CASE REPORT

Cerebral haemorrhage from a remote varix in the venous outflow of an arteriovenous malformation treated successfully by embolisation

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ABSTRACT. The “spot sign”, described in 2007, has shown that a focal area of contrast extravasation within an intracerebral haematoma (ICH) can be correlated with haematoma expansion. Here we describe a case where time-resolved dynamic CT angiography (dCTA) shows the appearance of the “spot sign” only in later images. This shows the importance of timing of static CT angiogram that, if performed too early, might result in a false negative diagnosis.

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Haemorrhage in patients with intracranial arteriovenous malformation (AVM) usually occurs from the AVM itself or from an associated arterial aneurysm. We report a case of intracerebral haemorrhage arising from a remote varix related to the venous outflow of an ipsilateral frontal AVM.

Case report

A 63-year-old male was admitted following a sudden onset of headache associated with neck stiffness and visual disturbances. There was no history of loss of consciousness or focal neurological deficit. On examination, the patient was photophobic and had nuchal rigidity. A visual field test showed a right homonymous hemianopsia but there were no other focal neurological signs.

A non-enhanced CT of the brain (Figure 1a) showed a large left temporal lobe intracerebral haemorrhage with secondary intraventricular and subarachnoid haemorrhage. There was also a calcified lesion in the left frontal lobe (Figure 1b), consistent with calcification within an AVM. A CT angiogram confirmed the presence of a left frontal AVM. An enhancing vascular structure adjacent to the left temporal lobe haematoma (Figure 1c) was demonstrated, representing either a flow aneurysm or a varix. MRI brain (Figure 1d) showed a left temporal intracerebral haematoma with an adjacent spherical area of flow void (Figure 1e), consistent with an aneurysm or varix. An AVM was noted in the left middle frontal gyrus (Figure 1d), distal from the area of haemorrhage. MR

angiography (Figure 1f) showed enlarged arterial feeders arising from the left anterior and middle cerebral arteries.

The conventional cerebral angiogram (Figure 2) confirmed the presence of a left frontal AVM (Spetzler–Martin grade 1-0-0) that was mainly supplied by branches of the left internal carotid artery. No arterial flow aneurysm was noted. The AVM had a peculiar way of draining. A primary single large collecting vein was divided into several smaller veins, majority of which were draining into the superior sagittal sinus via communication with superficial cortical veins. There was a single vein with a posteroinferior course. During late venous phase, two varices were seen on this vein (Figure 2c, f). Various degrees of stenoses were seen in the peripheral draining veins. The varix, adjacent to the area of haemorrhage, showed impaired outflow because of stenosed segments (Figure 2f) and was probably the most vulnerable part of this AVM with regard to haemorrhage.

During embolisation, the major right prefrontal middle cerebral artery feeder was initially catheterised and the tip of the microcatheter was placed intranidally within the AVM (Figure 3a). A total of 1.5 ml of Onyx 34 was injected (Figure 3b). Next, the enlarged frontal feeder from the left anterior cerebral artery was catheterised distally via the contralateral carotid and anterior communicating artery (Figure 3d), and a further 0.4 ml of Onyx was injected intranidally. This resulted in the complete obliteration of the AVM (Figure 4a, b), including the first 2 cm of the large diameter main draining vein. The smaller peripheral vein with the two varices was also no longer filled. The venous outflow of the hemisphere was not compromised. The patient had no neurological deficit post-procedure and made an uneventful recovery. Follow up angiography at three months and 1 year (Figure 4c, d) did not show any filling of the AVM or varices.

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Figure 1. (a) CT scan shows a left temporal haematoma with secondary subarachnoid haemorrhage. (b) A calcified left frontal AVM is noted in the higher axial image. (c) CT angiography shows an enhancing varix (arrowhead) at the inferior medial aspect of the left temporal lobe haematoma (white arrow). (d) MRI showing left frontal AVM (arrowhead) separate from the left temporal lobe haematoma. (e) Flow void (arrowhead) is noted within the venous varix adjacent to the haematoma. (f) MR angiography shows enlarged arterial feeders to the frontal AVM.

Discussion

A general definition of brain arteriovenous malformations (BAVM) is an abnormal tangle of vessels giving rise to arteriovenous shunting, that is non-nutritive blood flow [1]. About 53% of patients with BAVM present with haemorrhage [2]. The risk of haemorrhage is 1.3–3.9% per year for patients who present without haemorrhage [2]. For the patients who present with haemorrhage, the risk of re-haemorrhage is greater at 6.0 [3] to 6.9% [4] for the first following year; after that, the risk returns to that of the non-haemorrhagic group.

