CASE REPORT

Intracranial myopericytoma: a tumour in a rare location

Rui Caetano Oliveira,¹ Ricardo Velasco,² Gonçalo Roque Santos,³ Olinda Rebelo⁴

¹Department of Pathology, Hospitais da Universidade de Coimbra, Coimbra, Portugal

²Department of Neurosurgery, Centro Hospitalar Tondela Viseu EPE, Viseu, Portugal

³Department of Radiology, Centro Hospitalar Tondela Viseu EPE, Viseu, Portugal

⁴Department of Neuropathology, Centro Hospitalar e Universitario de Coimbra EPE, Coimbra, Portugal

Correspondence to
Dr Rui Caetano Oliveira,
ruipecrooliveira@hotmail.com

Accepted 3 January 2018

SUMMARY

A 49-year-old female with history of headache, nausea and vomiting with some weeks of evolution, without neurological symptoms. Radiology revealed an expansive lesion near the inferior vermix and cerebellar tonsils, with heterogeneous gadolinium uptake and mass effect on the fourth ventricle, representing a probable extraventricular origin for the lesion. Pathological examination showed a proliferation of oval/spindle cell proliferation with eosinophil cytoplasm and small and monotonous nuclei, without mitoses. The cells had a concentric growth, surrounding thin-walled blood vessels with foci of stromal myxoid degeneration and whorled pattern. The vessels had a haemangiopericytoma pattern and were lined by non-atypical endothelial cells. The tumorous cells expressed vimentin, alpha-smooth actin and heavy-chain caldesmon and were negative for epithelial membrane antigen, protein S100, HMB45, CD34, calponin and desmin, thus providing the final diagnosis of intracranial myopericytoma.

BACKGROUND

This case was written due to its rarity and to increase awareness of the entity. Myopericytoma is a benign entity more common in soft tissue, however, it may arise anywhere. Clinical and radiological features are unspecific with symptoms arising due to compression of nearby structures and may be diagnosed as a more aggressive tumour. Pathology examination provides the definite diagnosis, but the differential diagnosis is wide. The authors hope that this case raise awareness of this entity in intracranial location—only five cases reported, including this one, and help other pathologists in the differential diagnosis.

CASE PRESENTATION

The patient was a 49-year-old woman with history of headache associated with nausea and vomiting with some weeks of evolution, without neurological symptoms.

She had been subjected to radiological examinations outside the hospital, namely CT scan and MRI that showed an expansive lesion of the posterior fossa.

No regular medication was referred—only occasional analgesics. Familiar background was unremarkable.

INVESTIGATIONS

Patient was then referred to the Neurosurgery department for study and therapeutic decision.

MRI was repeated and revealed an expansive lesion near the inferior vermix and cerebellar tonsils, with heterogeneous gadolinium uptake and mass effect on the fourth ventricle, representing a probable extraventricular origin for the lesion (figure 1). The location and morphology are suspicious of medulloblastoma; however, the age and signal on T2 are not in favour of that hypothesis; a secondary lesion could not be excluded.

Surgery was performed: suboccipital craniotomy with lesion excision; specimen was then sent to pathology evaluation.

Histological examination was performed on H&E stained slides observed in light microscope (Nikon Eclipse 50i) and images obtained using a Nikon-Digital Sight DS-Fi1 camera.

The lesion was composed of oval/spindle cell proliferation with eosinophil cytoplasm and small and monotonous nuclei, without mitotic activity.

The cells had a concentric growth, surrounding thin-walled blood vessels with multiple foci of stromal myxoid degeneration and whorled pattern. The vessels had variable size, with haemangiopericytoma pattern, lined by monolayered of non-atypical endothelial cells (figure 2).

DIFFERENTIAL DIAGNOSIS

Conjugation of morphological characteristics and location of the lesion provided the following differential diagnoses: meningioma, fibroblastic/miofibroblastic tumour (fibrous solitary tumour/haemangiopericytoma), vascular lesion (arterious/venous malformation, cavernous haemangioma), pericytic tumour (angioleiomyoma, myopericytoma) and neural sheath tumour (schwannoma).

