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Case Report
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Middle Ear Leiomyoma Presenting as Granulomatous Otitis Media

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Abstract

Leiomyoma is a benign smooth muscle tumor of metastatic potential. In this paper we present the second reported case of leiomyoma in the middle ear cleft in a 59-year old Saudi lady who presented with complaint of chronic left ear discharge for 15 years. Computed tomography scan revealed soft tissue density with calcification in the epitympanum and mastoid antrum without bony erosion. The mass was successfully removed by canal wall up mastoidectomy approach.Theoriginof leiomyoma is likely to be the smooth muscle of the vasculature of a middle ear cleft, which is devoid of other smooth muscles.

Keywords: Leiomyoma; Benign smooth muscle tumor; Benign spindle cell proliferation; Granulomatous chronic otitis media; Ear exploration

Abbrevations: SDS: Speech Discrimination Threshold; PTA: Pure Tone Audiometry; SMA: Smooth Muscle Actin; MSA: Muscle Specific Actin; GFAP: Glial Fibrillary Acidic Protein

Introduction

Leiomyoma is a benign smooth muscle tumor of metastatic potential. [1]. Leiomyoma rarely occurs in the head a neck area, with reported prevalence of nearly 1% in adults, and 2.5% in children, with female preponderance [2]. The most common site of leiomyoma in the head and neck region is the lips (27.46%) followed by tongue (18.30%), cheeks and palate (15.49%), gingiva (8.45%) and mandible (5.63%) [2]. In the temporal bone the external ear is the commonest site of leimyoma [3-6,7], followed by inner ear [8-10]. Here we present the first reported case was vascular leiomyoma encasing the geniculate ganglion and the associated segments of facial nerve [11].

Case Report

This is a 59 year old Saudi lady, Arab ethnic, who is a known case of untreated allergic rhinitis, bronchial asthma for 20 years, mixed anxiety depression disorder for 5 years, lumbar spondylosis, polyarthropathy with bilateral carpal tunnel syndrome, and gastritis. She is not known to have diabetes mellitus, hypertension, or other chronic disease of note. She presented with complaint of intermittent yellowish left ear discharge mixed with blood for 15 years, progressive reduction in hearing of left ear and non-pulsatile tinnitus in left ear for 15 years, and a few attacks of rotational vertigo 6 months back, lasted for a few minutes provoked by standing. No otalgia.

The patient had a past history of lower section Cesarean-section four times, and repairs of para-umbilical incisional hernias on 2 occasions. She also had strangulated viable bowel secondary to adhesions with which required adhesiolysis and omentectomy.

She is non-smoker and has no history of alcohol intake. There is no family history of malignant disease.

On examination, she presents as well groomed elderly women with clear anxiety. She was vitally stable. The right ear examination showed dull retracted tympanic membrane and healthy external canal. The left tympanic membrane had small central wet perforation in the center of pars tensa with rolled edges. Middle ear mucosa was not visible owning to the small size of perforation. The external canal appeared healthy. No granulation, or keratin debris were present. The secretions are faint yellow-stained and odorless (there was large anterior hump obstructing the view of anterior part of tympanic membrane). Post-auricular area was normal. Nose examination revealed pale mucosa, watery secretions, and moderate hypertrophy of inferior turbinates. Septum was central. Throat was congested. Neck examination showed no cervical lymphadenopathy and facial nerve was intact. Cardiac, chest, abdomen, and lower limbs examinations did not reveal significant findings.

Medications history include Paroxetine 25mg PO OD, Seretide (fluticasone propionate/salmeterol xinafoate) inhaler 2 puffs inhaled BID, Ventolin (salbutamol) inhaler 2 puffs inhaled PRN, and ranitidine 150mg BID.

Blood investigations showed white blood cells WBC of 5.48 x 10^9/L, Hemoglobin Hb 12.7 g/L, fasting glucose was 92 g/dL, corrected calcium 9.0 mg/dL, alkaline phosphatase ALP 69 iu/L, CK 104 iu/L, lactate dehydrogenase LDH 201 iu/L.Pure tone audiometry (PTA) of the left ear showed moderate mixed hearing loss with excellent speech discrimination threshold (SDS). Pure tone average was 51.3 dB. Right ear PTA and tympanogram were normal. Dix-Hallpike maneuver was negative.

Computed tomography pre-contrast showed soft tissue density with small calcification in left epitympanum and mastoid antrum (Figure 1). Left mastoid air cells were sclerotic. Scutum and ossicles are intact. No associated bony erosion. Normal bilateral cerebropontine angles and internal auditory meatus were found with no evidence of focal lesion or post-contrast enhancement. Nose and paranasal sinuses are within normal. Compared to the earlier conventional tomography scan done in September 2009, the current computed tomography scan showed no interval changes.

