumors of the EAC are infrequent. Malignant tumors are the most frequent tumors. Adenoid cystic carcinomas, ceruminous ade-

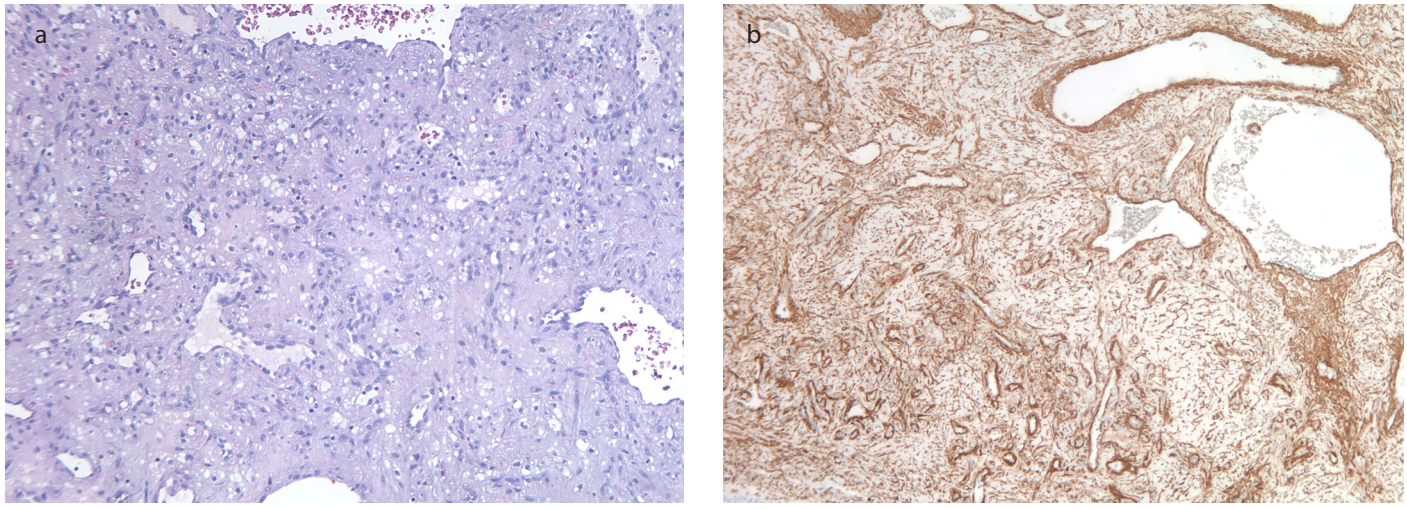


Figure 3. a, b. Leiomyoma shows a proliferation of cells (a), mostly spindle-shaped cells, organized in vortical bundles, in combination with many vascular structures (hematoxylin and eosin stain, photomicrograph, original magnification ×100). The second photomicrograph (b) shows leiomyoma expression of SMA protein by IHC in formalin-fixed paraffin-embedded tissue (IHC for SMA, photomicrograph, original magnification ×50).
IHC: immunohistochemistry; SMA: smooth muscle α-actin

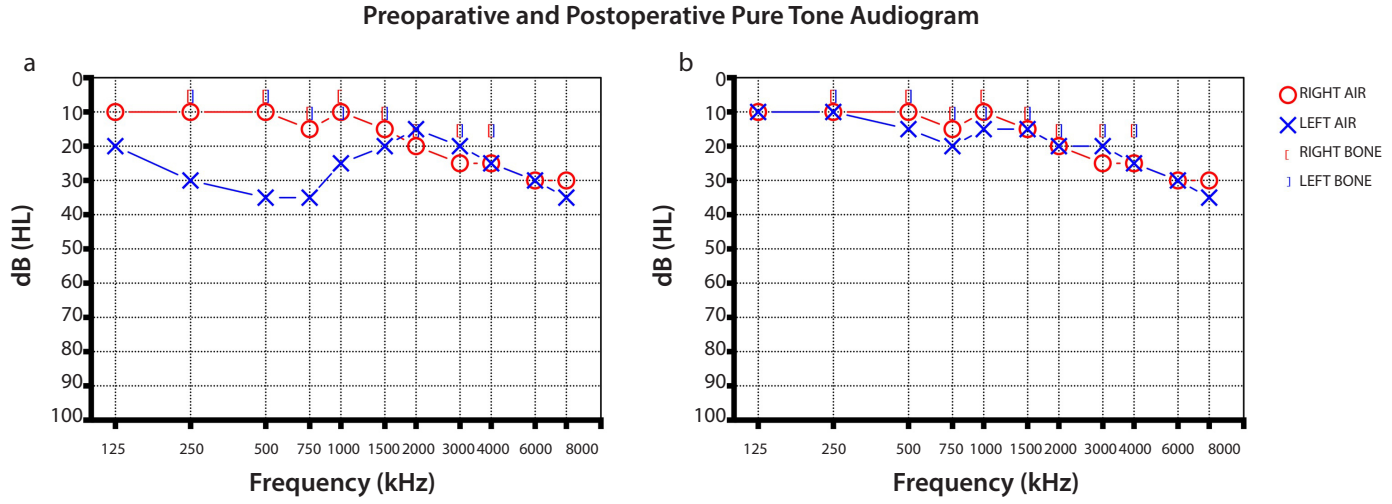


Figure 4. a, b. Preoperative and postoperative PTA. (a) Preoperative PTA shows a mild conductive hearing loss in the left ear with an average threshold of 26.40 dB HL in 125-8000 Hz frequency range. (b) Postoperative PTA in the left ear is within the normal range (19.55 dB HL). Normal hearing is found in the right ear (average PTA threshold: 18.20 dB HL).
PTA: pure tone audiometry

nocarcinomas, mucoepidermoid tumors, myoepithelial carcinomas, epidermoid carcinomas, basal cell carcinomas, and different types of sarcomas are among these.

