


Angioleiomyoma of the falx

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ABSTRACT

A 43-year-old man arrived at the emergency department following a syncopal episode. Computed tomography and magnetic resonance images demonstrated a small interhemispheric, anterior parafalcine mass that mimicked a meningioma. Surgical excision and subsequent pathologic evaluation revealed an angioleiomyoma and the patient recovered without incident. Angioleiomyomas are classified as benign smooth muscle tumors and are classically seen in adult females arising in the soft tissues of the lower extremities. Although rare, these masses have been described in various intracranial locations, usually extra-axially. A comprehensive review of the literature and discussion are provided, emphasizing histopathologic and imaging features of this uncommon intracranial neoplasm.

CASE REPORT

CASE REPORT

A 43-year-old right-handed Caucasian man with past medical history significant for diabetes mellitus arrived at the emergency department after becoming light-headed while doing yard work at home. The patient was found by his wife who reported that the syncopal episode had lasted approximately 5- 10 minutes. As per the wife, the patient was initially slow to respond but was alert and oriented within seconds of waking up.

At the emergency department, the patient was found to be neurologically intact on exam. A non-contrast head computed tomography (CT) was performed that demonstrated an extra-axial, non-calcified, hyperdense (average of 50 Hounsfield units), solid mass based at the anterior falx cerebri, without discernible surrounding edema (Figure 1). The most probable diagnosis at this point was thought to be a falcine meningioma or, less likely, an aneurysm.

The patient was then transferred to a tertiary- care center for further diagnostic evaluation and neurosurgery was consulted for management. Magnetic resonance imaging

(MRI) of the brain with and without contrast showed a well-defined, interhemispheric, soft tissue mass centered at the anterior portion of the falx, measuring 16 mm in greatest dimension, extending slightly to the right of midline. The mass was rounded and slightly irregular in shape and was found to be isointense on T1, hyperintense on T2. Enhancement was homogeneous and there was no adjacent parenchymal edema (Figure 2). The lesion demonstrated minimal mass-effect on neighboring structures. No aneurysms or vascular malformations were identified. With the MRI a diagnosis of parafalcine meningioma was proposed and surgical excision was recommended by the neurosurgery team.

Under general anesthesia, using navigation assistance, a bifrontal craniotomy was performed. The lesion was approached microscopically from the right, along the falx. The lesion was found to be intimately associated with the falx. Macroscopically, the tumor appeared well- circumscribed, did not adhere to the underlying brain parenchyma, was hypervascular, and its surface showed multiple 1-3 mm nodules (Figure 3). The mass bled readily with manipulation, although this was easily controlled with bipolar cautery. Gross

total resection was achieved and the patient had an uneventful postoperative course.

Pathology revealed a 1.0 x 0.6 x 0.4 cm gray-reddish, well-circumscribed mass composed of irregular vascular spaces and nodular fibrous tissue compatible with an angioleiomyoma on hematoxylin and eosin (H&E) stains. Immunohistochemical staining supported this diagnosis (Figure 4). The histopathological features of the mass were most consistent with the solid subtype with few intervening dilated vascular channels.

DISCUSSION

Etiology & Demographics:

According to the most recent soft tissue tumor classification developed in 2013 by the World Health Organization (WHO), angioleiomyomas, also known as angiomyomas and vascular leiomyomas, are considered benign smooth muscle tumors. Overall, angioleiomyomas are relatively uncommon neoplasms, accounting for around 4.4% of benign soft tissue tumors [1]. The exact etiology of these tumors is unclear, although infection, trauma, venous stasis and hormonal factors have been suggested. Furthermore, lipometaplasia detected within the mass may indicate a hamartomatous nature [2- 4]. The majority of these lesions are solitary, slow-growing masses, and patients most commonly present with pain as their chief complaint.

Histopathologically, these neoplasms are comprised of mature smooth muscle interwoven with vascular channels, and can be further subdivided into three types: solid, venous and cavernous. The solid subtype is composed of compact smooth muscle with multiple small vascular channels. Venous angioleiomyomas are made up of loosely organized smooth muscle bundles that intertwine with the thickened muscular walls of venous channels. Lastly, the cavernous subtype is characterized by small amounts of smooth muscle bundles and a predominance of dilated vascular channels [1].

