

Intracranial Myopericytoma: A Rare Benign Tumor at an Extremely Rare Location

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Abstract

A 50-year-old female presented with a history of seizures, headache, nausea, and vomiting. On imaging, parafalcine meningioma with mass effect features was rendered. She underwent right frontal tumor excision and craniotomy. Pathological examination showed a tumor composed of syncytial aggregates of round to plump fusiform cells forming whorls around prominent branching congested vessels. The tumorous cells expressed α -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.

Keywords

- ▶ pathological
- ▶ immuno-histochemical
- ▶ intracranial myopericytoma

The rarity of this benign tumor at an extremely rare location prompted this study. As preoperative radiological investigations are nonspecific in such cases, a detailed and comprehensive pathological examination is mandatory to come to a definitive diagnosis.

Introduction

Myopericytoma is a benign tumor that usually arises in the subcutaneous and superficial soft tissue of the extremities. Very few cases have been reported at other locations, intracranial presentation being exceptional. The neoplasm is believed to originate from the perivascular myoid cellular environment, and was previously classified as a variant of hemangiopericytoma.¹

Here, we describe this relatively rare tumor, at a rare location and aim to enhance the awareness of this entity by supplementing the literature. Clinical and radiological features are largely nonspecific and a definitive diagnosis is possible only with the help of histopathologic examination, supplemented by immunohistochemistry (IHC) to exclude similar entities.

Case History

A 50-year-old female presented with a history of seizure associated with nausea and vomiting and left-sided weakness since 1 month. There was no predisposing history of hypertension and diabetes mellitus.

Radiological Investigation

Her preoperative noncontrast computed tomography (CT) scan revealed a large hyperdense well-defined possibly extra-axial, right parafalcine mass with surrounding edema, causing a contralateral midline shift (▶ Fig. 1A).

Magnetic resonance imaging brain with contrast revealed a well-defined extra-axial mass measuring 48 × 50 × 43 mm (vertical × anteroposterior × transverse) appearing hyperintense on T2/fluid-attenuated inversion recovery images, in

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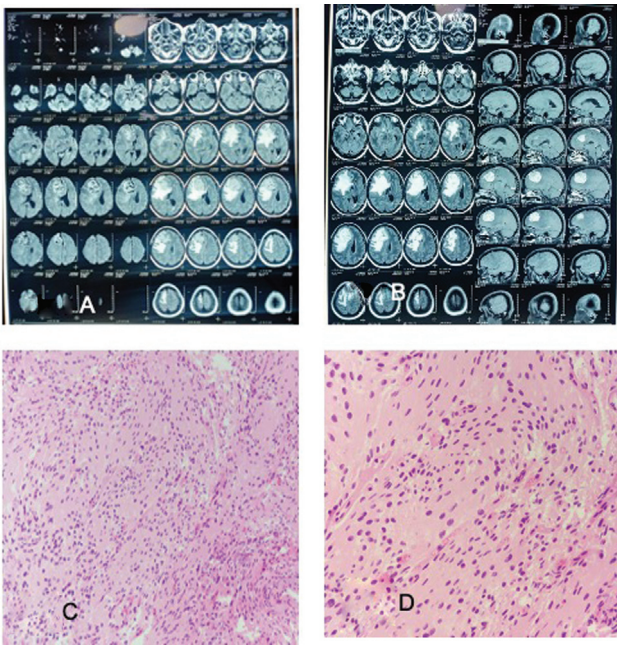


Fig. 1 (A and B) Magnetic resonance imaging (MRI) with contrast brain (preoperative)—hyperintense, well-defined extra-axial hypervascular and hypercellular mass showing features of parafalcine meningioma with mass effect. (C) Microphotograph showing the tumor having syncytial aggregates of round to plump fusiform cells forming whorls around prominent branching congested vessels. (hematoxylin and eosin [H&E] $\times 100$). (D) Microphotograph showing fusiform cells showing no mitotic activity, no pleomorphism (H&E $\times 400$).

the right parafalcine region with moderate surrounding edema causing mass effect in the form of effacement of adjacent sulcal spaces, compression of right lateral ventricle, contralateral midline shift (25 mm), and contralateral subfalcine herniation. The mass showed multiple flow voids with intense postcontrast enhancement and restriction of diffusion on diffusion-weighted imaging, suggesting a hypervascular and hypercellular mass. Enhancing dural tail is also seen. The vascularity/feeding vessels appear to arise from the convexity and falx. The mass is hyperdense on noncontrast CT scan. The imaging findings were reported as consistent with parafalcine meningioma causing mass effect (**► Fig. 1B**).

