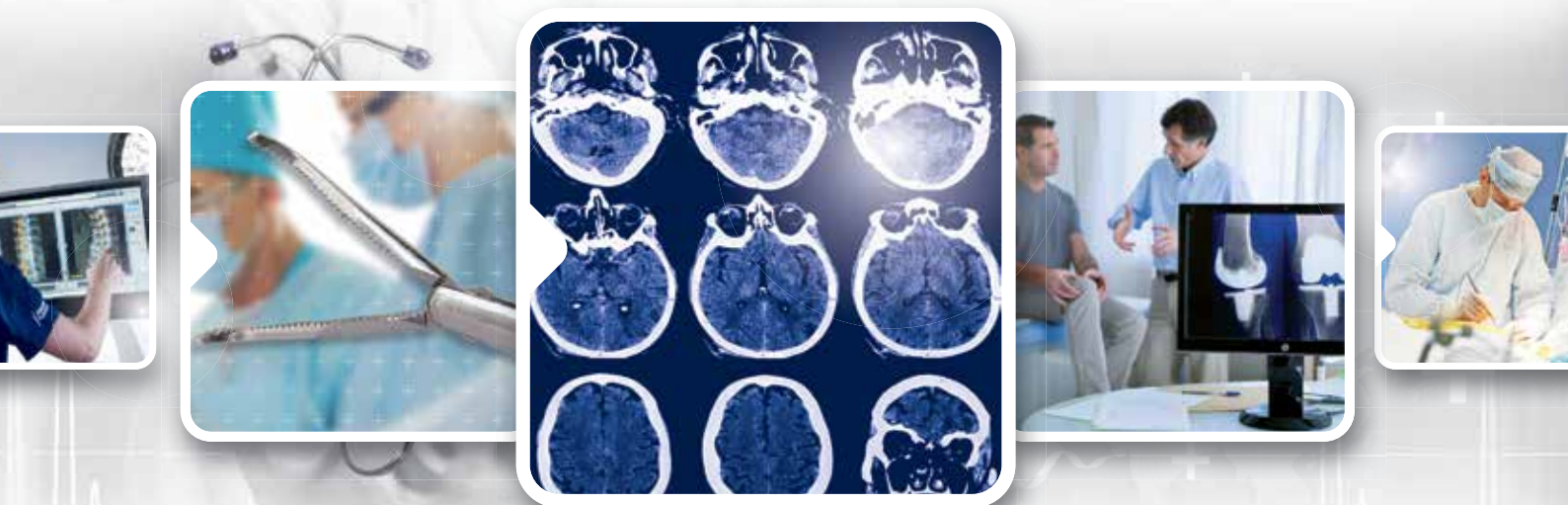


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Spontaneous thrombosis of the main draining vein revealing an unruptured brain arteriovenous malformation

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Abstract

Haemorrhage is the most frequent revealing condition of brain arteriovenous malformations (bAVMs). We report a rare case of unruptured parietal bAVM revealed by spontaneous thrombosis of the main draining vein, responsible for a focal neurological deficit. The bAVM was embolized in emergency conditions; complete regression of the neurological symptoms was observed within five days after the embolization. Potential mechanisms of such spontaneous thrombosis of the bAVM's main drainage pathway as well as an exhaustive review of the literature concerning this rare revealing condition are presented and discussed.

Keywords

Brain AVM, spontaneous, thrombosis, embolization

Background

Brain arteriovenous malformations (bAVMs) are rare vascular intracranial malformations that are supposed to be of a congenital origin.¹ Their angioarchitecture is complex and includes arterial feeders, a net of dysplastic vessels: the so-called nidus and draining vein(s).² bAVMs most frequently present with intracranial haemorrhages or seizure.^{3–7}

Flow reduction or even thrombosis of the main draining vein(s) is a well-known mechanism that may lead to haemorrhage.⁸ Thrombosis of the draining vein and subsequent haemorrhage may reveal a bAVM⁹ or may occur secondary to liquid embolic agent migration in the venous compartment in case of incomplete embolization of the nidus.^{2,10}

Thrombosis of the main draining vein without associated haemorrhage is an exceptional revealing condition of bAVM. We present such an unusual case in a parietal bAVM, and we review the literature on this topic.

