Association of intracerebral venous angioma and true arteriovenous malformation: a rare, distinct entity

Abstract  We report a mixed cerebrovascular malformation in which a true arteriovenous malformation drained into associated venous angiomas. We describe the MRI and angiographic appearances and review the literature on mixed vascular malformations.

Key words  Angioma venous - Malformation arteriovenous

Introduction

The classification of cerebrovascular malformations (CVM) into capillary, cavernous, venous and arteriovenous is widely accepted; each has distinct radiological features. Only recently have mixed CVM been appreciated as a distinct entity [1–5]. Their pathogenesis, incidence, natural history, clinical relevance and treatment remain controversial. The association of venous angioma (VA) and arteriovenous malformation (AVM) seems to be the least common; the association of cavernous angioma and VA seems more frequent [2]. There have been a few reports of “arterialised” VA shunts with [3] or without a typical nidus [1, 2, 6].

We report the MRI and angiographic findings of a combined VA and AVM.

Case report

A previously healthy 11-year-old-boy presented with episodes of headache and new-onset temporal lobe seizures. Neurological examination was normal. MRI (Fig. 1) revealed a vascular abnormality in the medial and anterior right temporal lobe. Two adjacent radiating spokewheel patterns (caput Medusae) of contrast-enhancing draining veins, characteristic of VA, were seen. Angiography (Fig. 2) showed an AVM of the right temporal lobe with enlarged feeding arteries and a nidus draining into two adjacent venous angiomas. There was no sign of intracerebral or intraventricular hemorrhage on MRI.

The patient was treated with gamma-knife radiosurgery guided by stereotactic angiography. The radiation field covered both the AVM and the VA. The target dose was 2000 cGy. He tolerated the treatment well with no side effects or complications. He is asymptomatic 6 months after treatment and will be followed with MRI and angiography.
In 1967, Wolf et al. [7] presented the first case of an intracerebral VA with angiographic arterial involvement. A hypertrophied posterior parietal branch of the middle cerebral artery was shown to supply one of multiple small VA. Our search of the literature revealed no other report of multiple venous angiomas, although Truwit [8], mentioned, without reference, that it was not rare to find two or more immediately adjacent to each other. In our case, two adjacent VA were associated with an AVM. Descriptions of combinations of AVM and VA have been restricted to VA with arterial components that correspond to arteriovenous (AV) shunting and lack the typical AVM nidus [1–3, 5, 7, 9–11]. We found one case of a true AVM with a nidus that drained into a VA [12]. The terminology used in the literature is not uniform. This association is usually described as mixed angiomas, arterialised VA, atypical AVM or as mixed AVM-venous malformations. The lesions are considered to be transitional forms between venous malformations and AVMs, or true mixed malformations of atypical AVM and VA, or small or atypical AVM with venous predominance [2, 3, 5, 12, 13].

In a retrospective review of 34 patients with angiographically occult venous malformations (AOVM), Robinson et al. [14] suggested that cavernous malformation, AOVM and the mixed lesions have similar nonspecific presentations (seizures, headache, focal neurological deficits) and haemorrhagic tendencies. In their series, 5 lesions previously thought to be solely AV, cavernous, or other vascular malformations were actually mixed vascular malformations with heterogeneous pathological features in the same lesion. Our patient had headache and temporal lobe seizures. VAs can be associated with cavernous malformations that have a bleeding tendency [1, 4, 12]. They also coexist with AV shunts, or capillary telangiectasia [2, 4, 13, 15]. These lesions have subtle angiographic findings or are angiographically occult [1, 2, 13–16].

To treat an AVM associated with a venous angioma could be a challenge because resection will necessarily lead to an unfavourable alteration in the haemodynamic situation, with removal of the venous drainage of nearby normal parenchyma. The controversy in the description of this rare entity also extends to its treatment [1, 6, 15, 17–21]. All recent reports show a low risk of intracerebral haemorrhage from pure VA [4, 6, 21]. Cere-