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
- immunohistochemical
- 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
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
Post surgery 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.

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 Roussel et al in a cohort of three patients, who documented the morphological, IHC, and ultrastructural features of this entity.

Zhang et al, Holling et al, Cox and Giltman, Hunald et al, and Agrawal and Nag have also reported cases of myopericytoma in the intrasplanic 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,
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 (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. IHC for S100 protein was also negative and allowed the exclusion of a schwannoma.

**Table 1** Immunohistochemistry evaluation

<table>
<thead>
<tr>
<th>Antibody</th>
<th>[Clone]</th>
<th>Interpretation</th>
</tr>
</thead>
<tbody>
<tr>
<td>CD34 -</td>
<td>[QBEnd10] -</td>
<td>Non-immunoreactive score “0” in neoplastic cells (immunoreactive in proliferating vascular channels)</td>
</tr>
<tr>
<td>Caldesmon -</td>
<td>[EP19] -</td>
<td>Immunoreactive score 2+ in neoplastic cells</td>
</tr>
<tr>
<td>EMA -</td>
<td>[E-29] -</td>
<td>Non-immunoreactive score “0” in neoplastic cells</td>
</tr>
<tr>
<td>HMB-45 -</td>
<td>[Melanoma] -</td>
<td>Non-immunoreactive score “0” in neoplastic cells</td>
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<tr>
<td>Melan-A -</td>
<td>[A-103] -</td>
<td>Non-immunoreactive score “0” in neoplastic cells</td>
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<tr>
<td>S100 -</td>
<td>[4C4–9] -</td>
<td>Non-immunoreactive score “0” in neoplastic cells</td>
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<tr>
<td>SOX-10 -</td>
<td>[EP263] -</td>
<td>Non-immunoreactive score “0” in neoplastic cells</td>
</tr>
<tr>
<td>SMA -</td>
<td>[1A4] -</td>
<td>Immunoreactive score 2+ in neoplastic cells</td>
</tr>
<tr>
<td>STAT-6 -</td>
<td>[EP325] -</td>
<td>Non-immunoreactive score “0” in neoplastic cells</td>
</tr>
<tr>
<td>GFAP -</td>
<td>[GA-5] -</td>
<td>Non-immunoreactive score “0” in neoplastic cells</td>
</tr>
</tbody>
</table>

Abbreviations: EMA, epithelial membrane antigen; GFAP, glial fibrillary acidic protein; SMA, smooth muscle actin; STAT-6, signal transducer and activator of transcription 6.
<table>
<thead>
<tr>
<th>Author/Year</th>
<th>Gender</th>
<th>Age/Gender</th>
<th>History</th>
<th>Clinical symptoms</th>
<th>Location</th>
<th>Imaging</th>
<th>Size</th>
<th>Follow-up</th>
<th>Clinical symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cox and Giltman/2003</td>
<td>Male</td>
<td>50/Male</td>
<td>Not relevant</td>
<td>Progressive weakness of arms and legs</td>
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<td>Not provided</td>
<td>Not provided</td>
</tr>
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<td>Rousseau et al/2005</td>
<td>Male</td>
<td>50/Male</td>
<td>Neonatal hypoxic ischemic brain damage and hypopituitarism</td>
<td>Vomiting, axial hypotonia</td>
<td>Pineal region</td>
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<td>Not provided</td>
<td>Not provided</td>
<td>Not provided</td>
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<td>Female</td>
<td>59/Female</td>
<td>Ectopic pregnancy, chronic depressive syndrome</td>
<td>Decreased visual acuity of the left eye</td>
<td>Anterior canal fossa and involving the optic chiasm</td>
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<td>Not provided</td>
<td>Not provided</td>
<td>Not provided</td>
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<tr>
<td>Rousseau et al/2005</td>
<td>Female</td>
<td>56/Female</td>
<td>Glaucoma and asthma</td>
<td>Decreased visual acuity of the right eye</td>
<td>Decreased visual acuity of the right eye</td>
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<td>Not provided</td>
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<tr>
<td>Brunschweiler et al/2009</td>
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<td>43/Male</td>
<td>History of osteomalacia due to T5 tumor– incomplete removal</td>
<td>Acute pain of the upper back, involving shoulders</td>
<td>Pain in the back with gradual onset of paraparesis</td>
<td>CT C-2 intradural</td>
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<td>Not provided</td>
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<td>Agrawal and Nag/2013</td>
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<td>Metastatic melanoma</td>
<td>Pain in the back with gradual onset of paraparesis</td>
<td>Medial orbital</td>
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<tr>
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<td>36/Male</td>
<td>Not relevant</td>
<td>Metastatic melanoma</td>
<td>Left-sided Bell’s palsy</td>
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<td>Not provided</td>
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<tr>
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<td>74/Male</td>
<td>Not relevant</td>
<td>Metastatic melanoma</td>
<td>Progressive pain in right doral calf</td>
<td>L5-S4 intraspinal</td>
<td>Metastatic</td>
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<td>Not provided</td>
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<td>Female</td>
<td>38/Male</td>
<td>Not relevant</td>
<td>Metastatic melanoma</td>
<td>Pain in S1 dermatoma</td>
<td>Intraspinal/periocular</td>
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<td>Male</td>
<td>58/Male</td>
<td>Not relevant</td>
<td>Metastatic melanoma</td>
<td>Diplopia</td>
<td>Intraspinal</td>
<td>Metastatic</td>
<td>Not provided</td>
<td>Not provided</td>
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<tr>
<td>Holling et al/2015</td>
<td>Female</td>
<td>61/Female</td>
<td>Not relevant</td>
<td>Metastatic melanoma</td>
<td>Right paraplegia</td>
<td>T9 intradural</td>
<td>Metastatic</td>
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<td>Not provided</td>
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<tr>
<td>Holling et al/2015</td>
<td>Male</td>
<td>63/Male</td>
<td>Not relevant</td>
<td>Metastatic melanoma</td>
<td>Back pain, bilateral lower limb numbness/weakness</td>
<td>Not relevant</td>
<td>Not provided</td>
<td>Not provided</td>
<td>Not provided</td>
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<tr>
<td>Chew et al/2017</td>
<td>Female</td>
<td>50/Female</td>
<td>Seizure</td>
<td>Back pain, bilateral lower limb numbness/weakness</td>
<td>Not relevant</td>
<td>Not relevant</td>
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</table>

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

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None.

**Conflict of Interest**

None declared.

**References**