Case presentation

A 31-year-old man, without any past medical history, attended our department for the sudden onset of a right hemiparesis. Four days before his admission, the patient experienced a severe frontal headache that was incompletely resolved by using oral paracetamol

medication. Two days after the headache, his family noticed slight aphasia when he spoke.

Glasgow coma scale was 15 at admission. Neurological examination revealed a motor deficit of the right inferior limb, paraesthesia of the right superior limb and mild motor aphasia (National Institute of Health Stroke Score (NIHSS) = 2).

Standard blood tests were within the normal range except for the C-reactive protein level that was slightly increased: 26 mg/L. Hypercoagulability workup (activated protein C resistance, Leiden factor V mutation, protein S deficiency, antithrombin, lupus anticoagulants) was performed and all tests were negative.

Investigations

The patient underwent an unenhanced brain computed tomography (CT) scan (16-row; Somatom Sensation

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16, Siemens, Erlangen, Germany) then a brain magnetic resonance imaging (MRI) (3 T; Signa HDX, GE, Milwaukee, USA) in emergency conditions. The CT showed an 11 mm tubular-shaped hyperdensity located close to the left parietal cortex connected to a 25 mm round hyperdense pouch (Figure 1(a)). Neither sub-arachnoid nor intraparenchymal haemorrhage was seen. The brain MRI (Figure 1(b)–(d)) showed an abnormal vascular network within the left parietal lobe, fed by parietal branches from the left anterior cerebral artery (ACA). The tubular-shaped structure, close to the enhancing vascular network, that appeared hypointense on the CT scan, presented a hypointense signal on both T2* and T2-weighted images (WI) (Figure 1(b) and (c)) and a hyperintense signal on T1-WI (Figure 1(d)). These findings were consistent

with a cortical vein thrombosis. Perivenous edema was also seen on T2-WI (Figure 1(c)). Time-resolved magnetic resonance (MR) angiography (time-resolved imaging with contrast kinetics (TRICKS)) did not show a arteriovenous shunt; i.e. no venous opacification on early arterial phase (not presented). These findings were consistent with the diagnosis of spontaneous thrombosis of the main draining vein in an unruptured bAVM.

Treatment

The patient was moved to the intensive care unit. Corticosteroids were started intravenously in order to decrease the perivenous edema (methylprednisolone 120 mg twice a day).

