Infratentorial angioleiomyoma: a new location for a rare neoplastic entity

Case report

JAIME GASCO, M.D.,1 BRODUS FRANKLIN, B.S.,1 LEONARDO RANDEL-CASTILLO, M.D.,1 GERALD A. CAMPBELL, M.D., PH.D.,2 MAHMOUD ELTORKY, M.D., PH.D.,2 AND PAUL SALINAS, M.D.1

1Division of Neurological Surgery; and 2Department of Pathology, Division of Neuropathology, University of Texas Medical Branch, Galveston, Texas

Angioleiomyomas are benign neoplasms most often located in the subcutaneous tissue of middle-aged individuals and usually confined to the subcuticular and deep dermal layers of the lower extremities. An intracranial site for this tumor is exceedingly rare, with very few reports documenting locations in the neuraxis. To the authors' knowledge the present case represents the first reported instance of an infratentorial angioleiomyoma. The authors conducted a review of selected English-language papers published since 1960 describing well-documented cases of intracranial vascular leiomyomas, with detailed information on the clinical presentation, radiology, pathology, and particulars of surgical management in each case. (DOI: 10.3171/2008.8.17645)

KEY WORDS • angioleiomyoma • angiomyloma • cerebellar tumor • smooth muscle tumor • vascular leiomyoma

Case Report

History and Examination. This 43-year-old left-handed man was transferred to the emergency department of the John Sealy Hospital, University of Texas Medical Branch, with symptoms of an acute-onset headache, blurred vision first noticed while reading, dizziness, and gait abnormalities and instability. His medical history was significant for a mild traumatic head injury he had incurred during a motor vehicle accident 2 months prior to the onset of his presenting symptoms. Computed tomography scanning and subsequent MR imaging of the brain at that time incidentally revealed a mass in the left cerebellar hemisphere. The patient was offered neurosurgical intervention, but he refused given a reported lack of symptomatology. (In retrospect and following surgical intervention at our institution, the patient admitted to having experienced acute intermittent episodes of frontal headaches for 1 year prior to the incidental finding, with episodes lasting ~ 12 hours.)

A neurological examination at our institution was significant for left upper-extremity dysmetria on finger-to-nose testing, left upper-extremity dysdiadochokinesia on testing of rapidly alternating movements, and mild gait ataxia. In the cognitive-affective domain, he exhibited a decreased attention span, depression, and emotional blunting.

A noncontrast head CT revealed a 4.4 × 3.2–cm isodense lesion without surrounding edema or mass effect and a patent fourth ventricle without evidence of obstructive hydrocephalus (Fig. 1A). Magnetic resonance imaging further delineated the lesion as a 4.4 × 3.9 × 3.9–cm mass with broad dural contact, appearing isoin-
transverse sinus. D: Preoperative axial FLAIR image revealing a lesion with extensive dural content and in proximity to the obtained after GTR. C: Preoperative sagittal T1-weighted MR image frenchnema. Note the patent fourth ventricle. B: Axial noncontrast CT with homogeneously increased density compared with surrounding pa-channels. within the tumor mass, in retrospect, could be interpreted as vascular ing no significant edema or mass effect. Some areas of hypointensity within the tumor mass, in retrospect, could be interpreted as vascular channels.

tense on T1-weighted sequences, hyperintense on T2-weighted sequences, and homogeneously enhanced after the administration of Gd. Thus, the lesion was originally interpreted as a possible meningioma pending histological confirmation (Fig. 1C and D).

Operation. The patient underwent a left suboccipit-al craniotomy for tumor resection and histological identifica-tion of the mass by pathology. On elevation of the bone flap during gross inspection the lesion was visible underneath the dura mater, revealing an abnormally dark red appearance of underlying tissue in the superolateral aspect of the craniotomy site. Once the dura was opened in a cruciate fashion, the tumor appeared adherent to the confluence of the left transverse and sigmoid sinuses, and the mass was otherwise embedded within surrounding parenchyma. The lesion had an encapsulated plane of dissection and was easily freed circumferentially from surrounding cerebellar tissue. Once freed from the dura at the superolateral margin, the tumor was removed in 3 separate specimens. On direct inspection of the dural attachment, the lesion appeared to infiltrate the dura as well as the left transverse sinus. The surface of the oval mass appeared red-tan with a gelatinous consistency. Hemosta-sis was achieved, and the dural defect was repaired with a porcine duraplasty supplement—that is, the Durasis dural substitute (Cook Medical) and Tisseel VH fibrin sealant (Baxter Healthcare).

