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Case Report

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

Abstract

A 50-year-old female 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 alpha-smooth muscle 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. The rarity of this benign tumor at an extremely location, prompted this study. As preoperative radiological investigations are nonspecific in such cases, hence a detailed and comprehensive pathological examination is mandatory to come to a definitive diagnosis.

Keywords: *Immunohistochemical, Intracranial myopericytoma, Pathological*

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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, the 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 and immunohistochemistry to exclude similar entities.

Case Report

A 50-year-old female presented with a history of seizures associated with nausea and vomiting and left-sided weakness for 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 [Figure 1a].

Magnetic resonance imaging brain with contrast revealed a well-defined extra-axial mass measuring 48 mm × 50 mm × 43 mm (vertical × anterio posterior × transverse) appearing hyperintense on T2/FLAIR images, in the right parafalcine region with moderate surrounding edema causing mass effect in the form of effacement of adjacent sulcal spaces, compression of the 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 (DWI), 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 [Figure 1b].

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The 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 the right frontal craniotomy and tumor excision.

Peroperative 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 supra sagittal 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 grey white to grey-brown soft-tissue bits measuring 5.5 cm × 4 cm × 0.8 cm in aggregates.

After processing of the tissue, histological examination was performed on H and E-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 [Figure 1c and d].

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

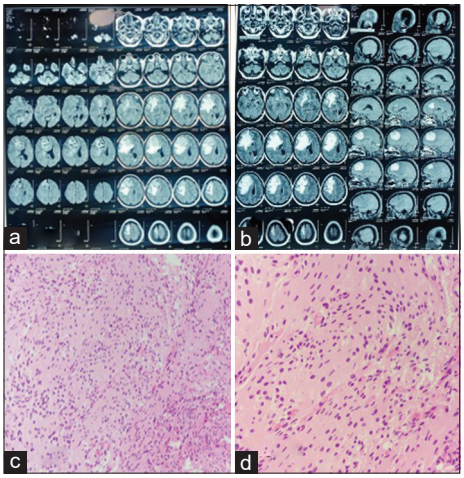

Figure 1: (a) Contrast-enhanced computed tomography brain-(preoperative)-large hyperdense well-defined possibly extra-axial, right parafalcine mass with surrounding edema, causing a contralateral midline shift. (b) Magnetic resonance imaging with contrast brain– (preoperative)-hyperintense, well-defined 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 (H and E, ×100). (d) Microphotograph showing fusiform cells showing no mitotic activity, no pleomorphism (H and E, ×400)

A detailed and comprehensive immunohistochemical evaluation was performed using the Ventana BenchMark XT autostainer [Table1]. The tumor cells expressed alpha-smooth muscle actin (SMA) [Figure 2a] and h-Caldesmon [Figure 2b]. EMA, [Figure 2c], HMB-45, Melan-A, SOX-10, STAT6 [Figure 2e], and S-100 [Figure 2f] were negative. A CD 34 immunostain highlighted the prominent branching vascular channels [Figure 2d]. Based on the morphologic appearance, a differential diagnosis of meningioma/perivascular myoid tumor/solitary fibrous tumor/hemangiopericytoma was considered.

The above morphologic and immunohistochemical features were deemed to be consistent with a mesenchymal,

Table 1: Immunohistochemistry evaluation

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

SMA – Smooth muscle actin

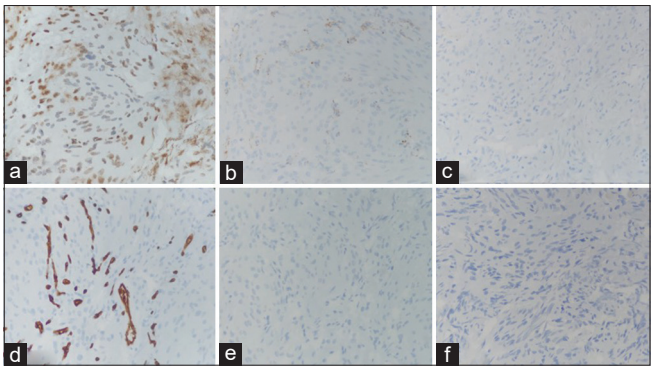

Figure 2: (a) Immunoreactivity score 2+ in neoplastic cells (IHC stain SMA, ×400). (b) Immunoreactivity score 2+ in neoplastic cells (IHC stain Caldesmon, ×400). (c) NonImmunoreactivity Score 0 in neoplastic cells (IHC stain EMA, ×400). (d) NonImmunoreactivity Score 0 in neoplastic cells (IHC stain CD34, ×400). (e) Nonimmunoreactivity Score 0 in neoplastic cells (IHC stain STAT-6, ×400). (f) Nonimmunoreactivity Score 0 in neoplastic cells (IHC stain S-100, ×400)

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 (cm)	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	6 months de death At from non-T tumorous can 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	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 re recurrence
Brunschweiler *et al.*/2009[7]	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	Not Provided
Zhang *et al.*/2015[3]	36/male	Not relevant	Left sided Bell's Palsy	Right cerebellar convexity	Meningioma	2.6	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	Schwanoma	Not provided	18months 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[8]	63/male	Not relevant	Back pain, bilateral lower limb numbness/weakness	T9 Intradural	Tumorous	1.6	12 months No tumor Recurrence
Current case	50/female	Seizure	Nausea, vomiting and left sided weakness	Right parafalcine region		4.8	6 months No tumor Recurrence

nonmeningothelial tumor favoring an intracranial myopericytoma.

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, re-exploratory craniotomy and hematoma excision were done through the previous coronal incision.

Peroperative 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 re-exploration, 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, immunohistochemical, and ultrastructural features of this entity.[2]

Zhang *et al*.[3] and Holling *et al*.,[4] Cox and Giltman,[5] Hunald *et al*.,[6] Agrawal and Nag,[8] have also reported cases of myopericytoma in the intraspinal and peripheral nervous system.

The clinical characteristics of the 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 such as paraparesis, decreased visual acuity, and diplopia and constitutional symptoms such as backache, vomiting, and headache.

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 (Arterio-venous malformation, cavernous hemangioma), pericytic tumors (angioleiomyoma, 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. Immunohistochemistry (IHC) for S100 protein was also negative and allowed the exclusion of a schwannoma.

HMB45, SOX-10, and Melan-A were also negative. CD34 was only positive in the endothelial cells, and STAT6 was also nonimmunoreactive 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, evaluation of histological and immunohistochemical features is mandatory for a definitive diagnosis. It is necessary to create awareness about this rare benign tumor among radiologists and neurosurgeons. The importance of a detailed and comprehensive histologic and immunohistochemical evaluation cannot be overemphasized.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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