There is no widely accepted prognostic grading system other than the Spetzler–Martin scale [5], which itself has limitations as pointed out in the Joint Writing Group Report on BAVM terminology [1]. This scale does, however, take into account three very important aspects of a BAVM, i.e., its size, location and

direction of venous drainage. All of these factors predict either the risk of haemorrhage or the risk of clinically significant adverse events following surgical and radio-surgical treatment.

The main clinical factor is a presentation with haemorrhage, and a clear dichotomy must be made between patients with haemorrhagic and non-haemorrhagic presentation. As mentioned, there is increased risk of re-haemorrhage in patients with haemorrhagic presentation [6, 7]. Other adverse angio-architectural features are also associated with increased haemorrhagic risk. These include intranidal and arterial flow related aneurysms. Intranidal aneurysms have a high correlation with haemorrhagic presentation and re-haemorrhage [8]. Flow aneurysms in the distal feeding arteries near the nidus have a high chance of regression following treatment of the BAVM [8]. Coexisting arterial aneurysm is more common in patients presenting with haemorrhage than in those without haemorrhage, and these associated

Figure 2. (a–c) Anteroposterior (AP) and lateral (d–f) views of the digital subtraction angiogram show left frontal AVM with venous varices. (c) The inferior varix (arrowhead) fills slowly in the late venous phase and (f) shows stenosis of its outflow (arrowheads).

aneurysms should be treated as an independent risk factor [9]. All factors that contribute to venous hypertension are likely to increase the potential risk of intracerebral haemorrhage (ICH). The number of draining veins appears to have an inverse relation to venous pressure and thus to haemorrhagic risk [10–12]. Venous stenosis is more commonly seen in patients presenting with ICH [13, 14]. Stenosis should be diagnosed as narrowing of a draining vein in at least two angiographic views. Likewise, venous ectasia/varix is defined as at least a doubling of the diameter of the draining vein. Venous ectasia is predicted to be associated with increased haemorrhagic risk [15]. Presence of angiogenesis (moya moya type phenomenon) also appears to increase the risk of haemorrhage associated with venous stenosis [13]. In the same study, however, no increased risk of ICH was noted in association with the presence of venous ectasia [13].

In our patient, the haemorrhage had occurred not from the AVM or an associated arterial aneurysm but rather from a distal varix. Associated stenoses of the outflow

made this varix the most vulnerable area of this arteriovenous malformation. There is only one reported case of a similar remote haemorrhage in the cerebellum from a varix in an unusual venous drainage of a thalamic AVM that was treated surgically [16]. The treatment of BAVM is a multidisciplinary task requiring embolisation, surgery and radiosurgery in various combinations. Our patient was treated endovascularly with Onyx. This is a non-adhesive liquid embolic agent comprised of EVOH (ethylene vinyl alcohol) copolymer dissolved in DMSO (dimethyl sulphoxide) and suspended micronised tantalum powder that provides radio-opacity for fluoroscopic visualisation. Onyx 34, used in this patient, has 8% concentration of EVOH. The complete cure rate of BAVM from Onyx embolisation is varied and in the range of 16% [17] to 49% [18]. In a recent study [19], treatment-related permanent disability was found to occur in 3.8% of cases, and procedure-related mortality was in the range of 2.8%. In our patient, endovascular embolisation successfully occluded the AVM and there was spontaneous thrombosis of the venous side of the malformation.

Figure 3. Images obtained during embolisation show (a) selective microcatheterisation of the major prefrontal feeder from the left middle cerebral artery and (b) injection of Onyx. (c) At the end of the injection, the lateral portion of the AVM is occluded. (d) Subsequently, the frontal feeder from the left anterior cerebral artery was selectively microcatheterised from the right carotid with further injection of Onyx to occlude the medial part of the AVM.

Conclusion

In patients with AVM, haemorrhage usually occurs either directly from the AVM itself or from associated arterial aneurysms. In our patient, haemorrhage occurred from a remote varix that showed stenotic outflow distally. This should be considered as a cause of an apparently unrelated remote haemorrhage in a patient with AVM.

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Figure 4. (a, b) At the end of the embolisation procedure, the angiogram images show complete occlusion of the frontal AVM. No filling of the enlarged draining veins and associated varices is seen. (c, d) One-year follow-up images show persistent occlusion of left frontal AVM. The abnormal venous drainage is no longer visible.

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