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Medical Imaging Quiz – Case 56

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A 40-year-old man presented to the emergency department due to severe headache for three days. He referred a history of mild migraine since he was 20 years old. Physical examination revealed none abnormal finding. A cerebral computed tomography (CT) was performed and revealed the diagnosis (fig. 1).

Comment

Cerebral arteriovenous malformations (CAVMs) are a common form of cerebral vascular malformation and are composed of a nidus of vessels through which arteriovenous shunting occurs. Although arteriovenous malformations are thought to be a congenital abnormality, they develop over time, thus mean age of diagnosis is 31 years old. AVMs occur in approximately 4% of the population, with no gender predilection, but become symptomatic in only 12% of affected individuals.

Arteriovenous malformations tend to be solitary in the vast majority of cases (>95%). When multiple, syndromic associations must be considered, including: hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu syndrome) and Wyburn-Mason syndrome.

CAVMs are the most common symptomatic vascular malformations. Possible clinical presentations include incidental finding in asymptomatic patients (15%), seizures (20%), headaches, ischemic events due to vascular steal from normal brain, and hemorrhage (65%) (parenchymal, subarachnoid, intraventricular).

The origin of arteriovenous malformations remains uncertain, although they are thought to be congenital due to dysregulation of vascular endothelium growth factor (VEGF). AVMs have numerous components such as feeding arteries, nidus and draining veins. The nidus is fed by one or more arteries and drained by one or more veins. The feeding arteries are enlarged due to the increased flow, and flow-related arterial aneurysms are encountered. Venous aneurysms also referred. It may contain dystrophic calcification, a small amount of gliotic tissue, and blood at different stages of aging.

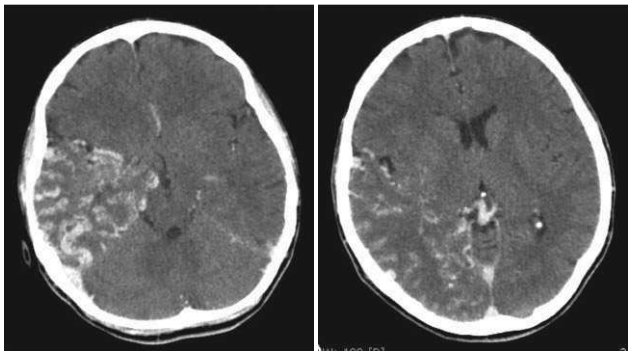

Figure 1. Cerebral computed tomography (CT) with contrast enhancement revealed feeding arteries, draining veins, and intervening nidus in the right supratentorial region, lesion compatible with cerebral arteriovenous malformation.

AVMs are located mainly supratentorial (85%) and infratentorial (15%). They can be divided into two types: compact (or glomerular) nidus with abnormal vessels without any interposed normal brain tissue or diffuse (or proliferative) nidus where no well-formed nidus is present, with functional neuronal tissue interspersed amongst the anomalous vessels. The Spetzler-Martin AVM grading system relates morphology and location to the risk of surgery.

Diagnosis can be difficult with non-contrast CT. The nidus is blood density and therefore usually somewhat hyperdense compared to adjacent brain. Enlarged draining veins may be seen. Although they might be very large in size, they do not cause any mass effect unless they bleed. Following contrast administration, and especially with CTA, the diagnosis is usually self-evident, with feeding arteries, draining veins, and intervening nidus visible in the so-called "bag of worms" appearance. The exact anatomy of feeding vessels and draining veins can be difficult to delineate, and thus, angiography remains necessary.

Angiography remains the gold standard, able to exquisitely delineate the location and number of feeding vessels and the pattern of drainage. Ideally, angiography is performed in a bi-plane system with a high rate of acquisition, as shunting can be very rapid.

On angiography, an AVM appears as a tightly packed mass of enlarged feeding arteries that supply a central nidus. One or more dilated veins drain the nidus and abnormal opacification of veins occurs in the arterial phase (early venous drainage), represents shunting.

On magnetic resonance imaging (MRI) fast flow generates flow voids, easily seen on T2 weighted images. Complications, including previous hemorrhage and adjacent edema, may be evident. MR angiography is often useful for subtracting the hematoma

components when an AVM complicated by an acute hemorrhage needs to be imaged.

Imaging differential diagnosis includes other cerebral vascular malformations, vascular tumor or glioblastoma.

Treatment options and rate of complications are dictated in part by the Spetzler-Martin grade. In general, the three options available are: microsurgical resection, endovascular occlusion or radiosurgery.

Occasionally, AVMs have been known to spontaneously resolve, usually in the setting of intracranial hemorrhage, resulting presumably in venous compression and thrombosis. The annual risk of hemorrhage for an untreated AVM is 2–3%, resulting from a flow-related aneurysm, intra-nidal aneurysm, or venous thrombosis (rarely). Following hemorrhage, the risk of a further bleed in the next 12 months is up to 18%.

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