The majority of benign tumors of the EAC develop from ceruminous glands as pleomorphic and ceruminous adenomas. Other benign tumors include hemangiomas, schwannomas, and plexiform neuromas.

To our knowledge, only five cases of LMA of the EAC are reported in the literature [4-8]. Most ALMAs present as a small, slowly enlarging mass usually of several years of duration. Pain is the most common subjective complaint in approximately half of patients with ALMA. Pain is exacerbated by wind, cold, pressure, pregnancy, or menses in some patients [4]. In our case, there was also an infection that was caused by a superimposed severe external otitis that promptly regressed with antibiotic treatment.

The mean age of onset of ALMA of the EAC is 46.5 (37-56) years. Men appear to be more affected than women with a 2:1 ratio, whereas it is the opposite for LMA of the uterus, skin, and esophagus.

A possible relationship of LMA with trauma has been previously described; however, in our case, there was no history of trauma in the previous years [6].

Histologically, LMAs are composed of mature and well-differentiated smooth muscle cells. Mitotic figures are usually absent or very rare.

Different classifications for LMA can be found in the literature. LMA is divided into two types. Tumors that originate from the arrectores pilorum muscles are called pilar LMA and usually occur in the skin, whereas those that derive from the muscular coats of blood vessels are called ALMAs and usually occur in smooth muscles of the lower extremities [5]. Morimoto et al. [9] also divided the ALMA into

three histological types: solid, venous, and cavernous. In the solid type, smooth muscle bundles are closely compacted, and vascular channels are many but usually small and slit-like. In the venous type, ALMAs have vascular channels with thick muscular walls and less compacted smooth muscle bundles. In the cavernous type, tumors are composed of dilated vascular channels with small amounts of smooth muscle. The World Health Organization Classification of Tumors also subdivided LMA of the uterus into four different types: cellular, epithelioid, myxoid, and atypical [10].

When conducting immunohistochemistry research, most cells of ALMAs are positive for alpha-smooth muscle actin, desmin, vimentin, and type IV collagen [4]. Mitotic activity is used to differentiate benign vascular LMA from malignant leiomyosarcomas. If there are no mitoses in ≥50 high-power fields, the lesion is benign. However, when there are one or more mitoses in every five high-power fields, the lesion is probably malignant and is certainly malignant when mitoses are found in every high-power field. In our case, there was no mitosis or necrosis, and the diagnosis was benign LMA [5].

The proper treatment for LMA is complete surgical excision. We used an endaural approach, which was also used by Iguchi et al. [5] and De Luca et al. [8], whereas other authors used a retroauricular approach, and in one case, this was combined with mastoidectomy [4-8].

CONCLUSION

Angioleiomyomas arising in the EAC are exceptionally rare. A possible diagnosis of ALMA should be considered when finding a tumor inside the external ear canal of adult male patients. Complete surgical excision is the treatment of choice, and an accurate pathological examination should be performed to eliminate any possibility of malignancy.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – M.T.; Supervision – M.d.V.; Materials - M.R.; Data Collection and/or Processing – R.C; Literature Search – A.G.; Writing – M.R.; Critical Reviews – A.G.

Conflict of Interest: The authors have no conflict of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

REFERENCES

1. Barnes L. Tumors and tumor like lesions of soft tissue. Surgical pathology of the head and neck. 2nd ed. New York: Marcel Dekker; 2001. p. 912-5.
2. Makino Y. A clinicopathological study on soft tissue tumors of the head and neck. Acta Pathol Jpn 1979; 29: 389-408. **[CrossRef]**
3. Barnes L. Pathology and Genetics of Head and Neck tumours. Lyon: IARC Press; 2005.
4. Petschenik AJ, Linstrom CJ, McCormick SA. Leiomyoma of the external auditory canal. Am J Otol 1996; 17: 133-6.
5. Iguchi H, Nakai Y, Yamane H, Goto K, Wakasa K. Smooth muscle tumor of the external auditory canal. Otolaryngol Head Neck Surg 1997; 116: 231-3. **[CrossRef]**
6. Yeh SY, Chao TK. Leiomyoma of the external auditory canal--a case report and brief review of the literature. Eur Arch Otorhinolaryngol 2005; 262: 397-9. **[CrossRef]**
7. Picciotti PM, Cantore I, La Greca C, Di Nardo W, Scarano E. Angioleiomyoma of the external auditory canal. Am J Otolaryngol 2007; 28: 235-7. **[CrossRef]**
8. De Luca LM, Karligkiotis A, Melis A, Pintus A, Trombetta M, Riu FG, et al. Angioleiomyoma of the External Auditory Canal. Otol Neurotol 2016; 37: e171-2. **[CrossRef]**
9. Morimoto N. Angiomyoma (vascular leiomyoma): a clinicopathological study. Med J Kagoshima Univ 1973; 54: 126-30.
10. Tavassoli F, Devilee P. Pathology and Genetics Tumours of the Breast and Female Genital Organs. Lyon: IARC Press; 2003.