Immunohistochemical studies were performed on one representative block of the lesion, resorting to avidin–biotin–peroxidase complex detection system and performed on Ventana Marker Platform Bench Mark ULTRA IHC/ISH using the following antibodies: epithelial membrane antigen (EMA, E29; Ventana, Arizona, USA), Ki67 (30–9, Ventana), melanosome (HMB45, Ventana), S100 protein (S100, 4C4.9, Ventana), alfa-smooth actin (1A4, Ventana), desmin (DE-R-11, Ventana), heavy-chain caldesmon (E89, Ventana), calponin (EP789Y), CD34 (QBEnd/10, Ventana) and vimentin (V9, Ventana).

The Van Gieson staining technique was also performed and did not show elastic fibres on the vessels walls, not supporting the diagnosis of a vascular lesion.

To cite: Oliveira RC, Velasco R, Santos GR, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-223111

Figure 1 MRI T1 sequence—sagittal (A) and axial (B) planes after gadolinium administration: expansive lesion in the vermis inferior area with heterogeneous contrast product uptake and poorly defined boundaries. The lesion has mass effect on the fourth ventricle inducing collapse.

The tumour expressed vimentin without EMA (figure 3A) expression, thus ruling out meningioma; S100 protein was also negative and allowed to exclude a schwannoma.

HMB45 was also negative. Regarding muscle markers, the tumour expressed alpha-smooth actin and heavy-chain caldesmon, with negativity for calponin and desmin (figure 3B,C). CD34 was only positive on the endothelial cells, not compatible with fibrous solitary tumour/haemangiopericytoma (figure 3D).

The proliferative index (Ki67) was low (inferior to 1%).

The conjugation of morphological and ancillary studies allowed the final diagnosis of intracranial myopericytoma.

TREATMENT

Treatment was surgical, and due to benign nature of the lesion no further therapy was preconised.

OUTCOME AND FOLLOW-UP

Patient was discharged 2½ weeks after surgery. Later she developed surgical wound infection, promptly resolved with short-term hospitalisation and use of antibiotics. One year after surgery, patient is okay, without nausea, vomiting and neurological symptoms. She will have annual MRI until 3 years after

Figure 2 Vessels with haemangiopericytoma pattern surrounded by oval/spindle cells (A; H&E ×40) with stromal myxoid degeneration (B; H&E ×20). The cells exhibit a whorled configuration (C; H&E ×100), and the vessels were lined by non-atypical endothelial cells (D; H&E ×400).

Figure 3 Immunoprofiling: the tumorous cells were negative for epithelial membrane antigen (A; ×40) and diffusely positive for alpha-smooth actin (B; ×100) and heavy-chain caldesmon (C; ×200). The cells were also negative for CD34, this one positive in the endothelial cells (D; ×200).

surgery, and if there is no tumorous relapse it will be performed each 2 years.

DISCUSSION

Myopericytoma is a benign perivascular neoplasm that originates from the myoid cells.¹ It is more common in middle-aged adults, however with wide range, and preferentially arises in dermal/subcutaneous tissue of distal extremities or retroperitoneum.² It was first described by Granter *et al*³ when he characterised a cohort of 24 patients with tumours that showed perivascular myoid differentiation and called ‘perivascular myoma’; nowadays, it is well known that they correspond to a continuum morphological spectrum, including myofibroma, myopericytoma and glomangiopericytoma.¹²

Intracranial myopericytoma are very rare, first reported by Rousseau *et al*⁴ in a cohort of three patients, well documented with morphological, immunohistochemical and ultrastructural studies.

After that, only two more articles make reference to intracranial myopericytomas—Zhang *et al*⁵ and Holling *et al*,⁶ the latter with reference to a cohort of five patients, where the authors include also one peripheral nervous system and two intraspinal myopericytoma; intraspinal location has also been reported in the past by Cox and Giltman,⁷ Hunald *et al*,⁸ Agrawal and Nag,⁹ Brunschweiler *et al*,¹⁰ Chew *et al*¹¹ and by Cobos I and Hedley-Whyte ET (personal communication, 2014).