Postoperative Course. The patient’s postoperative course was completely uneventful, and CT confirmed complete resection of the lesion (Fig. 1B). Microscopically, the tumor was discrete but lacked a definitive capsule. Cerebellar tissue was not present in the resection. The tumor was primarily composed of ectatic thick-walled vascular channels intermingled with thin fusiform stromal cells arranged irregularly in a circumferential manner in sparsely cellular interlacing bundles between and around the vessels. Atypia, pleomorphism, and mitoses were absent. The stromal cells evidenced prominent cytoplasmic acidophilia with indistinct cellular margins and uniform, thin elongated nuclei, all characteristic of smooth muscle cells. Special stains (Movat and Masson trichrome) revealed both collagen and muscle in the stroma and the complete absence of elastin. Immunohistochemistry evidenced a positive reaction to smooth muscle actin (M0851, Dako Corp.), the absence of reactivity for epithelial membrane antigen (M0613, Dako Corp.), and positive CD-34 reaction (326-01, Covance) for endothelial cells only. The proliferation index based on Ki 67 (M7240, MIB-1, Dako Corp.) nuclear reactivity was much less than 1% (Fig. 2). The immunohistochemical reaction for the Epstein-Barr virus was negative. All immunohistochemistry was pro-cessed using an antigen retrieval system (Target Retrieval Solution, Dako Corp.) and Dako Autostainer (LSAB-2).

Discussion

Angioleiomyoma is a benign tumor composed of smooth muscle and endothelium in which the predominant feature is an abundant number of vascular channels sepa-rated by stroma composed of loosely organized smooth muscle bundles with variable amounts of collagen, but not elastin. Surrounding the vascular lumens, the smooth muscle fibers are organized in a regular circumferential pattern, whereas in the intervening stroma they are more disordered.17 Based on the histological assessment of the relative proportions of its components (smooth muscle, vascular, and fibrous), the lesion can be further subdivided into 3 subtypes: solid or capillary (many small vascular spaces), cavernous (dilated vascular spaces and little intervening smooth muscle), and venous (vascular spaces surrounded by thick muscular walls).5,13 The lesion in the case reported here corresponds to the venous subtype. Angioleiomyoma is generally considered to be a variant of leiomyoma,16 although the presence of mature adipocytes in the stroma in a small subset of cases has suggested a hamartomatous origin to some authors.10

The first case series involving angioleiomyomas was described by Stout16 in 1937 after a review of some 110 similar cases. Since then, other reports have followed: in 1959 by Montgomery and Winkelmann12 as well as Duhig and Ayer,1 and in 1961 by Magner and Hill.11 The largest series of leiomyomas—562 cases—was reported by Hachisuga et al.5 None of these studies included intracranial angioleiomyomas.

According to Hachisuga et al.,5 these benign lesions are found mainly in middle-aged individuals, mostly fe-males (ratio 1.7:1), and the preferred locations are the sub-cutaneous tissue of the lower extremities (89%), followed by the head and neck region (48%) and the trunk (14%). In their series of 562 cases of angioleiomyoma, only 2

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patients had recurrences and none had evidence of malignant transformation or died as a result of this tumor within the limits of the patient follow-up.

An intracranial location of this tumor is extremely unusual. The first intracranial leiomyoma was reported in 1968 by Kroe et al., who described an intrasellar case. In 1994 Lach et al. described the first primary intracranial angioleiomyoma, in an extraaxial location (leptomeningeal). To the present time, 4 additional intracranial cases have been documented; of these, only the case reported by Ravikumar et al. was intraaxial (basal ganglia). Karagama et al. and Kohan et al. have verified lesions in the IAM. Figueiredo et al. have described a lesion in the cavernous sinus. Thus, our case is the first reported instance of an angioleiomyoma in an infratentorial location that extends into and compresses the brain (cerebellum). Like all but 1 of the previously reported cases, the lesion in our patient arises from an extraaxial (meningeal) origin. Table 1 summarizes the clinical, radiological, and surgical findings in all of the previously reported cases as well as our own findings.