Although capable of arising virtually anywhere in the body, angioleiomyomas classically occur in the soft tissues of the lower extremity within the subcutaneous or deep dermal layers. The peak incidence is between the fourth and six decades of life and, when occurring in the lower extremities, these tumors show a slight female predominance [1].

On the other hand, intracranial angioleiomyomas are uncommon and, unlike their soft tissue counterparts in the lower extremities, exhibit a male to female predominance of 4:1 [5]. To the best of our knowledge, approximately fifteen cases of intracranial angioleiomyomas have been reported in the literature. Although the solid subtype usually accounts for approximately 66% of cases in the lower extremities, intracranial angioleiomyomas are more commonly of the venous or cavernous variety [1].

Clinical & Imaging findings:

In a review of the literature performed by Sun et al. [6], eight out of twelve patients in their search, as well as one out

of their three personal case reports, complained of headache at presentation. As with the case being presented, it is unclear if non-specific neurological symptoms such as headache and dizziness are directly related to the mass, especially when the lesions are small and show no significant mass-effect on underlying structures. Likely, these symptoms trigger brain imaging that serves to detect ALM, possibly as an incidental finding. Other reported common clinical symptoms included seizure, visual disturbances, and muscle weakness. In contrast, focal neurological symptoms are generally related to mass-effect from the lesion on neighboring structures. The majority of masses exhibit an extra-axial origin, including sellar, supratentorial and posterior fossa locations. Generally, ALMs occur near large venous structures such as dural sinuses or the cavernous sinus, as was the case with our patient, with the mass in close proximity to the inferior sagittal sinus. In keeping with the classically described small (< 2 cm), round, well-defined, gray-white or brown macroscopic appearance of these lesions, our case showed a well-circumscribed, gray-reddish mass. Generally, these masses are isodense on CT, unlike this case, which showed mild hyperdensity on non-contrast CT. As mentioned previously, the majority of intracranial ALMs are venous or cavernous subtypes, which histologically show loosely organized smooth muscle. In contrast, this case was classified as a solid subtype, composed of compact smooth muscle, which may account for its greater density on non-contrast CT. Post-contrast CT typically exhibits small nodular enhancement [6- 8].

On MRI, ALM show intermediate signal intensity on T1-weighted images, high signal intensity on T2-weighted images and display what some authors have described as a “flame-like” appearance on T1 post-gadolinium images [5]. This pattern of contrast enhancement refers to enhancement arising centrally from the base of the tumor and ultimately filling out peripherally on more delayed images. Although described as a possible diagnostic clue to distinguish ALM from other extra-axial enhancing masses, this particular feature was not apparent in the present case. This is thought to be due to the timing on post-contrast images, which occurs approximately five minutes following contrast injection. Imaging the patient at a later phase, when contrast filling within the tumor had already occurred, possibly accounted for a homogeneous contrast enhancement pattern. Uncommon imaging findings include an intraparenchymal origin and presence of draining veins [6, 7, 9-11]. Although apparently highly vascular in nature, no associated intraparenchymal or extra-axial hemorrhage has, to our knowledge, been reported.

Treatment & Prognosis:

Prognosis is typically favorable, with surgical resection being generally sufficient for treatment and postoperative recurrence considered exceedingly rare [7]. Occasionally, depending on the location of the mass, complications related to mass effect may occur. Several complications affect ALM including hydrocephalus, seizure and visual impairment, mainly when located in the sellar and suprasellar region [7, 9-12]. Furthermore, due to the proximity of these lesions to large venous structures, surgical resection carries an increased risk of intraoperative hemorrhage.