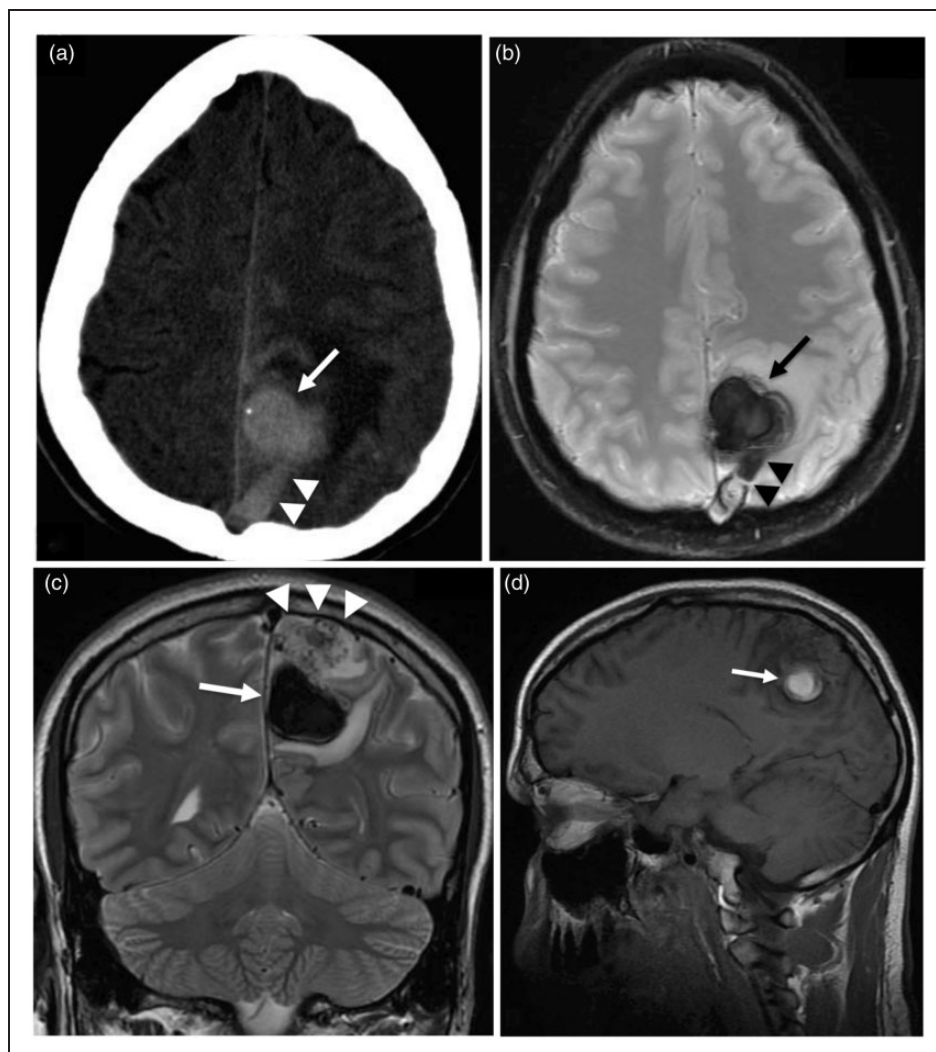


Figure 1. (a) Unenhanced 16-row computed tomography (CT) scan performed under emergency conditions; axial slice, parenchymal windowing. Spontaneous tubular-shaped hyperdensity (arrowheads) associated with a spontaneously hyperdense pouch (arrow) is seen along the left parietal cortex, suggestive of cortical vein thrombosis. Note the huge perilesional edema. Brain magnetic resonance imaging (MRI) (3 T) performed at day zero. (b) Axial T2*-weighted image (WI) showing tubular (arrowheads) and round (arrow) hypointense signals corresponding to the cortical thrombosed vein. (c) Coronal T2-WI showing the thrombosed venous pouch (arrow) and flow voids suggestive of a brain arteriovenous malformation (bAVM) nidus (arrowheads). (d) Sagittal T1-WI. The thrombosed venous pouch presents a hyperintense signal.

A digital subtraction angiography (DSA) was then performed and confirmed the diagnosis of bAVM (Figure 2). The DSA showed two main arterial feeders from the left pericallosal artery (superior and inferior parietal arteries). Tiny feeding branches from the left middle cerebral artery (MCA) were also depicted. The nidus was compact; its maximum diameter was 23 mm, making this AVM graded II in the Spetzler and Martin grading scale (nidus: 1 point, eloquence: 1 point, venous drainage: 0 point). Neither pedicle nor intranidal aneurysm was depicted. No high flow arteriovenous (AV) shunt was seen on the DSA at the arterial phase (Figure 3(a)); only a small cortical vein draining the AVM slowly into the superior sagittal sinus was seen at the late capillary phase (Figure 3(b)). These findings

confirmed the hypothesis of a thrombosis of the bAVM's main draining vein.

Due to the high estimated risk of bleeding, an endovascular treatment was considered under emergency conditions.

Under general anesthesia and full intravenous (IV) heparinization (activated clot time (ACT) between 2–3 fold the baseline), after positioning of a 6 F Envoy guiding catheter (Cordis Neurovascular, Miami Lakes, FL, USA) in the subpetrous left internal carotid artery via a 6 F femoral access, a 1.2 F Magic microcatheter (Balt Extrusion, Montmorency, France) was navigated over a 0.008 inches Mirage microguide wire (eV3/Covidien, Irvine, CA) successively in the two feeding branches from the left pericallosal artery.