After seven years of declining surgery because of fears and anxieties, on 22 December 2014, the patient finally underwent left ear exploration by canal wall up and atticotomy approaches; mastoid antrum and middle ear were filled with unhealthy looking abnormal, granulation tissue, which was engulfing the ossicles. The long process of incus was necrosed. The middle ear mucosa was polypoidal. Middle ear and mastoid granulation tissue was removed completely along with malleus and incus. Dexamethasone-impregnated gelfoam was applied to the middle ear cavity. Silk was applied lateral to the tympanic membrane to facilitate healing. Postoperatively, patient was fit for home discharge. Histopathological examinations revealed benign spindle cell proliferation (immunological studies revealed intensely positive for smooth muscle actin (SMA), muscle specificactin (MSA), desmin, and caldosmin and negative forCD34,S100 and Glial Fibrillary Acidic Protein (GFAP)) consistent with leiomyoma (Figure 2).

Patient was reviewed at 2, and 4 weeks in outpatient clinic. She complained of mild left facial weakness and autophony. Silk covering the tympanic membrane was removed. Tympanic membrane was of healthy appearance. There was grade 1 facial weakness, which markedly improved on subsequent visits. Autophony greatly reduced. Pure tone audiometry showed no interval changes postoperatively.

Pelvic ultrasound was ordered to rule out uterine leiomyoma of metastatic nature; there was no evidence of uterine leiomyoma seen.

Discussion

Smooth muscle tumors of the head and neck region are uncommon. They are reported more often in the nasal cavity and paranasal sinuses, pharynx, oral cavity, and auricle. The great majority of soft tissues tumors in the head and neck region are of benign nature (96%) [5]. The primary site of occurrence of leiomyoma in the body is the uterus (95%), followed by skin (3%) then gastrointestinal tract (1.5%) [2]. Benign smooth muscle tumors arising from the middle ear are very rare and this case represents the second reported case of leiomyoma in the English language literature to the best of our knowledge. The first case(reported in 2013)in which vascular leiomyoma was encasing the geniculate ganglion and the adjacent segments of facial nerve, was completely removed by canal wall up approach without injury to the facial nerve [11]. In the presenting case, leiomyoma was completely removed by canal wall up approach without evidence of recurrence during follow up.

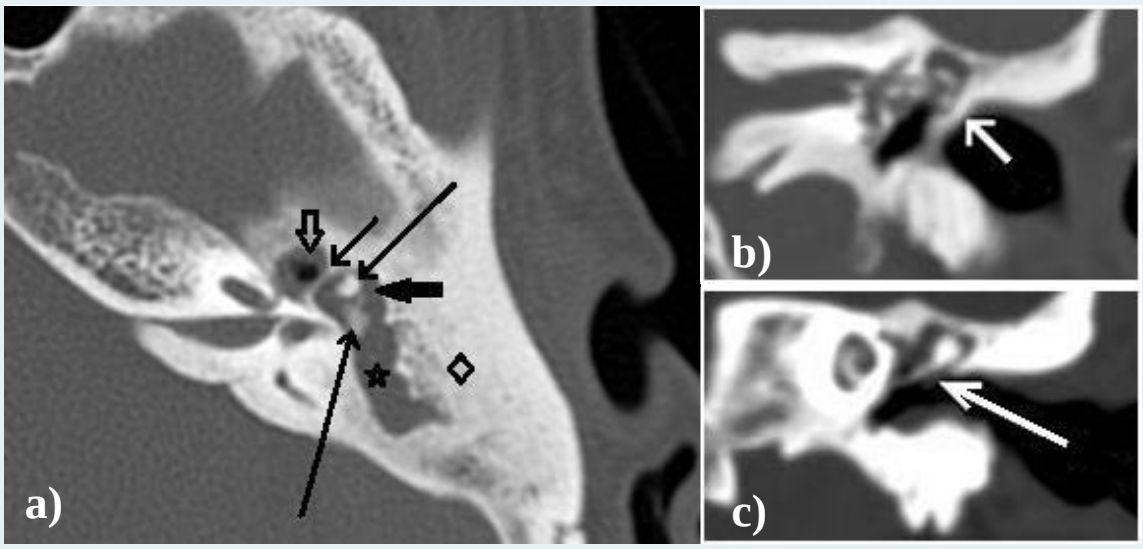

Figure 1: A. Axial Computed Tomography of the left middle ear demonstrating obliterative soft tissue density originating from left mastoid antrum [hollow star], travelling via aditus to the middle ear posterior epitympanum [solid short arrow] and completely obliterating it. This soft tissue density is partially delimited by the cog [short thin arrow] anteriorlyresulting in only partial obliteration of the anterior attic [hollow short arrow]. Note the sclerotic mastoid [hollow diamond], intact head of malleus [intermediate thin arrow] and intact body and short process of incus [long thin arrow]. B. and C. Coronal views showing intact scutum [short white arrow] and intact body and long process of incus [long white arrow].

