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ABSTRACT

SUMMARY: Vascular venous malformations of the cavernous sinus have multiple imaging features that can be used to distinguish them from other entities in the region. Accurate identification of these lesions is essential: Vascular venous malformation lesions carry considerable risk of intraoperative hemorrhage, so preoperative recognition of vascular venous malformations can greatly impact the treatment strategies used. Nevertheless, because of their scarcity, many radiologists are unfamiliar with the radiologic and clinical features of cavernous sinus vascular venous malformations. This article will describe a case of an asymptomatic vascular venous malformation; outline its imaging, clinical, and pathologic features; and review the relevant literature regarding this diagnosis.

ABBREVIATIONS: CCM = cerebral cavernous malformation; ISSVA = International Society for the Study of Vascular Anomalies; RBC = red blood cell; VM = vascular venous malformation

A 50-year-old man with no notable medical history presented to our institution with a 2-year history of bilateral nonpulsatile tinnitus. The patient's work-up included an audiogram that demonstrated asymmetric left-sided hearing loss. This prompted evaluation with internal auditory canal MR imaging, which incidentally discovered a mass in the left cavernous sinus.

Imaging and Biopsy

The initial MR imaging noted a well-circumscribed mass centered in the left cavernous sinus, which encroached on the left aspect of the sella. The mass encased the left ICA, which was deviated medially. Intralesional signal was hyperintense on FLAIR and isointense to gray matter on T1WI. On T2 sampling perfection with application-optimized contrasts by using different flip angle evolution (SPACE sequence; Siemens), the lesion was notably heterogeneous, with a somewhat coarsened texture. Intralesional enhancement was also heterogeneous (Fig 1).

Initially, the differential for this lesion included meningioma, schwannoma, and cavernous sinus hemangioma. However, there was also some concern that this represented a chondroid lesion, such as a chondrosarcoma. The neurosurgical team was consulted, and they planned to resect the mass via a craniotomy with cavernous sinus exploration. The preoperative stereotactic CTA revealed findings extremely atypical of chondrosarcoma. First, the mass lacked an intralesional chondroid matrix. Also, the adjacent osseous structures appeared remodeled rather than eroded, suggestive of a benign and long-standing process (Fig 2).

Two additional imaging examinations were then performed to evaluate the mass for features of a vascular venous malformation (VM). First, a technetium Tc99m-tagged red blood cell (RBC) scan was completed, which demonstrated avid radiotracer accumulation within the mass on delayed images. Then, a dynamic contrast-enhanced MR imaging was performed, which demonstrated progressive enhancement of the mass on sequential images (Fig 3). Altogether, the imaging was highly suggestive of a VM. The patient ultimately underwent a biopsy of the mass for pathologic confirmation. Complete resection was not attempted due to the patient's preoperative lack of symptoms and the known favorable response of such lesions to focal treatment with stereotactic radiation.

Pathology

The biopsy from the left cavernous sinus showed a collection of predominantly thin-walled, large-caliber, anastomosing vessels within collagenous stroma. Occasional thick-walled vessels were present, showing a relatively prominent smooth-muscle layer and lacking an internal elastic lamina. All vessels were lined by bland endothelial cells, and no mitotic activity or cytologic atypia was present (Fig 4). No proliferation of meningothelial cells was

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From the Departments of Radiology (J.C.B., D.J., D.K.K.), Laboratory Medicine and Pathology (K.L.E., A.R.), and Neurologic Surgery (J.V.G.), Mayo Clinic, Rochester, Minnesota.

Please address correspondence to John C. Benson, MD, Department of Radiology, Mayo Clinic, 200 1st St SW, Rochester, MN 55901; e-mail: benson.john3@mayo.edu

