

Myopericytoma of the external auditory canal and tragus

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Abstract

Background: Myopericytoma is a relatively recently described skin and soft tissue tumour that demonstrates perivascular myoid cell or pericytic differentiation. Whilst the range of anatomical locations has expanded to include visceral locations, head and neck myopericytomas are rarely documented. There have been no previous reports of aural myopericytoma.

Case report: This paper reports the clinicopathological features of a biopsy-proven, slow-growing, 20 × 20 mm, polypoid myopericytoma that involved the external auditory canal and tragus in an 18-year-old woman. Excision was curative.

Conclusion: Heightened clinicopathological awareness of the expanding anatomical distribution of myopericytoma is critical to its diagnosis when it presents in unusual and novel locations. Myopericytoma should be added to the range of external auditory canal neoplasms, especially those characterised by an admixture of spindle cells and a prominence of blood vessels, including those with a haemangiopericytomatous pattern.

Key words: Ear, External; External Auditory Canal; Tragus; Myopericytoma; Neoplasm; Pathology

Introduction

The term ‘myopericytoma’ was coined in 1998 to describe a benign neoplasm with a striking perivascular proliferation of spindle cells and differentiation towards perivascular myoid cells or pericytes.¹ Myopericytoma is closely related to the spectrum of tumours that demonstrate pericytic differentiation, including myofibromatosis, solitary myofibroma and infantile haemangiopericytoma. It most commonly develops in middle adulthood.² Myopericytomas are typified by a predilection for skin and soft tissue, and involvement of the extremities.³ However, the tumour is receiving increasing clinicopathological recognition as a distinct entity, which has resulted in an expanding, evolving anatomical distribution.² Myopericytomas have very occasionally been documented in the head and neck area.^{2,4–7} However, an extensive literature review using the PubMed search engine confirmed that aural involvement remains unreported in the English-language literature.

In documenting a polypoid myopericytoma of the external auditory canal and tragus, we aimed to heighten awareness of this tumour in the head and neck area. We propose that myopericytoma is added to the list of tumours that present with polypoid architecture in this location, and discuss the potential diagnostic pitfalls and mimicry.

Case report

Clinical features

An 18-year-old (human immunodeficiency virus negative) woman sought medical attention, for cosmetic reasons, for

a slow-growing mass that involved the left external auditory canal and tragus, which had developed over the previous 9 months (Figure 1a). The mass was not associated with pain, rapid growth, bleeding, or impedance to hearing or jaw movement. There were no other otorhinolaryngological complaints. The patient’s past medical, surgical and family history was non-contributory.

Examination revealed an otherwise healthy woman with a 20 × 20 mm, firm, smooth, well demarcated, non-tender, polypoid mass involving the lateral aspect of the left tragus, with no overlying skin changes. The findings of the otoscopic examination of the external auditory canal and tympanic membrane were normal. The Rinne tuning fork test (a clinical hearing test) was positive for frequencies 256, 512 and 1023 Hz bilaterally. There was no cervical lymphadenopathy or abnormalities of the skin and mucous membranes.

Incisional biopsy of the mass confirmed a spindle cell tumour with a myoid immunophenotype. Excision biopsy of the mass was conducted under general anaesthetic. The excision biopsy findings revealed incomplete excision margins. Wide local excision of the tumour site and left tragus was subsequently performed and tumour-free margins were confirmed.

The patient was well at follow up, with no recurrence at one, three or six months post-excision (Figure 1b).

Pathological features

On gross examination, the excised tumour measured 12 × 10 × 7 mm in size. It had a firm consistency and a grey-white appearance on cut section.