Patient was then referred to the neurosurgery department. At the time of admission, she was afebrile and her vitals were stable. She was taken up for right frontal craniotomy and tumor excision.

Preoperative Findings

The tumor was extra-axial, well-defined, firm to cystic in consistency, and highly vascularized. The tumor was adherent to the falx cerebri and suprasagittal sinus. The tumor capsule showed a large vascular channel with dilated vascular channels in the tumor substance as well.

Pathologic Findings

Postsurgery the excised tumor was sent for pathological evaluation.

Grossly the specimen consisted of multiple gray-white to gray-brown soft tissue bits measuring $5.5 \times 4 \times 0.8$ cm in aggregates.

After processing of the tissue histological examination was performed on hematoxylin and eosin-stained slides and observed in light microscope (Olympus BX 43).

Histologically, the tumor was composed of syncytial aggregates of round to plump fusiform cells forming whorls around prominent branching congested vessels (**► Fig. 1C, D**).

No evidence of significant pleomorphism or necrosis or mitotic activity was seen. No evidence of infiltration into neuroparenchyma seen.

A detailed and comprehensive IHC evaluation was performed using the Ventana BenchMark XT autostainer. The tumor cells expressed alpha-smooth muscle actin (SMA) (**► Fig. 2A**) and h-caldesmon (**► Fig. 2B**). Epithelial membrane antigen (EMA) (**► Fig. 2C**), HMB-45, Melan-A, SOX-10, signal transducer and activator of transcription 6 (STAT6) (**► Fig. 2E**), and S-100 (**► Fig. 2F**) were negative. A CD34 immunostain highlighted the prominent branching vascular channels (**► Fig. 2D**). Based on the morphologic appearance, a differential diagnosis of meningioma/perivascular myoid tumor/solitary fibrous tumor/hemangiopericytoma was considered.

The above morphologic and IHC features were deemed to be consistent with a mesenchymal and non-meningothelial tumor favoring an intracranial myopericytoma (**► Table 1**).

Following surgery, the postoperative noncontrast CT head findings were suggestive of extensive edema, hematomas, and mild pneumocephalus and a midline shift of approximately 2 cm.

On day 1 postsurgery, reexploratory craniotomy and hematoma excision was done through the previous coronal incision.

Preoperative findings: There was a hematoma present just below the frontal lobe and all around the residual tumor. The hematoma and part of the brain were excised.

After the reexploration, the patient was stable.

Discussion

Myopericytoma is a low-grade, benign perivascular neoplasm that originates from the myoid cells.¹

It is more prevalent in middle-aged adults, although it can occur at any age. The most common site of its occurrence is in the dermal or subcutaneous tissue of the distal extremities or retroperitoneum. An intracranial location is extremely rare. Myopericytomas show a female predominance tumor.

Intracranial myopericytoma was first reported by Rousseau et al in a cohort of three patients, who documented the morphological, IHC, and ultrastructural features of this entity.²

Zhang et al,³ Holling et al,⁴ Cox and Giltman,⁵ Hunald et al,⁶ and Agrawal and Nag⁷ have also reported cases of myopericytoma in the intraspinal and peripheral nervous system.

The clinical characteristics of patient reported in the literature with myopericytoma of the central nervous system are shown in **► Table 2**.

The most common clinical presentation of intracranial myopericytoma is compression symptoms like paraparesis,