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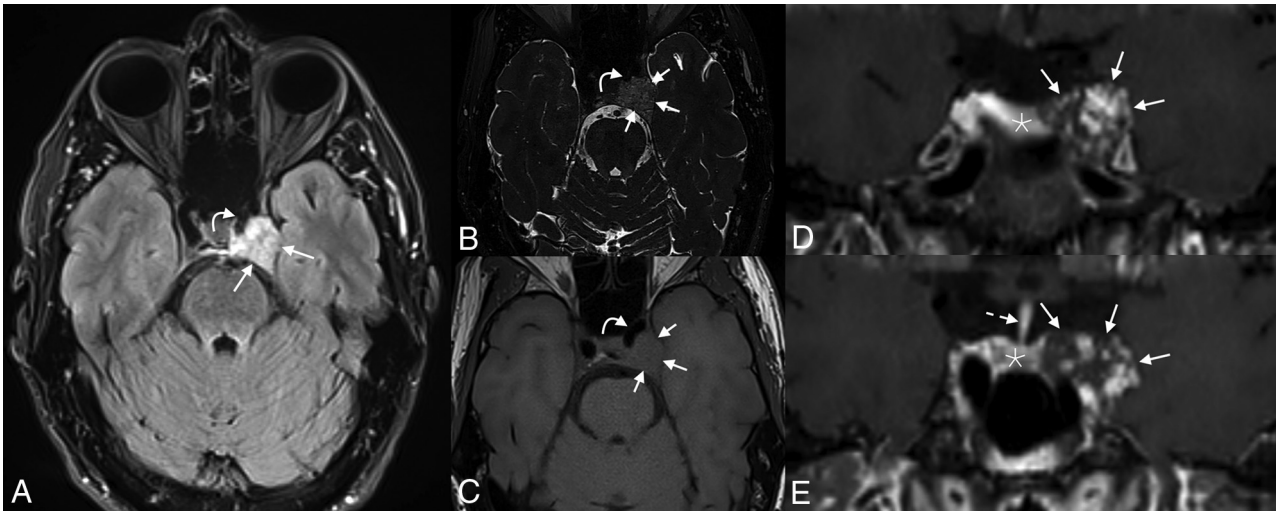

FIG 1. Initial MR imaging of the lesion demonstrates a well-circumscribed mass (*straight arrows*) in the left cavernous sinus with hyperintense signal on FLAIR (*A*). The left ICA is medialized and encased by the mass (*curved arrows*). Intralesional signal is notably heterogeneous and coarsened on T2 SPACE (*B*); enhancement is also heterogeneous (*C–E*). Coronal postcontrast images delineate encroachment of the mass into the sella, with mild mass effect on the pituitary (*asterisks*); the infundibulum (*dashed arrow*) has slightly deviated to the right.

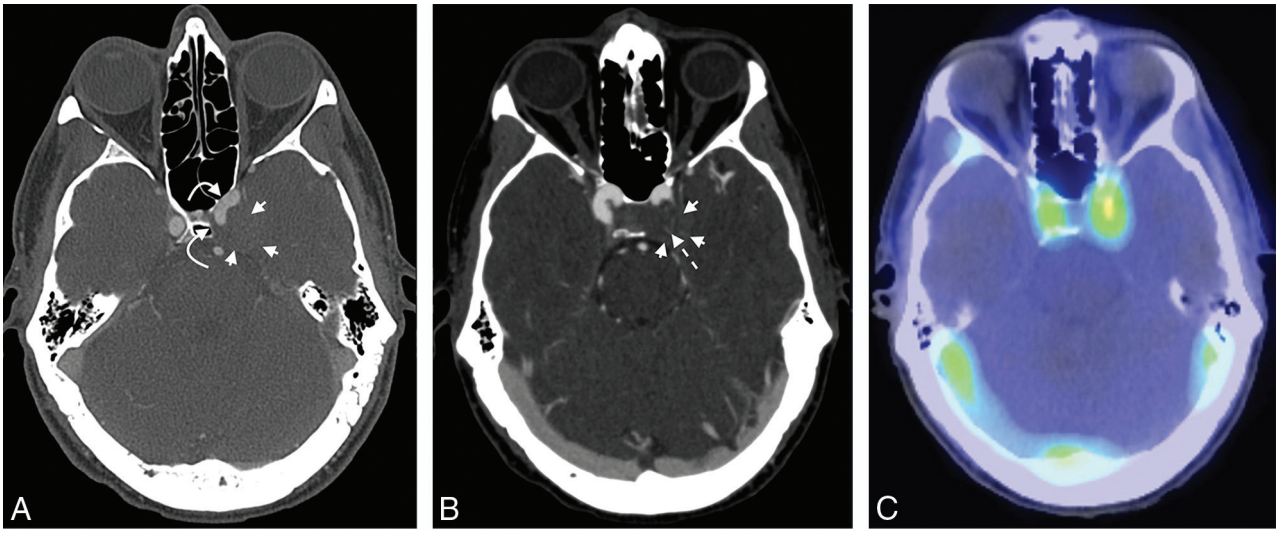

FIG 2. Preoperative CTA (*A* and *B*) demonstrates prominent bony remodeling about the margins of the mass (*curved arrows*). The mass itself is essentially isodense to the parenchyma (between the *short straight arrows*). Its inferior border is close to but does not appear to originate from the petroclival fissure (not shown). Small enhancing intralesional vessels are appreciated (*dashed arrow*). A subsequent technetium Tc-99m-tagged RBC SPECT scan (*C*) shows intense radiotracer accumulation within the mass on delayed images.