Differential Diagnoses:*Meningiomas*

Angioleiomyomas can often mimic other more common well-defined, extra-axial masses. Differential diagnoses include meningiomas, schwannomas, dural metastases, and aneurysms. Meningiomas are classically well-circumscribed, extra-axial masses, which often occur in the supratentorial compartment and may exhibit a dural tail and adjacent hyperostosis [9, 10]. Approximately 60% of meningiomas are hyperdense on non-contrast CT, and the rest are usually isodense. Furthermore, CT may aid in identifying varying degrees of associated calcification within the mass, which may occur in 20-30% of cases [10- 14]. On MRI, meningiomas tend to be isointense on T1 and iso- to hyperintense on T2. Their enhancement is classically intense and homogeneous on both CT and MRI. Meningioma was proposed as the most probable diagnosis in our case due to the location, CT features and signal intensity on MRI, yet unlike many meningiomas, no discernable dural tail or internal calcifications were evident [10, 13-14].

Dural metastasis

Dural metastasis may be another differential diagnosis to consider; nevertheless, the age of the patient, absence of known primary cancer, and the solitary nature of the lesion made this diagnosis less likely in the present case. Dural metastases can be solitary or multiple and vary greatly in imaging characteristics, according to the primary tumor in question and whether or not there is associated hemorrhage [10, 14]. On CT, dural metastases tend to be hypodense to mildly hyperdense, unless the primary tumor is a colonic adenocarcinoma, in which case the lesions are typically hyperdense [9]. MRI signal intensity and contrast enhancement is also variable, although generally hypointense on T1, hyperintense on T2 and homogeneously enhancing [10].

Schwannomas

Although schwannomas can be considered a possibility when extra-axial hyperdense masses are identified, the location of the tumor in our case is atypical for a schwannoma. Whereas schwannomas classically arise along cranial nerves when present intracranially, the tumor in our case originated along the falx in the interhemispheric region of the frontal lobes. Schwannomas rarely calcify, are generally solitary and can bleed in approximately 5% of cases [9-10, 15]. On CT imaging, these masses are generally iso- to hypodense, and on MRI tend to be T1 isointense and T2 hyperintense. While small schwannomas commonly exhibit intense homogeneous enhancement, larger tumors may show cystic degeneration and heterogeneous enhancement.

Saccular intracranial aneurysms

Saccular intracranial aneurysms account for the majority of intracranial hemorrhage and classically arise from the anterior circulation, specifically from the anterior cerebral artery/ anterior communicating artery complex. These well-defined, extra-axial vascular lesions can have varying CT and MRI characteristics, depending on the presence of internal thrombosis. Patent aneurysms are generally isodense on CT, show a flow void on T2-weighted images, are usually

hypointense on T2 and exhibit homogeneous contrast enhancement. On the other hand, thrombosed aneurysms may appear slightly hyperdense on CT, have heterogeneous signal intensity on T1 and T2, depending on the age of the clot, and can present a filling defect with peripheral enhancement on post-contrast images [10]. The location for the lesion in our case would correspond to a distal anterior cerebral artery aneurysm, which is uncommon.

Although generally uncommon, angioleiomyomas can be considered as a differential diagnosis of well-defined, solitary, enhancing, extra-axial masses. This is especially true when the post-contrast images show an enhancement pattern that begins at the central base region of the mass and gradually extends peripherally.

TEACHING POINT

Angioleiomyomas are benign smooth muscle tumors that classically arise in the lower extremities in adult females, although can develop anywhere in the body. When these tumors occur intracranially, they are usually extra-axial, well-defined and show significant homogeneous enhancement, occasionally beginning centrally at the base of the mass and extending outward in a “flame-like” pattern.

