More to what meets the eyes: Myopericytoma of the preauricular sinus

Azlina Ab Rania, Goh Bee See, Nordashima Abdul Shukur and Noor‘Ain Mohd Nasir

aDepartment of Otorhinolaryngology- Head & Neck Surgery, Universiti Kebangsaan Malaysia Medical Centre, Jalan Yaacob Latif, Kuala Lumpur, Malaysia; bDepartment of Pathology, Universiti Kebangsaan Malaysia Medical Centre, Jalan Yaacob Latif, Kuala Lumpur, Malaysia

ABSTRACT

Preauricular lesions such as cysts, pits, fissures are congenital malformations of the preauricular soft tissues. These lesions are common and seen in the paediatric population, located near the anterior aspect of the ear. Surgical excision of the lesion would usually be performed if the lesion is infected to avoid multiple episodes of infection. We report a case of a 9-year-old boy who presented with a lesion of the preauricular area. It was excised due to enlargement of the lesion over time. Histopathological examination revealed the lesion to be a rare myopericytoma of the preauricular sinus. In this report, we discuss regarding this rare soft tissue lesion with regards to the head and neck region.

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Introduction

Preauricular lesions may either be sinuses, cysts or fissures, found lateral and superior to the facial nerve and parotid gland. While some may be asymptomatic for life, recurrent infections may occur in cases where there is presence of bacteria sealed within the tract, the opening of the sinuses or the cyst. Myopericytoma is a benign tumor that is composed of myoid-appearing oval to spindle-shaped cells with a concentric perivascular pattern of growth and is rarely seen in the head and neck region. In this report, we describe a case of myopericytoma occurring in the head and neck skin region with involvement of the preauricular region, where it is known to occur very rarely.

Case report

A 9-year-old Malay boy presented to the otorhinolaryngology clinic with a history of left preauricular swelling since the age of 1 year old. The swelling gradually increased in size with age, however was not associated with pain, neither history of infection. Examination revealed a swelling at the preauricular area with bluish discoloration. It is soft in consistency and well circumscribed. The swelling was not blanchable, not pulsatile, non tender with no pustule present. There was no other swelling nor any neck nodes palpable. Patient underwent ultrasound which revealed a well-defined homogenously hypoechoic lesion with posterior enhancement within the subcutaneous tissue at the left pre auricular region. There was some mixed vascularity noted within the lesion, predominantly seen at the periphery and also calcification noted within the lesion. Otherwise, there was no fatty hilum or evidence of hematoma nor pus seen. A clinical diagnosis of non-involuting congenital hemangioma was made.

Patient underwent excision of the left pre auricular swelling. Intraoperative finding showed superficial mass measuring 1 × 1 cm which was firm and rubbery with a well-defined margin. It appeared vascularized but there was no active bleeding. The mass was completely excised and sent for histopathological examination.

Macroscopically, the lesion appeared as a greyish tissue measuring 10×10mm. Microscopically, it showed fibrous tissue with lobules neoplastic cells surrounding slit-like and haemangipericytoma-like blood vessels. The neoplastic cells were fairly uniform, small, round to oval nuclei concentrating around the blood vessels (Figure 1). There were scattered blood vessels and lobules of neoplastic cells with proliferation of...
smooth muscle cells surrounding the blood vessels (Figure 2). The neoplastic cells showed to be diffusely positive for smooth muscle actin immunohistochemistry (Figure 3) and negative for CD 31 immunohistochemistry where it only highlights the blood vessels (Figure 4).

Patient was discharged well after one day postoperatively. He has been followed up regularly and there was no clinical features of recurrence after 3 years.

Discussion

Myopericytoma (MPC) is a term recently proposed to describe a group to tumours originating from perivascular myoid cells and show a range of histological growth patterns [1]. This rare entity was firstly described by Granter et al. in 1998 [2] where the cases reported had distinctive concentric perivascular proliferation of spindle cells and were located in the subcutaneous and superficial soft tissues of distal extremities. This was followed by a large case series of cutaneous myopericytoma [3] where Mentzel et al. noted that out of 54 cases reviewed, 26 were cutaneous myopericytomas, including 20 cases confined to the dermis and 6 showing subcutaneous extension. In this case report, a preauricular myopericytoma can be considered as a cutaneous lesion as it only confined to the skin.

Mentzel et al. further described this condition as to be reported in patients ranging from 13 to 87 years (median 52 years) and to most commonly affect lower extremities (48%), followed by the upper extremities (29.6%), the head and neck region (7.4%) and the trunk (3.7%) [3]. This condition was reported to have a male preponderance of 1.5:1 and although some lesions were painful, most were asymptomatic [4]. Our patient was 9 years old, younger than the average age of patients where this lesion was identified.

The incidence of myopericytoma in the head and neck region is rare and some cases in the maxillofacial region. A. Valasidis et al. reported two cases of MCP, a 9 year old with a left mandibular mental foramen and a 37 year old male with a lesion at the vermilion of the lower lip, both excised and no recurrence was noted during follow up [5]. Another recent study by Wu-Tong Ju et al who studied 5 new patients with MCP in the oral and maxillary region has shown all lesions to demonstrate a benign biologic behavior and no tumor recurrence [6]. Further study of this lesion within the head and neck region

Figure 1. Fibrous tissue with lobules of neoplastic cells surrounding slit-like and haemangispericytoma-like blood vessels. Inset: Neoplastic cells are fairly uniform, small, round to oval nuclei concentrating around the blood vessels.
Figure 2. Scattered blood vessels and lobules of neoplastic cells. Inset: Proliferation of smooth muscle cells surrounding the blood vessels.

Figure 3. Neoplastic cells are diffusely positive for smooth muscle actin immunohistochemical stain.
may be benefit in understanding the occurrence and behavior in the region.

Aung et al. further depict cutaneous myopericytomas to be benign in nature with a rare rate of recurrence [4]. These lesions may be identified microscopically when having hemangiopericytoma-like areas which are seen as numerous thin-walled, dilated with branching vessels, it may be angioleiomyoma-like when having perivascular bundles of elongated spindle-shaped cells and also may have occasional prominent atypia and mitotic activity and immunohistochemical staining revealed the lesion to be actin-positive and caldesmon-positive, as well as negative for other smooth muscle markers namely S100 protein, Mart-1/Melan A, EMA, CD31 and CD34 [4]. This corresponds well to the finding of our histopathological examination where the neoplastic cells are positive to actin immunohistochemistry, but negative to CD31 immunohistochemistry.

Treatment of a myopericytoma lesion is surgical excision and recurrence of this lesion is rare. Jung et al. reports a benign myopericytoma of the parotid gland which was treated successfully with superficial parotidectomy [7]. Our patient underwent excision of the preauricular sinus and has not shown any signs of recurrence upon follow up.

**Conclusion**

Even though commonly seen, preauricular sinus lesions must not be taken lightly. A thorough evaluation including imaging when necessary may help in achieving the diagnosis, and when subjected for excision, histopathological examination must be performed to ensure rare diseases are not missed.

**Informed consent**

The parents of the above patient have given consent for the patient to be included in this report. Noted that there is no identification of the patient, nor any images that can directly identify the above patient.

**Disclosure statement**

No potential conflict of interest was reported by the author(s).
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