present to suggest the presence of a meningioma. There was no evidence of cartilaginous material or a proliferation of spindle cells to support the diagnosis of a chondrosarcoma or schwannoma. Overall, the histopathologic features were diagnostic of a cavernous venous malformation.[1,2] The diagnosis was cavernous sinus VM.

Discussion

Cavernous sinus VMs represent rare non-neoplastic entities and account for 3% of all cavernous sinus lesions.[3] The lesions overwhelming occur in women, accounting for up to 94% of cases. The mean age at the time of diagnosis is 44 years.[3] Because the masses tend to be slow-growing, symptom onset is often insidious, with gradually progressive headaches, cranial nerve palsies, hypopituitarism, and proptosis.[4,5] The masses may enlarge during pregnancy, resulting in symptom exacerbation that improves after delivery.[6]

Imaging features of cavernous sinus VMs have been extensively described and largely match those noted in this case. The masses are well-circumscribed and cause a local mass effect on adjacent structures. As they enlarge, VMs first displace and later encase the ICA; the vessel lumen is usually preserved (a schematic is shown in Fig 5).[7] CT often shows remodeling of the adjacent bone, while CTA may show small arterially enhancing intralesional vessels. On MR imaging, the lesions are markedly T2 hyperintense, with intralesional signal approaching that of CSF.[3]

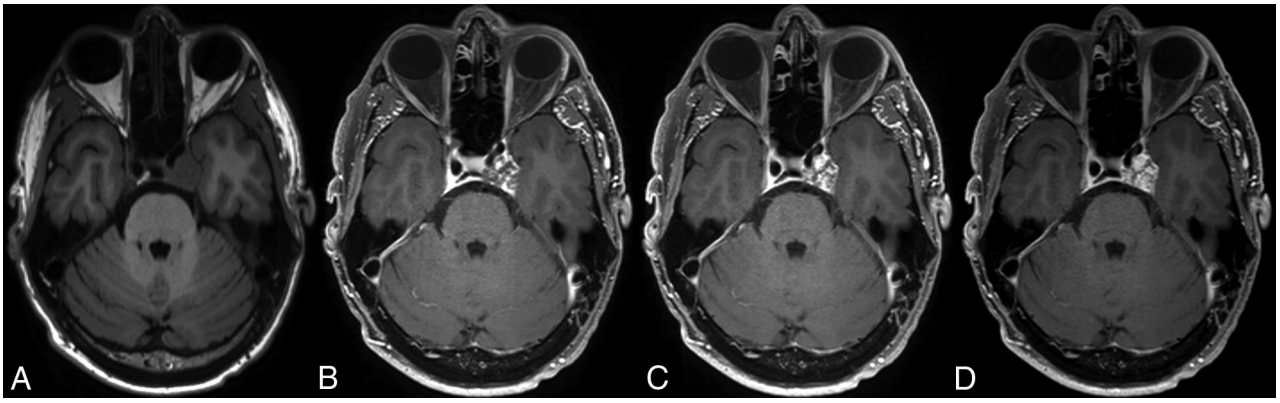

FIG 3. Dynamic contrast imaging. On precontrast (*A*) image, the mass is slightly isointense to nearby gray matter. Sequential postcontrast images (*B–D*) show progressive filling-in with contrast preferentially going to the periphery of the mass.