All reported cases of intracranial angioleiomyomas have been managed by surgical intervention. In all 6 cases, including ours, GTR was achieved. An easily identifiable plane of dissection and the lack of major feeding arteries may explain such success. Dural attachment varies and is significant in some cases, almost mimicking that with a lesion of meningeal origin. The presence of a large draining vein was seen in only 1 case. In the other 5 cases, the tumor blood supply (arterial and venous) did not seem to impose difficulties during dissection.

Overall, these lesions are paucisymptomatic, possibly because of their benign histopathology and scant cellular proliferation index, underscoring a slow proliferation rate.

Fig. 2. A: Photograph of the cut surface of the mass showing numerous blood-filled spaces separated by thick irregular septae. B: Low-magnification photomicrograph further delineating the overall pattern of irregular vascular spaces, some containing blood and surrounded by loose, concentric lamellar tissue in the septae. C: Photomicrograph demonstrating the presence of collagen (yellow) in the central regions of the septae and the total absence of elastin (black) in the walls. D: Photomicrograph demonstrating the presence of smooth muscle in the concentric lamellae surrounding the vascular lumens, made evident by immunohistochemistry using an antibody against smooth muscle actin. E: Higher-magnification photomicrograph clearly showing the immunoreactive thin, elongated muscle cells with bland nuclear morphology in the walls near the lumens. F: Photomicrograph revealing that CD34, an endothelial cell marker, reacts only with the endothelial cells lining the lumens and not with the stroma. H & E (B), Movat stain (C), and immunoperoxidase (D, E, and F). Bar = 0.5 cm (A), 200 μm (B), 100 μm (C), 50 μm (D and F), and 25 μm (E).
Infratentorial angioleiomyoma

According to our review, symptoms depend mostly on location, although headaches (3 of 6 cases), visual symptoms (3 of 6 cases), and ataxia (2 of 6 cases) are the most frequently reported clinical features (Table 1). Despite the vascular nature of these lesions, hemorrhagic presentation has not been described to date. The association of these tumors with immunodeficiency, in particular, AIDS and Epstein-Barr virus, deserves consideration. Although the HIV status of our patient is unknown, his immunohistochemical reaction for Epstein-Barr virus was negative. Antibody testing for these viral entities should probably be considered once a diagnosis has been confirmed.

Other examples of leiomyomas that deserve consideration and are important for an overall neurosurgical perspective include one reported by Steel et al.15 in 1993 describing a spinal epidural location and those found in the orbit (22 well-documented cases) according to the recent review presented by Arat et al.1 Intraorbital angioleiomyomas are also rare; the first reported case was described by Henderson and Harrison6 in 1970. Since then, only one case has been reported.1 Notably, none of the patients with intraorbital lesions has presented with intracranial extension.

This lesion generally appears as an isodense lesion on CT and homogeneously enhances with intravenous contrast administration. The CT-weighted images are usually isointense and the T2-weighted are hyperintense, also with uniform enhancement following Gd administration. The physiopathological explanation for atypical imaging findings such as the one reported by Ravikumar et al.14—that is, enhancing mural nodule with cyst formation—remains unclear. The presence of a large draining vein visible intraoperatively and in retrospect on imaging has only been described by Lach et al.10 The differential diagnosis of lesions with a similar imaging appearance is broad, and cavernous hemangiomas, meningiomas,