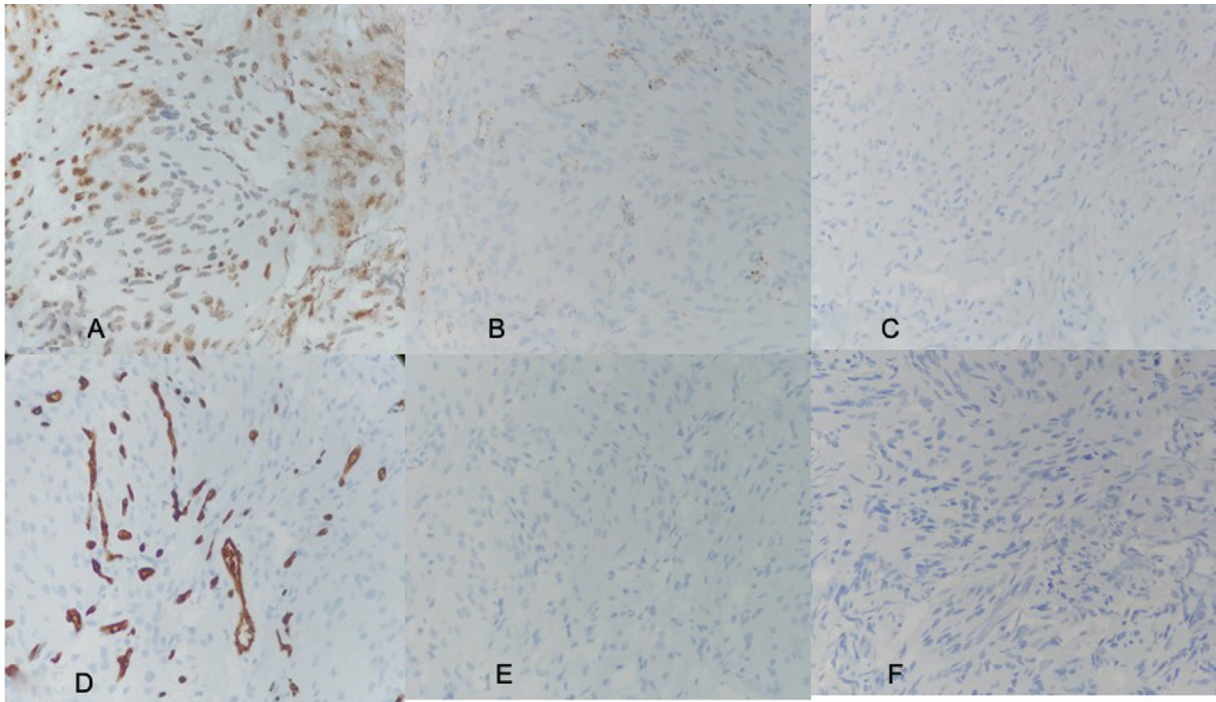


Fig. 2 (A) Immunoreactivity score 2+ in neoplastic cells (immunohistochemistry [IHC] stain smooth muscle actin [SMA] $\times 400$). (B) Immunoreactivity score 2+ in neoplastic cells (IHC stain caldesmon $\times 400$). (C) Nonimmunoreactivity score 0 in neoplastic cells (IHC stain epithelial membrane antigen [EMA] $\times 400$). (D) Nonimmunoreactivity score 0 in neoplastic cells (IHC stain CD34 $\times 400$). (E) Nonimmunoreactivity score 0 in neoplastic cells (IHC stain signal transducer and activator of transcription 6 [STAT6] $\times 400$). (F) Nonimmunoreactivity score 0 in neoplastic cells (IHC stain S-100 $\times 400$).

decreased visual acuity, and diplopia, and constitutional symptoms like backache, vomiting, headache, etc.

In our case, the patient presented with seizures and weakness in the lower limbs associated with constitutional symptoms of vomiting and nausea.

Myopericytoma is part of a morphologic continuum that includes myofibroma, myopericytoma, and glomangiopericytoma.¹

The histological findings in myopericytoma reveal a concentric perivascular proliferation of bland spindle-shaped pericytic cells with myoid features.

The differential diagnosis in the intracranial location includes meningioma, myofibroblastic/fibroblastic tumors (solitary fibrous tumor/hemangiopericytoma), vascular lesion (arteriovenous malformation, cavernous hemangioma), pericytic tumors (angioliomyoma, myopericytoma), and neural sheath tumor (schwannoma).

In our case, the tumor cells expressed smooth muscle cell markers, namely, SMA and heavy-chain caldesmon with negative EMA expression, thus excluding meningioma. IHC for S100 protein was also negative and allowed the exclusion of a schwannoma.

Table 1 Immunohistochemistry evaluation

Antibody	[Clone] -	Interpretation
CD34 -	[QBEnd10] -	Non-immunoreactive score "0" in neoplastic cells (immunoreactive in proliferating vascular channels)
Caldesmon -	[EP19] -	Immunoreactive score 2+ in neoplastic cells
EMA -	[E-29] -	Non-immunoreactive score "0" in neoplastic cells
HMB-45 -	[Melanoma] -	Non-immunoreactive score "0" in neoplastic cells
Melan-A -	[A-103] -	Non-immunoreactive score "0" in neoplastic cells
S100 -	[4C4-9] -	Non-immunoreactive score "0" in neoplastic cells
SOX-10 -	[EP263] -	Non-immunoreactive score "0" in neoplastic cells
SMA -	[1A4] -	Immunoreactive score 2+ in neoplastic cells
STAT-6 -	[EP325] -	Non-immunoreactive score "0" in neoplastic cells
GFAP -	[GA-5] -	Non-immunoreactive score "0" in neoplastic cells

Abbreviations: EMA, epithelial membrane antigen; GFAP, glial fibrillary acidic protein; SMA, smooth muscle actin; STAT-6, signal transducer and activator of transcription 6.