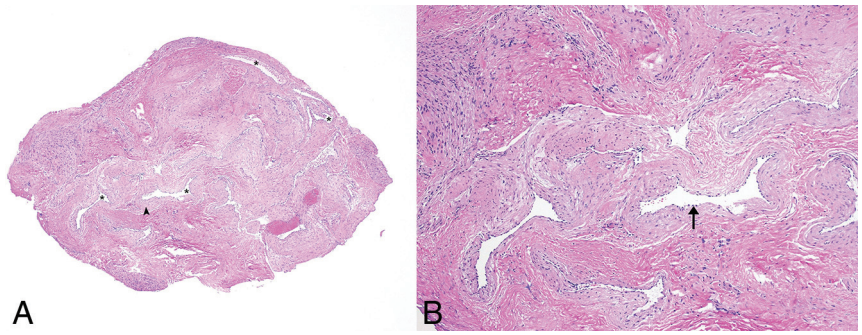

FIG 4. Histopathologic examination reveals that the mass is composed of numerous, predominantly thin-walled, anastomosing vascular channels (*asterisk*) embedded within delicate collagenous stroma. The vessels are predominantly thin-walled, with few vessels showing thickened muscular walls (*arrowhead*) (*A*, original magnification, ×40). Higher-power magnification shows that these abnormal vascular channels are lined with cytologically bland endothelial cells (*arrow*) and lack internal elastic laminae (*B*, original magnification, ×100). The specimen is stained with H&E.

Dural tails have also been reported in some lesions, though they seem to be rare.[8] He et al,[9] in an assessment of 133 cavernous sinus lesions (24 were cavernous sinus VMs), found 4 imaging biomarkers suggestive of VMs: ultra-high intralesional T2 signal, signal uniformity, infiltration of the sellar region, and dumbbell-shape. If all 4 characteristics were present, the sensitivity and specificity of the findings were 87.5% and 96.3%, respectively

Nevertheless, even with these features present, it can be difficult to confidently distinguish VMs from their closest mimickers: meningiomas and schwannomas.[10] Schwannomas, in particular, are also often dumbbell in shape.[11] When large, schwannomas typically follow the path of the nerves from which they arise. Schwannomas along the trigeminal nerve, for example, might extend posteriorly into the adjacent Meckel cave or inferiorly through the foramen ovale. Meningiomas are usually hypointense to gray matter on T2—not markedly hyperintense like hemangiomas—and often have an associated dural tail that extends along the ipsilateral tentorium.[11]

In such cases, dynamic MR imaging can help confirm the diagnosis. Like their hepatic counterparts, cavernous sinus VMs demonstrate characteristic centripetal "filling in" of enhancement on dynamic contrast imaging.[12,13] This is thought to represent a gradual influx of contrast into the large slow-flow vascular regions.[14] Still, centripetal enhancement is not universally present. Some authors have posited that 3 pathologic subtypes of VMs exist, with notable differences in their vascular makeups.[15,16] Per the classification by Yao et al,[15] type A lesions are spongelike with an intact pseudocapsule, largely composed of thin-walled sinusoids; type B lesions have incomplete or absent pseudocapsules and contain well-formed vasculatures; and type C lesions have features of both. Regardless of the recognition of such classifications, histologic differences may certainly play a role in imaging differences. For example, lesions with greater numbers of thin-walled vessels may fill with contrast earlier, thereby leading to early homogeneous enhancement that is present on all phases. Smaller lesions, too, may lack centripetal enhancement.[12]

If diagnostic uncertainty exists, a nuclear medicine–tagged RBC scan using technetium Tc99m–labeled RBCs may be used. Classically, such examinations are used to detect VMs within the liver. However, prior studies have shown similar accumulation of technetium Tc99m when used in cavernous sinus VMs.[17] Labeled RBC studies typically show photopenia during initial dynamic phase images. Subsequent slow accumulation of tracer results in marked intralesional uptake on 30-minute and 3-hour delayed images.[4]

The terminology of cavernous sinus VMs deserves explanation. Ever since Virchow first categorized vascular anomalies in the mid-19th century, the classification of vascular anomalies has undergone numerous adjustments. Today, the 2014 classification created by the International Society for the Study of Vascular Anomalies (ISSVA) is considered the mainstay resource for vascular anomaly classification.[18] Unfortunately, however, terminology from prior classification attempts has been passed on, often