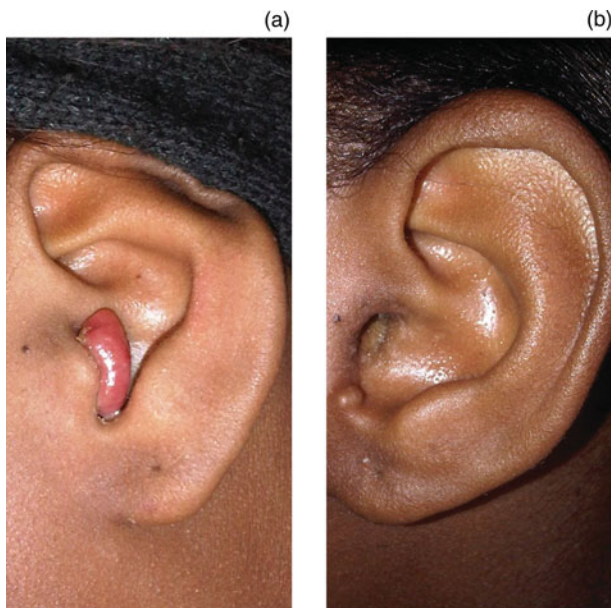


FIG. 1

(a) Pre-operative view of the patient showing a polypoid mass involving the external auditory canal and tragus, and (b) the post-operative view at six months follow up.

On microscopic assessment, the tumour demonstrated the presence of blood vessels of varying thickness, with a perivascular concentric arrangement of ovoid to plump spindle-shaped cells (Figure 2). In some fields, the spindle-shaped cells appeared to spin off the vessel walls. Thin-walled vessels, some with an antler-like, haemangiopericytomatous architecture, were also evident (Figure 2). The mitotic activity was less than 1 mitotic figure per 10 high-power fields. Cellular pleomorphism, tumoural necrosis and lymphovascular invasion were not seen. Admixed lymphocytes and plasma cells were noted. The tumour cells demonstrated muscle-specific actin (Figure 3a), alpha smooth muscle actin (Figure 3b), h-caldesmon immunopositivity (Figure 4a), desmin immunonegativity (Figure 4b) and Epstein–Barr virus-encoded RNA negativity. A diagnosis of myopericytoma was made.

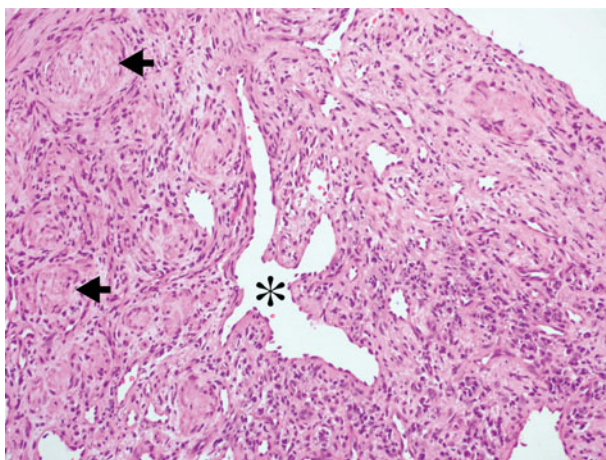


FIG. 2

Microscopic view of the vascularised spindle cell tumour, with stag-horn-type vessel architecture (asterisk) and vessels surrounded by whorls of plump myoid cells (arrows). (H&E; $\times 240$)

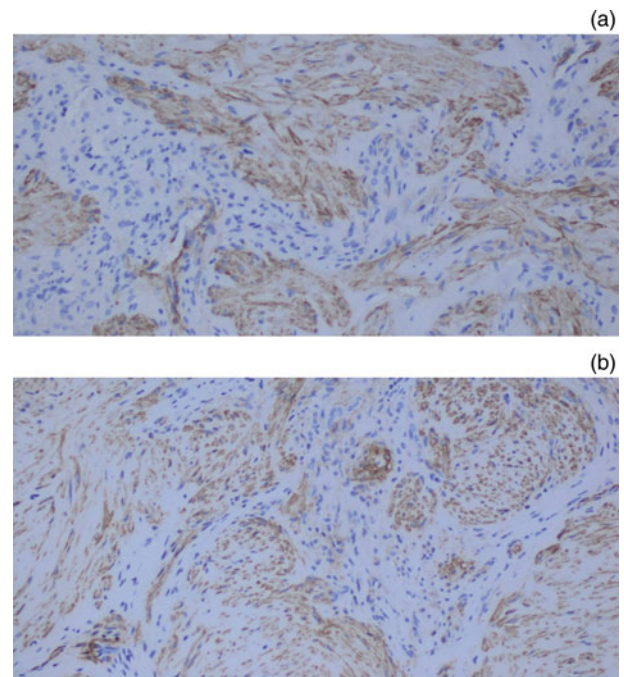


FIG. 3

Microscopic view of the tumour cells showing the immunohistochemistry staining for (a) muscle-specific actin and (b) alpha smooth muscle actin. ($\times 480$)

