Isolated Cerebral Intraaxial Varix

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Summary: A case of isolated parenchymal venous varix not seen on angiography is reported. CT demonstrated a well-defined cystic lesion with peripheral enhancement deep within the left temporal lobe. MR demonstrated a high-signal-intensity lesion with hemosiderin rim.

Index terms: Veins, abnormalities and anomalies; Veins, cerebral

Venous varices rarely present as isolated intraparenchymal lesions. The clinical, radiologic, and pathologic characteristics of this case are discussed, and the literature reviewed.

Case Report

A 17-year-old girl presented with 3 months of intermittent “dizzy spells” and three episodes of transient loss of consciousness. She also reported episodic word-finding difficulty, memory loss, and progressive difficulty with spelling, as well as increasing left frontal headaches. Physical exam was unrevealing and there were no neurologic signs. An electroencephalogram was within normal limits. Computed tomography (CT) and magnetic resonance (MR) of the brain were performed (Fig 1A–D). The differential diagnosis after CT was vascular malformation, cavernous angioma, partially thrombosed aneurysm, “cystic” neoplasm, or a brain abscess. The MR findings were consistent with a thrombosed cystic vascular malformation with a hemosiderin ring and surrounding edematous and gliotic central nervous system tissue. A cerebral angiogram was normal. At surgery, a soft, pliable blue-gray lesion containing semisolid blood, surrounded by gliotic brain tissue was found. At its margins were several partially thrombosed venous attachments. The lesion was resected in its entirety. Pathologic examination revealed a thin-walled, vascular lesion, containing semisolid blood clot. Microscopically, the wall of the lesion consisted of a blood vessel composed of connective tissue and fibroblastic cells with endothelial lining and macrophages with hemosiderin pigment (Fig 1E). A single vascular channel was present, with no nodular component. Several small venous channels within adjacent gliotic central nervous system tissue were noted, not in continuity with the cavity.

After surgery, the patient did well and was discharged without neurologic sequelae. She was asymptomatic at 14 months after surgery, but had experienced temporal lobe seizures with subtherapeutic anticonvulsant levels in the interim.

Discussion

Vascular malformations generally are classified as telangiectasias, arteriovenous malformations, cavernous angiomas, venous angiomas, or venous varices (1, 2). The differentiation of venous varices versus angiomas has been debated in the literature. They have similar wall characteristics (intima and adventitia with no media). Venous varices are a focal dilatation of a single vein and, therefore, contain no neural tissue (2). Venous angiomas contain multiple dilated anomalous veins with interposed neural tissue and one or more dilated draining veins (3). Imaging of varices has demonstrated single, enhancing saccular lesions on CT, with MR showing saccular lesions with signal flow void. Angiograms have shown focal dilation of an individual vein in each case (5, 6). They are most commonly associated with the venous drainage of high-flow vascular lesions, although isolated cases occur rarely. Angiomas appear as one or more dilated veins within the parenchyma or leptomeninges, with postcontrast enhancement on CT or signal flow void on MR (3, 4). Angiographic findings include multiple enlarged draining veins.

Typical cavernous angiomas consist of masses of sinusoidal vessels or irregular vascular spaces, which are immediately apposed and separated by fibrous tissue. As with venous varices, cavernous angiomas characteristically lack interposed neural tissue. The walls contain no elastic or muscle fibers. Cystic-appearing
Cavernous angiomas can occur (nine reported cases [7–9]), and could have an appearance similar to a large venous varix. Although the predominant radiographic findings were cystic masses, eight of these also demonstrated enhancing mural nodules or solid components on CT. The ninth lesion, although not demonstrating a nodular component on imaging studies, contained typical sinusoidal elements with large cystic components histologically. At least four of the other cases also were histologically similar.

In this case, CT and MR showed the lesion to be completely surrounded by parenchyma, initially raising the suspicion of a neoplasm such as cystic astrocytoma. Abscess and throm-
bosed aneurysm or vascular malformation also were considerations. The high signal on both T1 and T2 weighting could be consistent with either hemorrhage within a cystic tumor or thrombosis within a vascular malformation. However, the presence of the low-signal-intensity rim with magnetic susceptibility effect on long-repetition-time, long-echo-time images would be more consistent with a hemosiderin rim associated with a vascular malformation. Peritumoral hemorrhage also might have this appearance, although this is less likely. Pathologically, this case is distinguished from a typical cavernous angioma by its lack of associated neural tissue. It does not have sinusoidal characteristics or a mural nodule, therefore excluding a cystic-appearing cavernous angioma (Fig 1E).

A review of the literature reveals 26 reported cases of isolated venous varices (1, 2, 6, 10). Of these, 5 have been demonstrated radiographically. When compared with these cases, ours has several unique features. Considering all cases, 15 varices were leptomeningeal in origin, with 1 documented intraparenchymal lesion. All of the lesions radiographically imaged were leptomeningeal. The present lesion was entirely intraparenchymal. The present lesion was entirely intraparenchymal. Four prior cases were studied angiographically, and each time the lesion was identified. Ours was angiographically occult. The 2 previous cases studied with MR demonstrated signal void on T1- and T2-weighted imaging, from flowing blood within the lesions. In our case, there is high signal on both sequences. This is explained by thrombosis of the varix and also explains its angiographically occult nature.

Clinically, the two other cases to present with neurologic symptoms related to a venous varix were secondary to hemorrhage. Our patient’s symptoms were felt to be attributable to local mass effect, most likely secondary to recent expansion and thrombosis. Thus, although rare, a thrombosed venous varix should be considered during the evaluation of symptomatic intracranial cystic lesions.

References