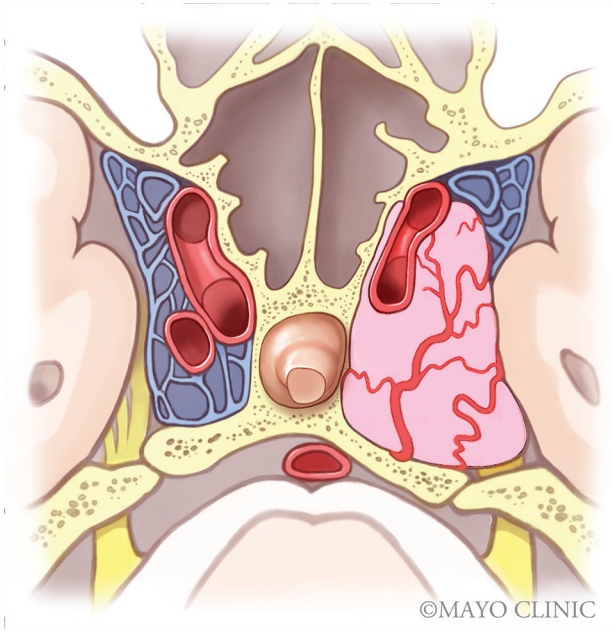

FIG 5. Illustration of a cavernous sinus vascular venous malformation with typical lesion characteristics. The mass has multiple benign characteristics, including remodeling of the adjacent bone and displacement—or encasement—of the ICA without high-grade luminal narrowing. Reproduced with permission from the Mayo Foundation for Medical Education and Research, all rights reserved.

as colloquial descriptors of various anomalies. VMs such as the one seen in this case are often still called "hemangiomas," both in the literature and clinical contexts. However, it is more precise to label such lesions vascular venous malformations.[19]

Thus, cavernous sinus VMs should not be confused with intraaxial cerebral cavernous malformations (CCMs) (often imprecisely referred to as cavernous hemangiomas or cavernomas). Per the ISSVA nomenclature, the cavernous sinus VM seen in this case is a common type of venous malformation and is morphologically distinct from other subtypes of venous malformations including CCMs. CCMs are specifically located within the parenchyma of the brain or spinal cord and have characteristic imaging findings that are distinct from common venous malformations.[18,20]

Histologically, the closest comparison with the cavernous VM in this case would be an AVM. An AVM is composed of variably sized veins, as well as arteries characterized by identifiable internal elastic lamina—a finding that was absent in our case.[1] In addition, some authors suggest that cavernous VMs may have an associated capsule or pseudocapsule, which may differentiate these lesions from CCMs.[20] However, this finding may represent reactive changes in the tissue surrounding the malformation rather than originating from the lesion itself, so it may not be a reliable indicator of these distinct pathologic entities. Also, as mentioned above, some authors have noted that not all proposed subtypes of VMs have a pseudocapsule.

Identification of cavernous sinus VMs on imaging is of utmost importance because surgical management of the lesions carries a considerable risk. The lesions are highly prone to hemorrhage and are in close anatomic proximity to the ICA and multiple cranial nerves.[3] Perioperative mortality from uncontrolled bleeding reached 12%–36% in older studies, though recent series have reported better success.[21–23] The lesions can also recur.[24] Thus, many physicians opt to treat VMs with stereotactic radiosurgery, either as a primary treatment strategy or as adjuvant therapy following surgery.[25,26] This has been shown to be successful: Lee et al,[27] in a study of 31 patients treated with stereotactic radiosurgery, showed that all patients had >50% lesion-volume reduction at 6 months. A meta-analysis by Wang et al[28] found that most patients have lesion regression after stereotactic radiosurgery.

The patient reported here recovered well from his biopsy, with only minimal double vision when looking downward. He elected to undergo gamma knife radiosurgery, which was completed 2 months after the biopsy. The patient will be followed with serial imaging to monitor the treatment response.

Case Summary

1. Cavernous sinus vascular venous malformations have multiple characteristic imaging features, including marked T2 hyperintensity and centripetal filling-in on dynamic contrast-enhanced MRI. Use of the latter, in suspected cases, can help avoid a potentially dangerous biopsy.
2. Technetium Tc99m–tagged RBC scans typically demonstrate intralesional radiotracer accumulation on delayed images.
3. Histologically, cavernous VMs are characterized as a mass-forming aggregate of venous-type vessels that lack an internal elastic lamina.
4. Cavernous sinus VMs are notoriously difficult to resect due to their propensity to hemorrhage. Radiosurgery is a viable alternative and is effective at decreasing lesion volume.

Disclosure forms provided by the authors are available with the full text and PDF of this article at www.ajnr.org.

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