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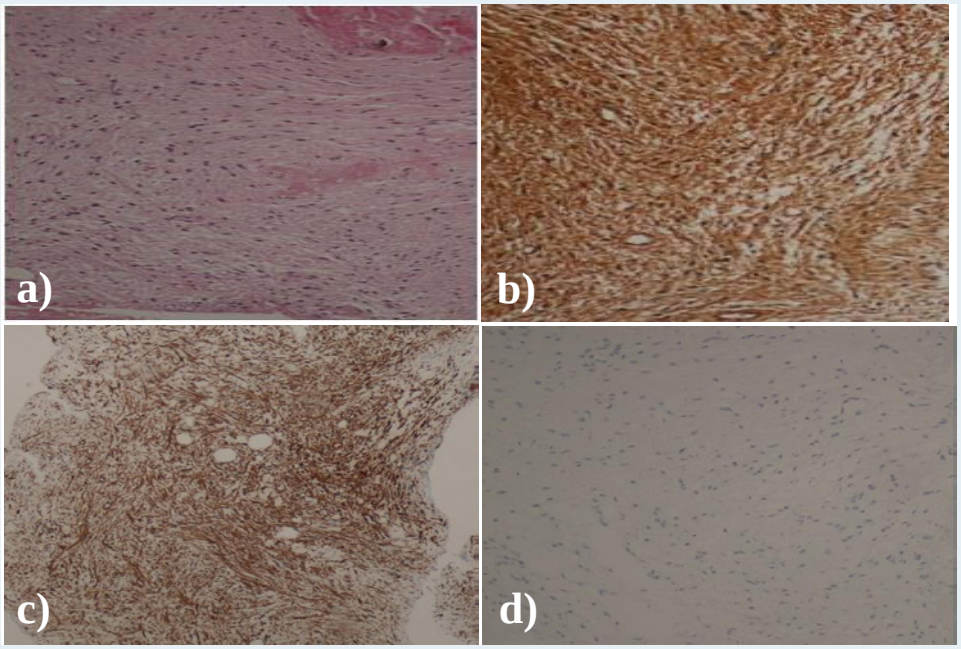

Figure 2: Photomicrographic panel of middle ear mass. A. Hematoxylin and eosin stained section (intermediate magnification) showing well circumscribed soft tissue mass containing spindle shaped lesional cells which pose bluntended nuclei, moderate volume of cytoplasm and indistinct outline. There was no evidence of mitosis or necrosis. A blood vessel closely related to the lesional cells is demonstrated in the right upper corner of photograph]. B. Immunohistochemical stain for smooth muscle actin (SMA) demonistrate diffuse strong reactivity. C. Immunohistochemical stain for muscle specific actin (SMA) demonistrate diffuse strong reactivity. **D.** Immuno histochemical stain for S100 demonstrates negative reactivity.

Leiomyoma is classified by the World Health Organization (1969) into three main variants; solid (conventional), vascular (angioleiomyoma) and epithelioid (leiomyoblastoma) [2]. The most common variants of leiomyoma in the head and neck region are either the solid (71%) or vascular types originating from smooth muscles of the vasculature (27%), followed by epithelialoid type (1.2%) and mesectodermal type (0.8%) [12,13].

Vascular leiomyoma was sub-classified by Mariamoto in 1973 into capillary (solid type, 66%), venous (23%), and cavernous (11%). Vascular leiomyoma are predominantly found in the lungs [14], with venous type being the commonest in the head and neck region [2,14]. In a review of 562 cases of vascular leiomyoma over a 17-year period, only 48 were seen in the head and neck [14]. In 1985, Barnes conducted literature review to identify 257 cases of leiomyoma in the Head and Neck and he found that 92 cases were associated with cervical esophagus, 58 with skin including skin of ear, 52 with oral cavity, 22 with larynx, 12 with eye and orbit, 8 with nose and paranasal sinuses, and the remainder are associated with trachea, salivary glands, thyroid, bone of jaws, soft tissue of the neck, hypopharynx, wall of thyroglossal duct cyst [12]. The majority of the reported cases of leiomyoma in the temporal bone are vascular type [3-6,8-10,15]. In the presenting case the leiomyoma originated from the smooth muscle of the vasculature of the middle ear, as there is no other source of smooth muscles in the middle ear cleft.

Leiomyoma of vascular origin used to be known as angiomyoma, and angioleiomyoma. However, vascular leiomyoma is the most wide accepted name given that it more descriptive of the nature of the tumor [14].

