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Case reports article – Rapidly growing mass in the ear canal:
A rare case of isolated superficial angiomyxoma
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Abstract:
Superficial angiomyxoma is a rare benign skin tumour and usually present as an asymptomatic lesion. It is known to be highly recurrent and complete wide excision is the best treatment. This case reported of 56 yr old gentleman presented with fleshy, pink coloured right external auditory mass. The mass arisen from the posterior wall of the canal at the bony–cartilaginous junction. Clinical diagnosis of an anaural polyp was made and the mass was excised. However, it rapidly recurred bigger than its actual size within a week. The histopathological examination was reported as superficial angiomyxoma. A differential diagnosis of isolated superficial angiomyxoma should be considered in the case of external auditory canal mass and Carney complex needs to be considered. Complete excision and regular follow up is recommended.

Keywords: Benign tumour, external auditory canal, myxoma

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Introduction
Superficial angiomyxoma (SA) is a rare cutaneous, benign lesion that is still poorly recognized and less in number of cases(1). SA can be present either as solitary or multiple lesions. External auditory canal (EAC) is a rare site for SA and rarely reported in the literature. Furthermore, an occurrence of SA in the EAC is usually having multiple lesions and associated with Carney’s complex (2). We report a rare case of isolated superficial angiomyxoma in the EAC.

Result - Case History
A 56 yr old Malay gentleman presented to Otorhinolaryngology (ORL) outpatient department with painless right ear fullness and reduced hearing for 2 wk. Otherwise, he denied a history of ear discharge, bleeding, tinnitus, vertigo, trauma or frequently used of a cotton bud or metal ear stick to clean his ears. Nor was there any other skin lesion, neoplasm or history of endocrine abnormalities. Examination under microscope revealed a pink fleshy mass filled part of the External Auditory Canal (EAC). The mass was arising from the bony cartilaginous junction of the posterior wall of EAC. No pus discharge or whitish debris seen. Part of the tympanic membrane was visualized beyond the mass and was unremarkable. A clinical diagnosis of an aural polyp was made and polypectomy was performed. The EAC was packed using ear wick mixed with framycetin sulfate and gramicidin eardrops. During followed up one week post polypectomy, the mass rapidly recurred, bigger than its actual size and totally obstructed the ear canal [figure 1]. Wide local excision with an adequate margin surround its base was performed under local anaesthesia. Following total excision, Trichloro Acetic Acid (TCA) was applied at the base area. On subsequent followed up and at the time of writing, no evidence of recurrence and the skin was healthy [figure 2].
Figure 1 Otoscopic view of a mass one week following the initial polypectomy

Figure 2 Endoscopic finding of the surgical site at two weeks post excision

Histopathological examination (HPE) and its features of the mass is shown in figure 3 and figure 4. Immunohistochemistry study showed positive to Vimentin and smooth muscle actin (SMA) with features keeping consistent with superficial angiomyxoma.

Figure 3 HPE showing stellate and spindle shape tumour cells in the abundant myxoid background with scattered neutrophils (Hematoxylin and eosin stain 10 ×10)
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**Figure 4** Low power view of HPE showing tumour tissue composed of spindle cells in the background of myxoid stroma and scattered congested blood vessels (Hematoxylin and eosin stain 4 x10)

**Discussion**

Superficial angiomyxoma was first described by Carney et al. in 1985 as a cutaneous myxoma of Carney’s complex (2). Carney’s complex is a familial disorder transmitted as an autosomal dominant trait: It includes myxomas, spotty pigmentation, endocrine overactivity and schwannomas. In this case patient was presented with solitary mass at external ear canal without any syndromic components associated with Carney’s complex. Several cases was reported to have solitary SA without association with Carney’s complex which occurred at parotid, inguinal and toe (3). The term SA was used to differentiate it from aggressive angiomyxoma. Aggressive angiomyxomas differ from the superficial angiomyxomas by the locations which is more deeper and occurring in the genital, pelvic and perineal regions as single and larger nodules (4). For the prevalence, SA has slightly higher prevalence in the male of about 56 % and the age of patient ranged from 12 yr to 82 yr with a mean age of 36.45 yr (4). Patient with SA usually presents with asymptomatic fleshy and pink coloured lesion mainly affecting the trunk, lower limbs, head, and neck regions. However, in this patient, the lesion occurred in isolation at a very rare site, within the EAC. There are several differential diagnoses for EAC mass according to its characteristics and sites of the lesions.

The commonest is a simple granulation tissue or aural polyps that occur due to repeated irritation or infection. In the majority of cases, aural polyps present with features of chronic supplicative otitis media, arising both in tubotympanic and atticoantral disease. Osteoma of the external ear canal can also be considered for its common presentation as a single and a unilateral mass. However, osteoma is usually attached to the tympanosquamous or tympanomastoid suture and hard in consistency. The other possible differential diagnosis could be canal cholesteatoma. It is usually presented as unilateral mass with a dull ache and chronic foul smelly ear discharge. Ear canal malignancy such as squamous cell carcinoma (SCC) also needs to be ruled out in this case in view of rapid recurrence immediately following the first excision and the patient’s age factor. However, in SCC of external ear canal patients usually presented with canal mass and persistent blood stained ear discharge. For investigations, imaging plays a minor role in the diagnosis, since there are no characteristic imaging features, although avid contrast enhancement will occur due to its vascular nature. Even though it is rarely been mentioned in common otorhinolaryngology textbooks or literature, otorhinolaryngologist should be aware of this disease entity and further diagnostic evaluation may be necessary after surgical excision.

Histological examination is essential for confirmation of the lesion. Histologically, SA is characterized by a well–defined border with subcutaneous involvement, reticulin network, mucinous stroma, and stellate to spindle shaped cells. It also commonly presents with prominent small thin walled blood vessels and stromal inflammatory cells particularly neutrophils. Clinically it has a tendency to locally recur. Immunohistologically the stromal cells are positive for vimentin but negative for S–100, SMA and desmin (5). It differs from aggressive angiomyxoma by the presence of thick or dilated blood vessels and immunopositive for desmin, smooth muscle actin, muscle–specific actin, vimentin, CD 34, estrogen, and progestin receptors in immunohistochemical analyses (6). As for this patient, HPE findings present all the characteristics mentioned above for the diagnosis of superficial angiomyxoma. The main treatment for SA is complete wide excision due to its high tendency of local recurrence. In view of its behaviour, complete excision and long term follow–up are recommended to all patients. Rodriguez et al reported that local recurrence of the tumour was estimated about 30 % to 40 % when associated with in–adequate resection (4).
Conclusions
In conclusion, isolated SA of the external ear canal is a rare diagnosis for benign external ear canal mass. Otorhinolaryngologist should be aware of the disease and to be considered as a differential diagnosis of EAC mass, especially in the presence of multiple myxomas in carney complex, to prevent further morbidity and recurrence. Complete excision and regular follow up is recommended for disease surveillance.

References