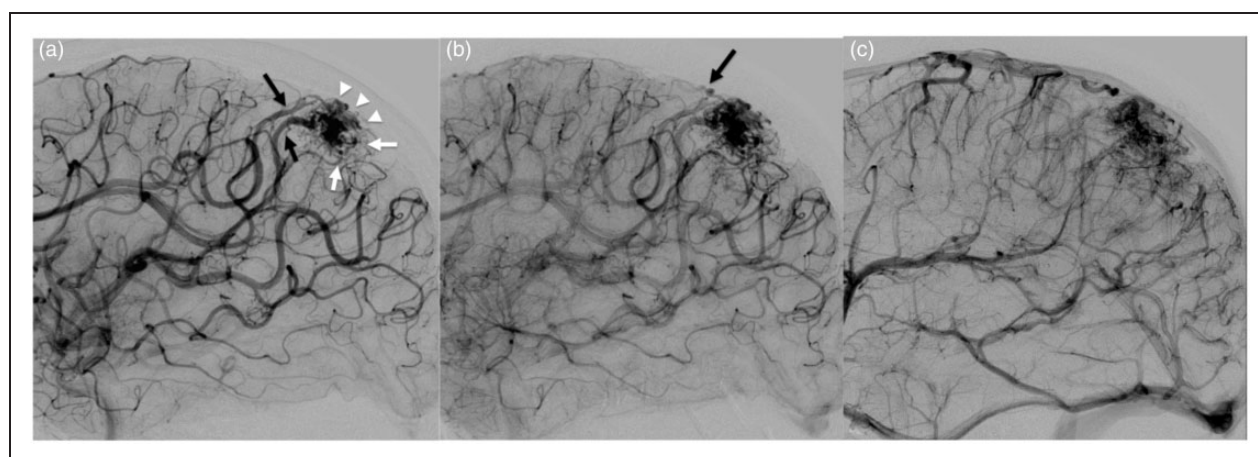


Figure 2. Left internal carotid artery (ICA) digital subtraction angiography (DSA) performed at day one: (a) lateral projections at the arterial phase, (b) capillary phase and (c) late venous phase. (a) Nidus is seen in the left parietal lobe (arrowheads). The arteriovenous malformation (AVM) is supplied by two main arterial feeders from the left pericallosal artery (black arrows) and by small branches from the left middle cerebral artery (MCA) (white arrows). (b) The tiny cortical vein is seen at a late capillary phase, slowly draining the brain arteriovenous malformation (bAVM) (black arrow). (c) No venous pouch is seen on the late venous phase; this situation is consistent with the thrombosis of the bAVM main draining pathway.