Histopathological appearance of leiomyoma is characterized by bundles of intersecting elongated spindle shaped cells, owing to its smooth muscles origin [2], together with perinuclear vacuoles and eosinophilic cytoplasm [13,15]. Secondary degenerative changes of leiomyoma, such as edema, hyalinization, hemorrhage, calcification, cystic degeneration, and rarely ossification, are uncommon [13]. Leiomyoma can usually be differentiated from most other spindle cell tumors by itspositive expression of smooth muscle markers (Smooth muscle actin SMA, muscle specific actin MSA and smooth muscle heavy chain myosin SMMS-1), vementin, and may rarely be positive for desmin [13]. Although leiomyoma is CD34 negative, in vascular leiomyoma is CD34 positive owing to the labeling of the endothelial cells of the vascular spaces [14]. Other spindle cell tumors that may express smooth muscle markers and contain calcifications include myofibroma,which contains a hybrid of smooth muscles and fibroblasts [13]. Masson trichrome stain and SMMS-1 help differentiate myofibroma from smooth muscle lesions in the head and neck region [13,17].

Active mitosis helps differentiate leiomyoma from leiomyosarcoma; the presence of one or more active mitoses in every five high-power fields, indicates probable malignant tumor, and the presence of mitoses in every high-power field, indicates certain malignancy [5]. If no active mitosis is seen, the tumor is certainly benign. The presence of atypia suggests malignant nature of tumor [16].

Leiomyoma has a potential to metastasize to distant sites,

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despite its benign nature and well-differentiated appearance. In 1939, Steiner proposed as first the contradictory term of benign metastasizing fibroleiomyoma [19]. There are just less than 100 reported cases of benign metastasizing leiomyoma in the English literature. Lung is known to be the most common metastatic site. In addition to lung, the extra uterine sites that these tumors can localize to include skin, pelvis, abdomen, muscle, greater omentum, inferior vena cava, right atrium, brain and bones [20]. Ultrasound scan of the pelvis did not show uterine leiomyoma in the presenting case, which rules out benign metastasizing leiomyoma to a great extent. Skeptics casted doubts about the existence of such benign metastases. Paley and Fornasier reviewed 10 cases of leiomyosarcoma with bone metastasis. In eight of the ten patients the primary tumor of the uterus was initially diagnosed as a leiomyoma but, after review, appeared to be an unrecognized low-grade leiomyosarcoma [1]. Therefore, if a primary tumor is found then careful histopathological re-examination is warranted.

Proposed theories to describe the origin of vascular leiomyoma include progression from aberrant undifferentiated mesenchyme, progression from vascular malformation, and neoplastic proliferation of smooth muscles of the walls of the vasculature [14]. Provocative factors for disease are theorized to include trauma, steroid therapy, and hormonal imbalance [18].

Leiomyoma usually presents as a slowly growing small spherical mass, which can be painful in solid type and painless in venous and cavernous types, and causes no complications [20,21]. In the presenting case the patient had ear discharge for 15 years before consenting to undergo surgery, and no complications from the disease were observed.

The chief differential diagnosis of leiomyoma in the middle ear is granulomatous otitis media. Besides granulomatous otitis externa differential diagnosis include cholesterol granuloma, cholesteatoma, calcified vascular malformation, soft tissue osteoma, schwannoma, neurofibroma of facial nerve, paraganglioma, hemangioma, vascular malformation, fibrous dysplasia, myositis ossificans, meningoencephalocoele, extracrainal meningioma, lymphatic malformation, adenoma (includes epithelial and neuroendocrine carcnoid types), myospherulosis, endolymphatic sac tumor, schneiderian-type mucosa papilloma (includesfungiform, inverted, and oncocytic types), choristoma, Langerhans cell histocytosis (Eosinophilic granuloma type), angiofibroma, angiomyolipoma, and angiomyosarcoma.

Computed tomography of leiomyoma demonstrates non-specific appearance of well-defined homogenously enhanced mass [22].

Proper treatment of leiomyoma is complete extirpation [5,15]. This will also help establish histopathological diagnosis if the diagnosis is uncertain. The surgical approach varies according to the site involved and extent of tumor. In this case typical middle ear exploration by classic canal wall up approach was employed to extirpate the tumor.

Recurrence of leiomyoma is very rare even with positive resection margins. Billings et al. [22] had only one recurrence out of 36 operated cases, 10 of which had positive resection margins. While Yoon et al. [20] had no recurrence in any of 12 head and neck cases after mean follow up of 52 months.

Conclusion

In conclusion, although leiomyoma of temporal bone is very rare benign tumor, it should be considered in the differential diagnosis of mass lesions in patients with risk factors. Surgical extirpation appears to carry an excellent prognosis.

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