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FIGURES

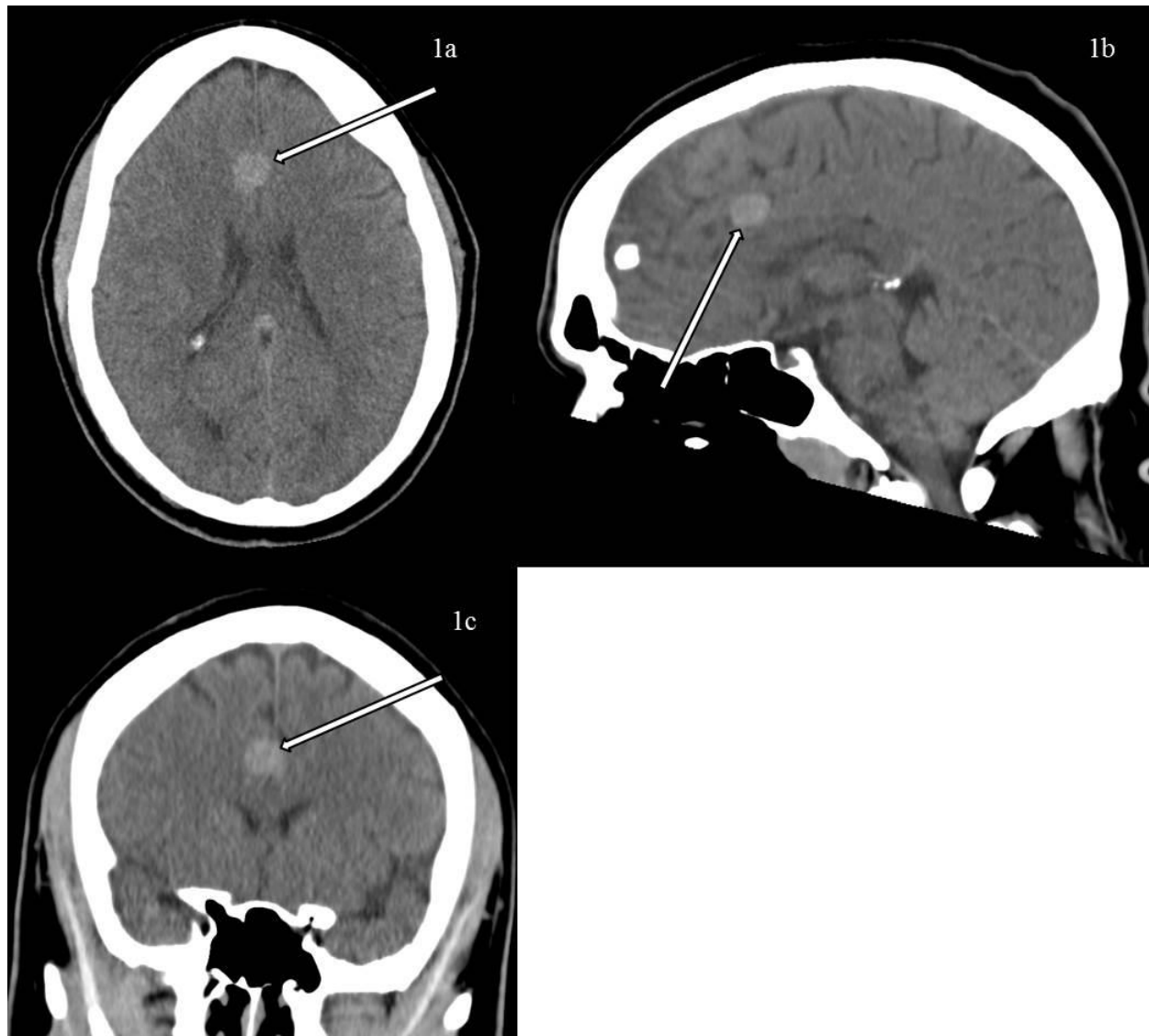


Figure 1: 43-year-old male with an angioleiomyoma of the falx. FINDINGS: Non-contrast-enhanced axial CT of the brain (1a), with sagittal (1b), and coronal (1c) reconstructions show a well-defined, hyperdense mass (arrows), with an average of 50 Hounsfield units, located at the anterior falx cerebri, slightly to the right of midline, without discernable edema of the adjacent frontal lobes. TECHNIQUE: Axial CT of the brain, 120 kV, 2.0 mm slice thickness, no intravenous contrast was administered