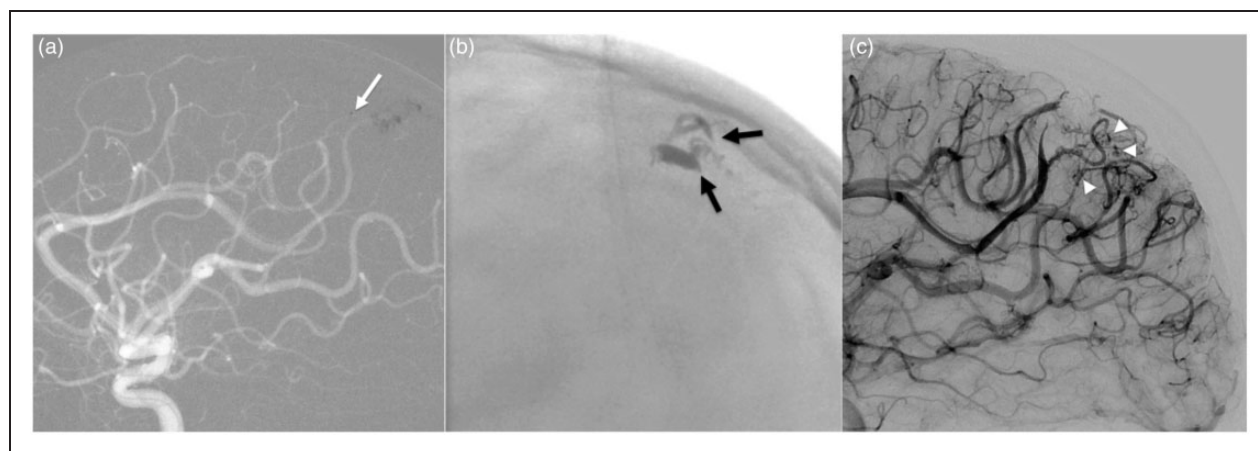


Figure 3. Endovascular treatment performed at day one. (a) Roadmap in lateral projection. Catheterization of one of the parietal branches feeding the arteriovenous malformation (AVM) by means of a 1.2 Magic microcatheter (Balt, Montmorency, France) (arrow). (b) Unsubtracted X-ray snapshot in lateral projection after glue injection (N-butyl cyano-acrylate (NBCA) diluted at 20%). (c) Control digital subtraction angiography (DSA) at the end of the procedure in lateral projection. Occlusion of the AVM is seen; only a small peripheral capillary network surrounding the embolized nidus is depicted (arrowheads).

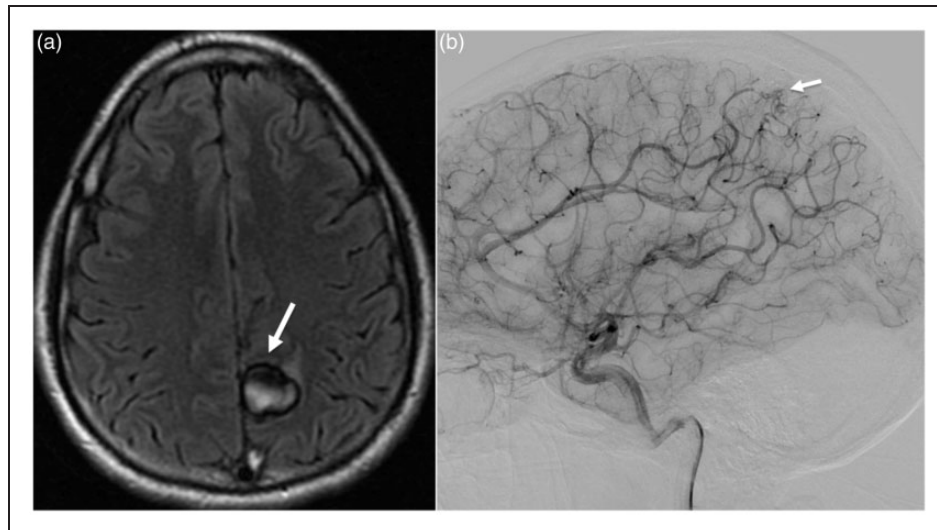


Figure 4. (a) Brain magnetic resonance imaging (MRI) (3 T) performed at three months. Axial T2-fluid attenuation inversion recovery (FLAIR)-WI showing shrinkage of the thrombosed draining vein (arrow). Note the disappearance of the perinidal edema. (b) Control digital subtraction angiography (DSA) performed at 39 months; left internal carotid artery (ICA) angiograph in lateral projection. Persistence of a very small capillary network is seen (white arrow) without residual arteriovenous (AV) shunt.

A positioning close to the nidus was obtained for each pedicle. N-butyl cyano-acrylate (NBCA) with a 20% dilution was injected under blank road map.

Postembolization control DSA showed occlusion of the nidus. Only a small peripheral capillary network surrounding the embolized nidus was seen.

Outcome and follow-up

After the embolization, the patient fully recovered from his neurological deficit and was discharged at day five uneventfully. Corticosteroid therapy was maintained for seven days and was progressively decreased. Prophylactic levetiracetam was introduced to prevent seizure.

A brain MRI performed at the three months' follow-up revealed a decrease in the size of the thrombosed vein and a complete disappearance of the perivenous edema (Figure 4). Control DSA, performed at the 39 months' follow-up, showed a very small perinidal capillary network remnant appearing as small tortuous vessels with no residual AV shunt (at the arterial or capillary phases).