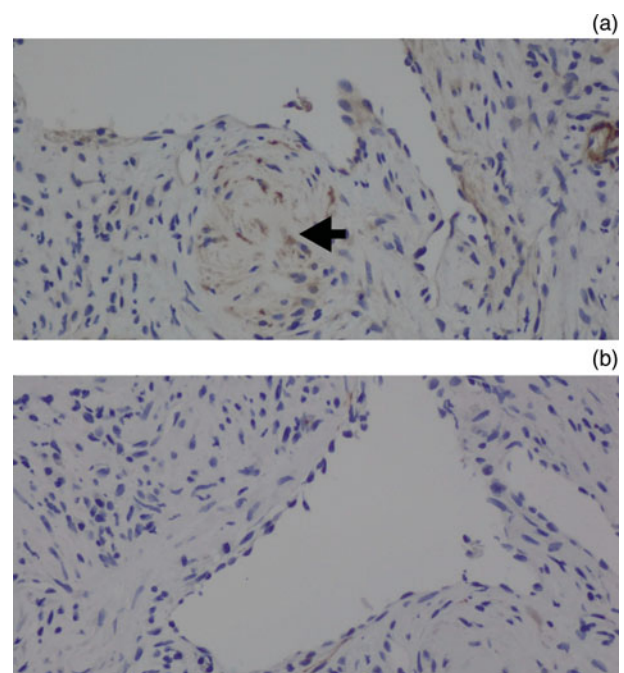


FIG. 4

Microscopic view of the tumour cells with immunohistochemistry staining showing (a) h-caldesmon immunopositivity (arrow) and (b) desmin immunonegativity. ($\times 480$)

Discussion

The pericyte, first described by Zimmermann in 1923, is a pluripotent cell that may differentiate into smooth muscle cells, adipocytes and osteoblasts.⁸ In coining the term 'myopericyte' in 1992, Dictor *et al.* referred to it as a cell transitional between pericytes and smooth muscle cells of

vessels.⁹ Neoplasms that display putative pericytic differentiation were first documented as haemangiopericytomas by Stout and Murray in 1942; however,⁸ the lack of reliable and reproducible diagnostic features for haemangiopericytoma has resulted in an ill-defined entity that is now largely regarded as a growth pattern rather than a distinct tumour.¹⁰ It is only in the last 13 years that myopericytoma has been increasingly recognised and defined as a distinct entity.^{1–9,11–16} Myopericytoma is categorised as a tumour that originates from the perivascular myoid cell, and it shares features of smooth muscle and glomus cells.¹¹ Myopericytoma is currently recognised by the World Health Organization as belonging to a subset of perivascular tumours with myoid features.¹³

Most reported myopericytomas are slow-growing, benign tumours with a good prognosis. However, local recurrences have been recorded; these are most commonly the result of incomplete excision.⁷ The management of myopericytomas involves local excision, with further excision if there is recurrence.

Since its first description, the clinicopathological spectrum has expanded to include tumours occurring in intravascular locations,¹⁴ those associated with the Epstein–Barr virus infection and immunosuppression,⁵ and tumours with a malignant profile.⁸ Malignant myopericytoma, which is rarely reported, is typified by disseminated disease and patient mortality.¹⁵