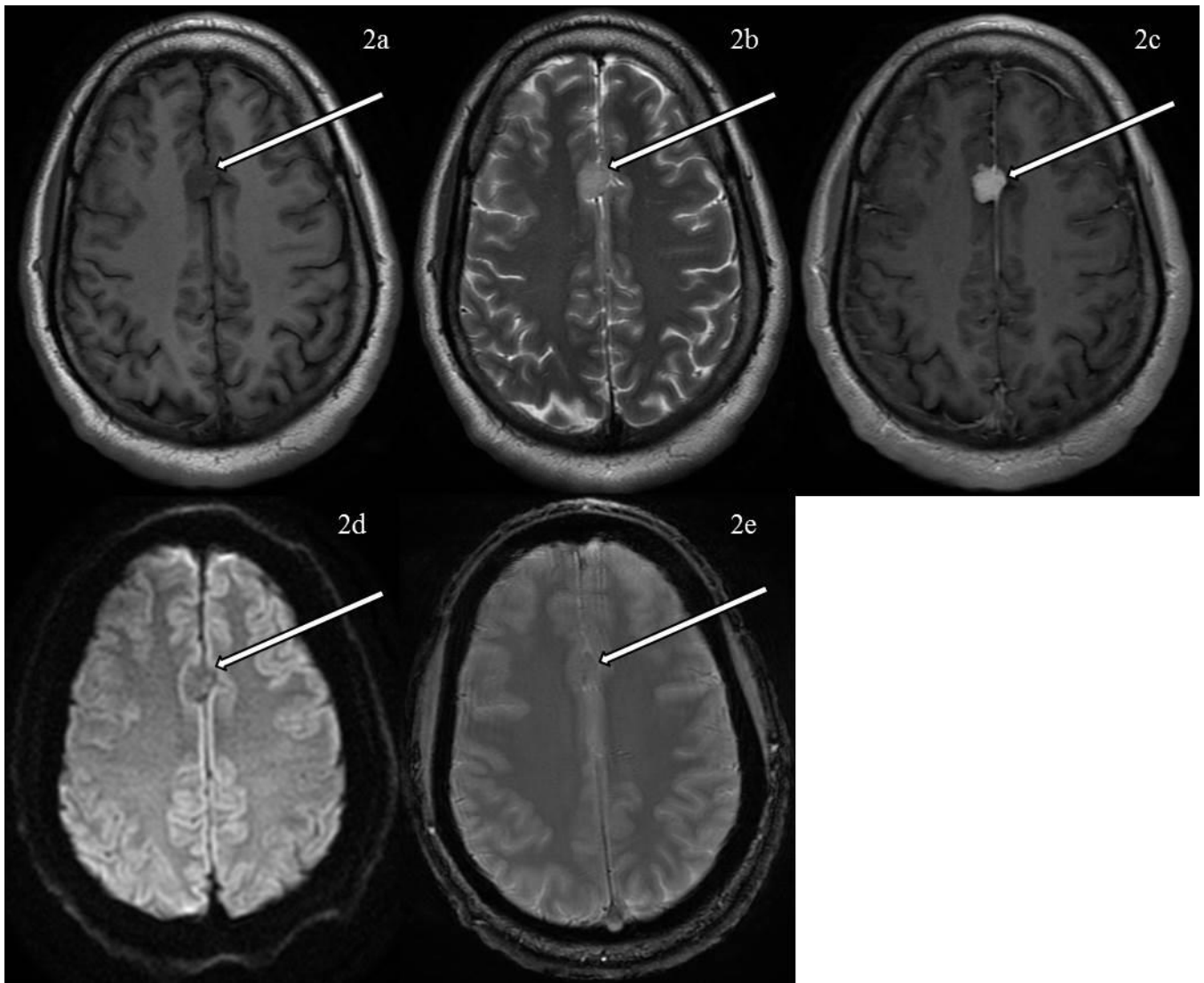


Figure 2: 43-year-old male with an angioleiomyoma of the falx.

FINDINGS: Axial MRI of the brain demonstrates a well-defined, extra-axial, interhemispheric mass, centered at the anterior aspect of the falx cerebri, slightly to the right of midline. The mass is isointense on T1 FLAIR (2a), hyperintense on T2 (2b), shows homogeneous enhancement on T1 FLAIR post-contrast images (2c), does not exhibit restricted diffusion on diffusion-weighted images (2d), and does not show susceptibility artifact on T2* (2e).