However, in the last years, myopericytoma has been described in several places, including in the nasal fossae.¹²

The patient’s clinical characteristics, including the one reported in this article and excluding the case with peripheral nervous system myopericytoma, can be consulted on table 1.

All the cases had a wide range of age, with slight female predominance (6:4). The histomorphological findings were similar, reporting a tumour composed of large vessels with thick walls and myxoid changes, lined by oval and monotonous endothelial cells. The atypical location invoked several differential diagnoses, mainly meningioma, angioleiomyoma and solitary fibrous tumour/haemangiopericytoma. Perhaps, the most difficult distinction is with angioleiomyoma, due to morphological

Table 1 Clinical characteristics of patients reported in the literature with myopericitoma of the central nervous system

Author/year	Age/gender	History	Clinical symptoms	Location	Radiology assumption	Size (cm)	Follow-up
Cox and Giltman ⁷ /2003	50, female	Not relevant	Progressive weakness of the arms and legs	T3	Not provided	Not provided	Not provided
Rousseau <i>et al</i> ⁴ /2005	50, female	Neonatal hypoxic-ischaemic brain damage and tetraparesis	Vomiting and axial hypotonia	Pineal region	Not provided	2.5	6 months, death from non-tumorous causes
Rousseau <i>et al</i> ⁴ /2005	56, female	Glaucoma and asthma	Decreased visual acuity of the right eye	Right orbital apex	Cavernous haemangioma	0.9	9 months, no tumour recurrence
Rousseau <i>et al</i> ⁴ /2005	59, female	Ectopic pregnancy, asthma, chronic depressive syndrome	Decreased visual acuity of the left eye	Anterior canal fossa and reaching the optic chiasm	Meningioma	3.5	12 months, no tumour recurrence
Brunschweiler <i>et al</i> ¹⁰ /2009	43, female	History of osteomalacia due to T5 tumour—incomplete removal	Acute pain of the upper back, involving shoulders	T5	Not provided	Not provided	24 months, no tumour recurrence
Agrawal and Nag ⁹ /2013	50, female	Not relevant	Pain in the back with gradual onset of paraparesis	T8	Infectious/tumorous	Not provided	32 months, no tumour recurrence
Cobos and Hedley-Whyte (personal communication)/2014	64, female	Metastatic melanoma	Progressively worsening headaches in left portion of the neck	C1–C2, intradural	Vascular lesion?	1	Not provided
Zhang <i>et al</i> ⁵ /2015	36, male	Not relevant	Left-sided Bell's palsy	Right cerebellar convexity	Meningioma	2.6	Not provided
Holling <i>et al</i> ⁶ /2015	74, male	Lung cancer	Progressive swelling in medial corner of left eye	Medial orbita	Metastasis	Not provided	19 months, no tumour recurrence
Holling <i>et al</i> ⁶ /2015	38, male	Not relevant	Progressive pain in right dorsal calf	L5–S1, intradural	Schwannoma	Not provided	18 months, no tumour recurrence
Holling <i>et al</i> ⁶ /2015	58, male	Larynx carcinoma	Pain in S1 dermatoma	S1–S4, intraspinal	Metastasis	Not provided	84 months, no tumour recurrence
Holling <i>et al</i> ⁶ /2015	61, female	Not relevant	Diplopia	Intrasellar/perisellar	Pituitary adenoma	Not provided	55 months, no tumour recurrence
Chew <i>et al</i> ¹¹ /2017	63, male	Not relevant	Back pain, bilateral lower limb numbness/weakness	T9, intradural	Tumorous	1.6	12 months, no tumour recurrence
Current case	49, female	Not relevant	Headache, vomiting and nausea	Inferior vermix/cerebellar tonsils	Medulloblastoma/secondary lesion	3.5	12 months, no tumour recurrence

overlap between the two identities—feature also common in the soft tissue but with scarce literature report of intracranial cases¹³; however, normally, desmin is negative in myopericytoma and positive in angioleiomyoma.²

Regarding its origin, some studies found peculiar molecular changes: t(7;12)(p22;q13) and del(6)(q12q15), the latter in infantile tumours.¹ Some authors postulate a probable relation between Epstein-Barr infection in AIDS context and myopericytomas; however, the reports on this association are very rare.^{14 15}

Regarding overall survival and tumour relapse, intracranial myopericytoma is a low-grade tumour, with excellent survival in all patients—only one patient died and it was due to non-tumour-related causes.