To date, myopericytoma has most commonly been documented in the lower limbs, followed by the upper limbs, trunk, and head and neck area. Recently, visceral involvement of the kidney and periauricular region have been described.^{12,15} The age of afflicted patients ranges from 13 to 87 years.¹³ There is a distinct male predilection. Whilst the head and neck region is a recognised site,^{4,6,7,16} there has been no previous report in the English-language literature of myopericytoma involving the external auditory canal. Masses in the external auditory canal pose clinicopathological challenges to otolaryngologists and histopathologists because of the diverse aetiological possibilities and overlapping presentations. Whilst the clinical diagnostic possibilities include congenital and acquired infective, metabolic, genetic and neoplastic diseases, the histopathological assessment narrows this spectrum. The cytoarchitectural hallmark of myopericytomas is the variable concentric perivascular growth of tumour cells with an oval to spindle morphology. The immunohistochemical profile of myopericytoma, which is now better defined, is characterised by

tumoural immunopositivity for alpha smooth muscle actin and h-caldesmon.¹³ Desmin may be focally positive or negative.¹³

Despite the known typical pathological features of myopericytoma, diagnostic histopathological challenges arise because of shared phenotypic and immunophenotypic features of benign and malignant tumours in this location (Table I). Infantile myofibroma and myofibromatosis are associated with fibroblastic or myofibroblastic proliferation, and have a biphasic zonation pattern and haemangiopericytomatous vasculature that lacks the concentric perivascular cellular arrangement and h-caldesmon immunopositivity.^{8,11} In contrast to myopericytoma, angioleiomyoma and leiomyoma demonstrate a desmin-immunopositive mature muscle phenotype.^{8,13} Whilst spindle cell haemangioma contains spindle cells and blood vessels, the latter are of cavernous morphology.¹⁷ The whirling pattern of tumour cells around blood vessels is reminiscent of a meningioma; however, meningiomas are usually epithelial membrane antigen positive, and alpha smooth muscle actin and muscle-specific actin immunonegative.¹⁸ Myopericytoma, haemangiopericytoma and solitary fibrous tumour demonstrate intratumoural staghorn-type vasculature, but only myopericytoma demonstrates a striking multilayered growth of spindle cells with a myoid immunophenotype around blood vessels. Myopericytoma lacks the CD99 and CD34 immunopositive spindle cells that are typical of haemangiopericytoma,¹⁶ and the cytoarchitectural spectrum and CD34 immunopositive cells of solitary fibrous tumour.¹⁹ Glomus tumours usually lack a spindle cell component and the typical ‘spinning-off’ of tumour cells directly from the blood vessels, as seen in myopericytoma.⁸ Jugulotympanic paraganglioma, a highly vascular tumour, has a typical nest-like growth pattern of variably spindle cells that are synaptophysin and chromogranin immunopositive.¹⁷ Kaposi sarcoma, a human herpesvirus 8 positive, spindle cell tumour, demonstrates an endothelial rather than a myopericytic immunophenotype.²⁰

TABLE I
HISTOPATHOLOGICAL MIMICKERS OF HEAD AND
NECK MYOPERICYTOMA

<i>Blood vessel-rich pattern</i>
Angioleiomyoma
Glomus jugulare
Kaposi sarcoma
Spindle cell haemangioma
Haemangiopericytoma
<i>Spindle cell-rich pattern</i>
Infantile myofibroma & myofibromatosis
Leiomyoma
Solitary fibrous tumour
<i>Whirled pattern</i>
Glomus tumour
Meningioma

- Myopericytoma is a relatively recently described cutaneous and soft tissue tumour
- It presents rarely in the head and neck area, but its occurrence in the ear is undocumented
- It is characterised by a perivascular proliferation of spindle cells, with perivascular myoid cell or pericytic differentiation
- This paper describes a novel case of myopericytoma involving the external auditory canal and tragus
- Heightened clinicopathological awareness of myopericytoma is pivotal to diagnosis
- Myopericytoma needs distinguishing from other benign and malignant tumours with similar features

In conclusion, this report details the hitherto unreported occurrence of a myopericytoma that involved the external auditory canal and tragus. Heightened clinicopathological awareness of the expanding anatomical distribution of myopericytoma is critical to its diagnosis and differentiation from microscopic look-alikes that are site-dependent.¹⁵

Myopericytoma should be added to the range of external auditory canal neoplasms, especially those characterised by an admixture of spindle cells and a prominence of blood vessels, including those with a haemangiopericytomatous pattern.