TECHNIQUE: 3.0 T MRI of the brain with and without contrast

Parameters:

T1 FLAIR: TR= 2322 ms, TE= 8 ms, slice thickness= 5.0 mm, field of view= 512 x 512

T2: TR= 4000 ms, TE= 130 ms, slice thickness= 5.0 mm, field of view= 512 x 512

T1 FLAIR post-contrast: TR= 2322 ms, TE= 8 ms, slice thickness= 5.0 mm, field of view= 512 x 512, intravenous injection of 20 cc of OmniScan® (gadodiamide)

DWI: TR= 9000 ms, TE= 92 ms, slice thickness= 5.0 mm, field of view= 256 x 256

T2*: TR= 600 ms, TE= 15 ms, slice thickness= 5.0 mm, field of view= 512 x 512

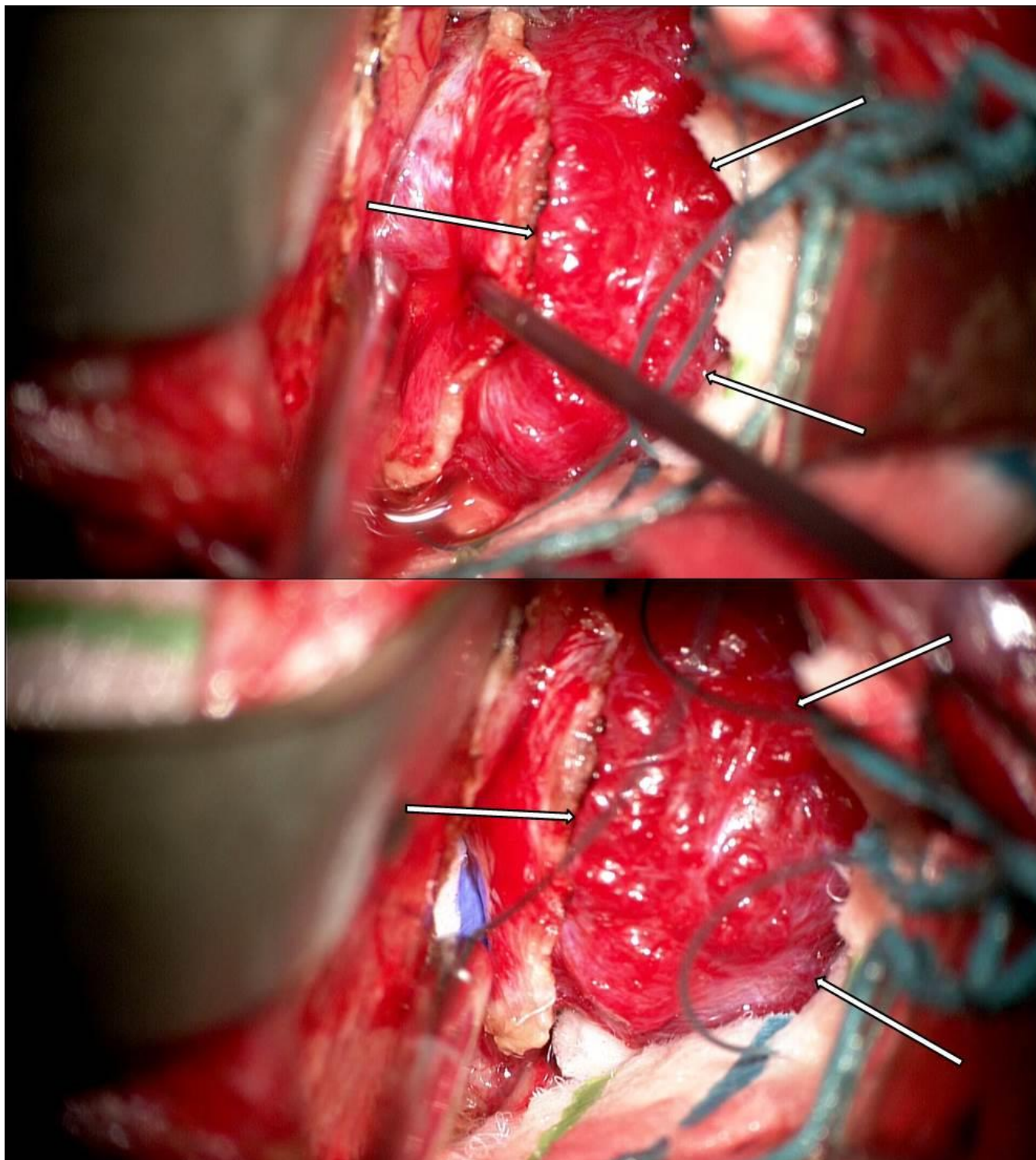


Figure 3: 43-year-old male with an angioleiomyoma of the falx.

FINDINGS: Intraoperative photographs show a well-circumscribed, hypervascular-appearing mass, with a slightly nodular surface, adjacent to the falx, without adherence to the underlying brain parenchyma (arrows).