### TABLE 1: Literature review of studies on intracranial angioleiomyomas*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Patient Age (yrs), Sex</th>
<th>Lesion Size (cm) &amp; Location</th>
<th>Type &amp; Duration of Signs/Symptoms (mos)</th>
<th>Imaging Procedure &amp; Results</th>
<th>Surgical &amp; Postop Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lach et al., 1994</td>
<td>47, M</td>
<td>2 × 2 × 2; parietal lobe, leptomeninges</td>
<td>gait abnormality, rt-sided parkinsonism; 14</td>
<td>CT: homogeneous enhancement, no mass effect, large draining vein</td>
<td>approach not specified, GTR, no dural attachment, clear plane of dissection, large draining vein</td>
</tr>
<tr>
<td>Ravikumar et al., 1996</td>
<td>12, F</td>
<td>NA; caudate nucleus</td>
<td>diplopia, headache, seizures (1.5), Lt hemidystonia, eyelid apraxia; 2</td>
<td>CT: hypodense lesion &amp; mass effect, enhancing mural nodule w/ cyst, secondary hyperdense lesion in Lt globus pallidus (untreated)</td>
<td>frontal craniotomy, lesion well circumscribed, adjacent to lat ventricle, GTR, 1 yr FU, no recurrence, untreated lesion remained quiescent at FU</td>
</tr>
<tr>
<td>Kohan et al., 1997</td>
<td>NA</td>
<td>NA; IAM</td>
<td>sensorineural hypoacusis/tinnitus; NA</td>
<td>hyperdense, homogeneously enhancing, bone erosion, no surrounding edema; T1: isointense; T2: hyperintense; post-Gd: homogeneous enhancement, ipsilat carotid artery &amp; optic chiasm displacement</td>
<td>suboccipitalretsosigmoid approach, GTR</td>
</tr>
<tr>
<td>Figueiredo et al., 2005</td>
<td>52, M</td>
<td>6 × 6 × 5; cavernous sinus</td>
<td>diplopia, headache, facial numbness (6), impaired visual acuity (6); 24</td>
<td>hyperdense, homogeneously enhancing, bone erosion, no surrounding edema; T1: isointense; T2: hyperintense; post-Gd: homogeneous enhancement, ipsilat carotid artery &amp; optic chiasm displacement</td>
<td>frontotemporal/pretemporal approach, complete resection, soft mass, blush red, no reported dural attachment, ipsilat ICA encasement by tumor</td>
</tr>
<tr>
<td>Karagama et al., 2005</td>
<td>47, F</td>
<td>1; IAM</td>
<td>sensorineural hearing loss; 12</td>
<td>T1: isointense; T2: hyperintense; post-Gd: homogeneous enhancement</td>
<td>translabyrinthine approach, complete resection</td>
</tr>
<tr>
<td>present study</td>
<td>43, M</td>
<td>4.4 × 4.4 × 4; cerebellar hemisphere</td>
<td>blurry vision, ataxia (acute), headache; 12</td>
<td>CT: hyperdense, no edema; T1: isointense; T2: hyperintense; post-Gd: homogeneous enhancement</td>
<td>suboccipital approach, complete resection, dural attachment, easily identifiable dissection plane, multiple small feeding/draining vessels</td>
</tr>
</tbody>
</table>

* FU = follow-up; ICA = internal carotid artery; NA = not available.
schwannomas, and neurofibromas as well as other well-encapsulated masses can be difficult to differentiate exclusively by imaging.

Postresection tumor shrinkage was observed in our case. Using the ellipsoid approximation, we determined that the tumor mass was 6.1 cm³. The sum of the volumes of the 3 specimens submitted to pathology generated a value of 5.1 cm³ using the same method. This value represents a volumetric reduction of almost 17%. A possible explanation is the absence of blood flow through the multiple vascular channels of the tumor once the specimen was obtained. The fact that the volumetric size of the resected lesion did not exactly match the measurement provided by MR imaging is an indicator of how vascular flow can affect and overestimate the volume calculation. Thus far, no reports of spontaneous or intraoperative intracerebral hemorrhage have been described in cases of angioleiomyomas. Notably, even though the case presented by Kroe et al.⁹ was purely a leiomyoma, its surgical management involved the hemorrhagic rupture of the capsule and contents that was amenable to intracavitary suctioning. Despite this lack of evidence, caution should lead neurosurgeons to an awareness of this potential hemorrhagic risk, especially if during the preoperative assessment the possibility of angioleiomyoma is contemplated.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References


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Address correspondence to: Jaime Gasco, M.D., Division of Neurological Surgery, University of Texas Medical Branch, 301 University Boulevard, Galveston, Texas 77555-0517. email: jagasco@utmb.edu.