References

- 1 Granter SR, Badizadegan K, Fletcher CD. Myofibromatosis in adults, glomangiopericytoma, and myopericytoma: a spectrum of tumours showing perivascular myoid differentiation. *Am J Surg Pathol* 1998;**22**:513–25
- 2 Cao JH, Xu JP, Li YC, Lai J, Li Q. Pulmonary myopericytoma: a case report and review of the literatures. *Chin Med J (Engl)* 2009;**122**:755–7
- 3 Woollard AC, Southgate C, Blair JW. Intravascular myopericytoma of the superficial palmar arch. *J Hand Surg Eur* 2007;**32**:475–6
- 4 Datta V, Rawal YB, Mincer HH, Anderson MK. Myopericytoma of the oral cavity. *Head Neck* 2007;**29**:605–8
- 5 Lau PP, Wong OK, Lui PC, Cheung OY, Ho LC, Wong WC *et al.* Myopericytoma in patients with AIDS: a new class of Epstein-Barr virus-associated tumor. *Am J Surg Pathol* 2009;**33**:1666–72
- 6 Terada T. Minute myopericytoma of the neck: a case report with literature review and differential diagnosis. *Pathol Oncol Res* 2010;**16**:613–16
- 7 Wilson T, Hellquist HB, Ray S, Pickles J. Intranasal myopericytoma. A tumour with perivascular myoid differentiation: the changing nomenclature for haemangiopericytoma. *J Laryngol Otol* 2007;**121**:786–9
- 8 Mentzel T, Dei Tos AP, Sapi Z, Kutzner H. Myopericytoma of skin and soft tissues: clinicopathologic and immunohistochemical study of 54 cases. *Am J Surg Pathol* 2006;**30**:104–13
- 9 Paek JO, Kang HS, Yeo KY, Yu HJ, Kim JS. A case of myopericytoma on the lower leg. *Ann Dermatol* 2011;**23**:201–4
- 10 Fletcher CDM. Hemangiopericytoma – a dying breed? Reappraisal of an ‘entity’. *Curr Diagn Pathol* 1994;**1**:19–23
- 11 Dray MS, McCarthy SW, Palmer AA, Bonar SF, Stalley PD, Marjoniemi V *et al.* Myopericytoma: a unifying term for a spectrum of tumours that show overlapping features with myofibroma. A review of 14 cases. *J Clin Pathol* 2006;**59**:67–73
- 12 Lau SK, Klein R, Jiang Z, Weiss LM, Chu PG. Myopericytoma of the kidney. *Human Pathol* 2010;**41**:1500–4
- 13 McMenamin ME. Myopericytoma. In: Fletcher CDM, Unni KK, Mertens F, eds. *Pathology and Genetics of Tumours of Soft Tissue and Bone*. Lyon: IARC Press, 2002;138–9
- 14 McMenamin ME, Calonje E. Intravascular myopericytoma. *J Cutan Pathol* 2002;**29**:557–61
- 15 Ramdial PK, Sing Y, Deonarain J, Singh B, Allopi L, Moodley P. Periampullary Epstein-Barr virus-associated myopericytoma. *Human Pathol* 2011;**42**:1348–54
- 16 Xia L, Chen Y, Geng N, Jiang J, Yang M, Zhang W. Multifocal myopericytoma in the maxillofacial region: a case report. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2010;**109**:e59–62
- 17 Weiss SW, Goldblum JR. Benign tumours and tumor-like lesions of blood vessels. In: Weiss SW, Goldblum JR, eds. *Enzinger and Weiss’s Soft Tissue Tumours*. St. Louis: Mosby Elsevier, 2008;633–79
- 18 Perry A, Louis DN, Scheithauer BW, Budka H, von Deimling A. Meningiomas. In: Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, eds. *WHO Classification of Tumours of the Central Nervous System*. Lyon: IARC Press, 2007;164–72
- 19 Gengler C, Guilloi L. Solitary fibrous tumour and haemangiopericytoma: evolution of a concept. *Histopathology* 2006;**48**:63
- 20 Lamovec J, Knuutila S. Kaposi sarcoma. In: Fletcher CDM, Unni KK, Mertens F, eds. *Pathology and Genetics of Tumours of Soft Tissue and Bone*. Lyon: IARC Press, 2002;170–2

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