Table 2 Clinical characteristics of patient reported in the literature with myopericytoma of the central nervous system

Author/year	Age/ Gender	History	Clinical symptoms	Location	Imaging	Size	Follow-up
Cox and Giltman/2003 ⁵	50/Male	Not relevant	Progressive weakness of arms and legs	T3	Not provided	Not provided	Not provided
Rousseau et al/2005 ²	50/Male	Neonatal hypoxic ischemic brain damage and tetraparesis	Vomiting, axial hypotonia	Pineal region	Not provided	2.5 cm	6 months, death from nontumorous causes
Rousseau et al/2005 ²	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 cm	12 months, no tumor recurrence
Rousseau et al/2005 ²	56/Female	Glaucoma and asthma	Decreased visual acuity of the right eye	Right orbital apex	Cavernous hemangioma	0.9	9 months, no tumor recurrence
Brunschweiler et al/2009 ⁸	43/Female	History of osteomalacia due to T5 tumor— incomplete removal	Acute pain of the upper back, involving shoulders	T5	Not provided	Not provided	24 months, no tumor recurrence
Agrawal and Nag/2013 ⁹	50/Female	Not relevant	Pain in the back with gradual onset of paraparesis	T8	Infectious/ Tumorous	Not provided	32 months, no tumor recurrence
Cobos and Hedley-Whyte, 2014	64/Female	Metastatic melanoma	Progressively worsening headaches in left portion of the neck	C1-C2 intradural	Vascular lesion	1 cm	Not provided
Zhang et al/2015 ³	36/Male	Not relevant	Left-sided Bell's palsy	Right cerebellar convexity	Meningioma	2.6 cm	Not provided
Holling et al/2015 ⁴	74/Male	Lung cancer	Progressive swelling in medial corner of left eye	Medial orbital	Metastasis	Not provided	19 months, no tumor recurrence
Holling et al/2015 ⁴	38/Male	Not relevant	Progressive pain in right dorsal calf	L5-S1, intradural	Schwannoma	Not provided	18 months, no tumor recurrence
Holling et al/2015 ⁴	58/Male	Larynx cancer	Pain in S1 dermatoma	S1-S4 intraspinal	Metastasis	Not provided	84 months, no tumor recurrence
Holling et al/2015 ⁴	61/Female	Not relevant	Diplopia	Intrasellar/perisellar	Pituitary adenoma	Not provided	55 months, no tumor recurrence
Chew et al/2017 ⁷	63/Male	Not relevant	Back pain, bilateral lower limb numbness/weakness	T9 intradural	Tumorous	1.6 cm	12 months, no tumor recurrence
Current case	50/Female	Seizure	Nausea, vomiting, and left-sided weakness	Right parafalcine region		4.8 cm	6 months, no tumor recurrence

HMB45, SOX-10, and Melan-A, were also negative. CD34 was only positive in the endothelial cells, and STAT6 was also non-immunoreactive thereby ruling out solitary fibrous tumor/hemangiopericytoma.

The combination of a detailed morphologic evaluation accompanied by comprehensive IHC allowed a final diagnosis of intracranial myopericytoma to be rendered.

Some studies have found that the genetic changes associated with myopericytomas including t(7;12)(p22;q13) and del(6)(q12q15).¹

As it is a low-grade tumor patients have an excellent chance of survival with no recurrence. Surgical excision is the mainstay of treatment of such tumors.

Conclusion

Intracranial myopericytomas are rare tumors. They are benign and low-grade lesions and may be clinically and radiologically be diagnosed as more aggressive entities. As preoperative radiological findings are nonspecific, an evaluation of histological and IHC features is mandatory for a definitive diagnosis. It is necessary to create an awareness about this rare benign tumor among radiologists and neurosurgeons. The importance of a detailed and comprehensive histologic and IHC evaluation cannot be overemphasized.

Funding

None.

Conflict of Interest

None declared.

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