Conservative management and imaging follow-up have been chosen for this tinny capillary network remnant since no residual AV shunt was visible on the control DSA.

Discussion

Review of the literature

Spontaneous angiographic regression of bAVMs occurs in less than 1.5% of cases.^{9,11} Several causes of bAVM spontaneous regression have been postulated, such as hemodynamic alteration due to haemorrhage, hypercoagulability, atherosclerosis or thromboembolism. In most cases, these bAVMs present a single draining

vein. In about 75% of these cases, the bAVM occludes after symptomatic intracerebral or subarachnoid haemorrhage.

Spontaneous venous thrombosis in unruptured bAVM is rare and only six cases have previously been published.^{9,12–14} Reported cases were three males and three females; the mean patient age was 54.5 years (range 42–67). There was no pregnant or post-partum female. The revealing clinical symptom was in most cases headache (3/6) or seizure (3/6). One patient experienced a progressive focal deficit. The remaining patient was asymptomatic.

Among those six bAVMs, 67% (4/6) were small (<3 cm) and 33% (2/6) were medium in size (3 to 6 cm); all (6/6) bAVMs had a supratentorial location. Venous drainage was superficial in 67% of the cases (4/6) and mixed in the remaining 33%; a unique draining vein was observed in 50% of the cases.^{9,12–14} None of the patients had coagulation disorder.

There are no clear data in the literature regarding the type of nidus in case of spontaneous bAVM draining vein occlusion. However, we can infer that these bAVMs probably present a fistulous or mixed plexiforme and fistulous angioarchitecture.

Physiopathologic mechanism

Pathomechanisms proposed to explain spontaneous thrombosis of the draining vein are venous stagnation related to stenosis of the draining vein, alteration of the endothelium of the main draining vein, thrombophilia/acquired coagulation disorders and mass effect related to the nidus on the main draining vein.¹⁴

In the particular case of bAVMs endovascularly treated, uncontrolled migration of the embolic agent may be responsible for delayed draining vein

thrombosis leading to subsequent haemorrhage in case of incomplete nidus occlusion.^{10,15,16}

Management

Treatment of an unruptured bAVM with thrombosed main draining vein is not consensual. Indeed, in our review of the literature we found that four patients were managed conservatively and the remaining two were treated by transarterial embolization. It is noteworthy that no patient was treated by means of stereotactic radiation therapy or surgery.

In our case, we decided to embolize the bAVM because we thought that there was an imminent risk of bleeding. Indeed, the fact that only a low flow secondary venous outlet was draining the malformation led us to think that there was a high risk of rupture. Surgery was not considered as a good therapeutic option since the huge edema surrounding the bAVM would probably increase the difficulty of the intervention. We did not use liquid embolic agent such as Onyx (eV3/Covidien, Irvine, CA) due to the eloquent location of the bAVM and due to the estimated risk of migration of the embolic agent into normal cortical branches related to reflux. We think that both corticosteroids¹⁷ and embolization triggered the reduction of the edema and avoided the bleeding of the malformation.

Conservative management may be a valuable option in bAVMs with venous drainage spontaneous thrombosis since few cases of total bAVM regression secondary to progressive spontaneous occlusion of the venous outlet have been reported in the literature.⁹ Maybe, a simple full anticoagulation therapy would have helped to reach a satisfactory clinical evolution in our case of bAVM draining vein thrombosis. However, the fact that most of the bAVM spontaneous occlusions present intracranial haemorrhage when they are diagnosed leads us to propose an aggressive treatment to prevent such haemorrhage. Additionally, full anticoagulation therapy alone was not considered because of the estimated risk of massive bleeding.

Conclusion

Even if very rare, spontaneous thrombosis of the main draining vein without subsequent haemorrhage should be known as a possible revealing condition of bAVMs and that this may incite treatment in emergency conditions. A review of the literature showed that most of the reported cases shared the same characteristics: small nidus and unique cortical venous drainage.

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Conflict of interest

The authors declare no conflict of interest.

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