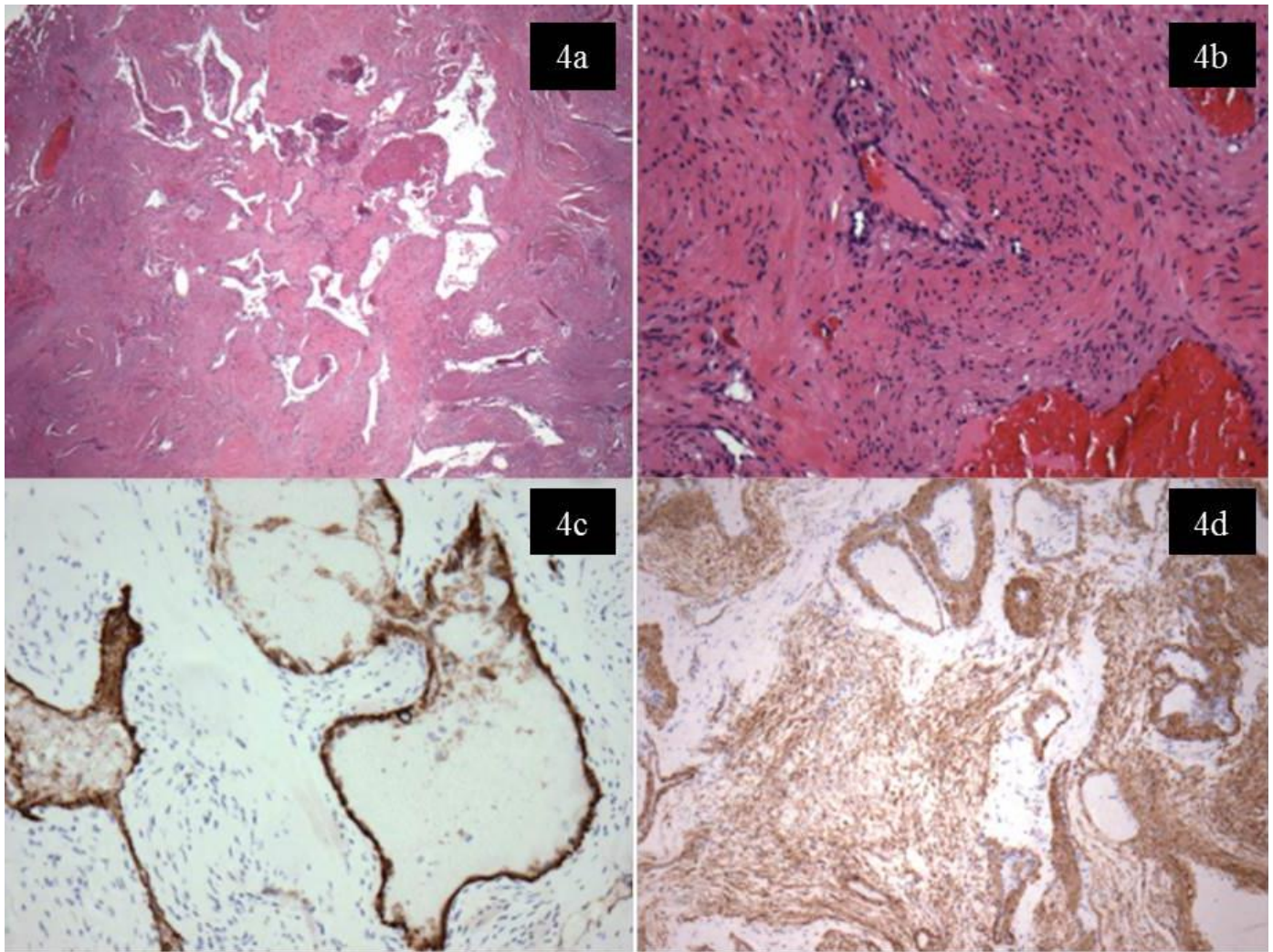


Figure 4: 43-year-old male with an angioleiomyoma of the falx.

H&E staining shows tumor tissue composed of large blood vessels and intervening spindle cell regions (4a). H&E staining at higher power shows small vessels that are noted to have muscular walls with intervening spindle cells appearing similar to the cells in the vascular walls (4b). CD31 immunohistochemistry highlights the endothelial cells lining the muscular vessels (4c). Immunohistochemistry for smooth muscle actin demonstrates strong staining in the muscular vessel walls as well as in the intervening spindle cells (4d).

Etiology	Although unclear, minor trauma, hormonal factors and venous stasis are proposed mechanisms.
Incidence	4.4% of benign soft tissue neoplasms, although intracranial variant is uncommon
Gender ratio	Generally more common in females, however, intracranial variant more likely in males
Age predilection	Between the fourth and sixth decades of life
Risk factors	Unclear
Treatment	Surgical excision
Prognosis	Generally carries a good prognosis, with surgical resection usually curative.
Imaging findings	CT: Isodense on non-contrast images and nodular enhancement following contrast administration. MR: T1 isointense, T2 hyperintense, enhancement progresses from the center of the mass outward

Table 1: Summary table for Angioleiomyoma of the falx.

Differential Diagnosis	General	CT	MRI	Enhancement
Angioleiomyoma	Well-circumscribed, usually extra-axial	Isodense	T1- isointense T2- hyperintense	Enhancement beginning centrally within the mass and extending peripherally
Meningioma	Well-circumscribed, extra-axial, usually supratentorial, dural tail, possible adjacent hyperostosis	Usually hyperdense although can be isodense, Internal calcifications in approximately 20-30%	T1- isointense, T2- isointense or hyperintense	Intense, homogeneous
Dural metastasis	Solitary or multiple, extra-axial, can present a dural tail	Hypodense to mildly hyperdense, Typically hyperdense if secondary to colonic adenocarcinoma	Variable signal intensity according to primary tumor characteristics and secondary hemorrhage, usually hypointense on T1 and hyperintense on T2	Variable according to primary tumor, usually intense and homogeneous
Schwannoma	Well-circumscribed, calcification is rare, hemorrhage present in 5%, most are solitary, usually arise along cranial nerves when intracranial	Isodense to hypodense	T1- isointense T2- Hyperintense	Typically homogeneous and intense when small, may show heterogeneous enhancement when large
Saccular cerebral aneurysm	Well circumscribed, extra-axial, round, usually arise from the anterior circulation (majority from ACA/ACoA complex)	Depends on presence of intraaneurysmal thrombus, if present the lesion appears slightly hyperdense, may present with calcification	T1- may show flow void if patent, heterogeneous signal intensity if thrombosed T2- usually hypointense	Intense and homogeneous if patent, rim enhancement with central filling defect if thrombosed

Table 2: Differential diagnosis table for Angioleiomyoma of the falx.

ABBREVIATIONS

ALM = angioleiomyoma
 CT = Computed tomography
 H&E = Hematoxylin and eosin
 MRI = magnetic resonance imaging
 WHO = World Health Organization

KEYWORDS

Angioleiomyoma; falx; dural; tumor; benign; extra-axial; neoplasm; intracranial; computed tomography; CT; magnetic resonance imaging; MRI; angiomyoma; vascular leiomyoma; angioma; falx

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