In conclusion, myopericytomas of the central nervous system are very rare, and from those only five cases with intracranial location are described in the literature, including the one described in this manuscript. They are benign neoplasm, successfully manageable by surgery and with excellent follow-up.

Learning points

- Myopericytoma is a benign perivascular neoplasm with origin in the myoid cells.
- It is more common in the soft tissue; however, it may arise everywhere.
- Intracranial location is extremely rare and may be clinically and radiologically diagnosed as a more aggressive tumour.
- Surgery is the treatment of choice with excellent prognosis.

Contributors Conception and design or acquisition of data or analysis and interpretation of data: RCO, RV, GRS, OR; Drafting the article or revising it critically for important intellectual content: RCO, OR; Final approval of the version to be published: RCO, RV, GRS, OR.

Funding This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

© BMJ Publishing Group Ltd (unless otherwise stated in the text of the article) 2018. All rights reserved. No commercial use is permitted unless otherwise expressly granted.

REFERENCES

- 1 Mentzel T. Myopericytoma, including myofibroma. In: *WHO classification of soft tissue and bone tumors*. 4th edn. Lyon: IARC, 2013.
- 2 Lindberg MR. *Diagnostic pathology of soft tissue tumors*. 2nd edn, 2015:344–5. section 8.
- 3 Granter SR, Badizadegan K, Fletcher CD. Myofibromatosis in adults, glomangiopericytoma, and myopericytoma: a spectrum of tumors showing perivascular myoid differentiation. *Am J Surg Pathol* 1998;22:513–25.
- 4 Rousseau A, Kujas M, van Effenterre R, et al. Primary intracranial myopericytoma: report of three cases and review of the literature. *Neuropathol Appl Neurobiol* 2005;31:641–8.
- 5 Zhang CH, Hasegawa H, Johns P, et al. Myopericytoma of the posterior cranial fossa. *Br J Neurosurg* 2014;29:90–1.
- 6 Holling M, Wildförster U, Verheggen R, et al. Myopericytoma: a series of 5 cases affecting the nervous system. *World Neurosurg* 2015;84:1493.e5–1493.e8.
- 7 Cox DP, Giltman L. Myopericytoma of the thoracic spine: a case report. *Spine* 2003;28:E30–2.
- 8 Hunald FA, Rakotoarisoa JC, Razafimahatratra R, et al. [Thoracic myopericytoma in a child. A case report]. *Arch Pediatr* 2008;15:1660–2.
- 9 Agrawal N, Nag K. Myopericytoma of the thoracic spine: a case report and review of literature. *Spine J* 2013;13:e23–7.
- 10 Brunschweiler B, Guedj N, Lenoir T, et al. Oncogenous osteomalacia and myopericytoma of the thoracic spine: a case report. *Spine* 2009;34:E857–60.
- 11 Chew LS, Han XJ, Tan KK, et al. Hemangiopericytoma of the thoracic spine: a case report. *J Surg Case Rep* 2017;2017:rjx121.
- 12 Wilson T, Hellquist HB, Ray S, et al. Intranasal myopericytoma. A tumour with perivascular myoid differentiation: the changing nomenclature for haemangiopericytoma. *J Laryngol Otol* 2007;121:786–9.
- 13 Sun L, Zhu Y, Wang H. Angioleiomyoma, a rare intracranial tumor: 3 case report and a literature review. *World J Surg Oncol* 2014;12:216.
- 14 Calderaro J, Polivka M, Gallien S, et al. Multifocal Epstein Barr virus (EBV)-associated myopericytoma in a patient with AIDS. *Neuropathol Appl Neurobiol* 2008;34:115–7.
- 15 Lau PP, Wong OK, Lui PC, et al. Myopericytoma in patients with AIDS: a new class of Epstein-Barr virus-associated tumor. *Am J Surg Pathol* 2009;33:1666–72.

Copyright 2017 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow