Spontaneous obliteration of a dural arteriovenous fistula after treatment of polycythemia in a patient with Factor V Leiden mutation: case report

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Dear Sirs,

A 62-year-old man experienced sudden shaking and dysesthesia of the right arm that lasted about 5 min. Several hours later, he developed dysesthesia in the right hand that rapidly spread to his shoulder and right side of his face and tongue. After 10 min, these symptoms disappeared completely. Four more similar events occurred in the following 2 weeks. A few months earlier, he had experienced three episodes with visual disturbances in his right hemifield that lasted 10 min and were followed by headache. His past medical history revealed myocardial infarction, renal insufficiency due to longstanding hypertension, hyperlipidemia, and polycythemia that was untreated for several years.

Neurological examination was normal. Laboratory investigations showed an increased hemoglobin (11.6 mmol/l, reference values 8.6–10.7 mmol/l), increased hematocrit (0.56, reference values 0.41–0.50), and heterozygosity for Factor V Leiden (FVL). EEG revealed no signs of epilepsy. Subarachnoid hemorrhage was seen on CT (Fig. 1b). DSA showed a DAVF (Fig. 1c) with cortical reflux (Cognard grade 2b; Borden type II) [1, 3] and thrombosis of the superior sagittal sinus. Venesections were initiated resulting in normal hemoglobin (10.2 mmol/l) and hematocrit (0.50). No more attacks occurred afterwards.

Peroperative embolization using a burr hole directly above the fistula point was planned because tortuous feeders hampered a femoral approach and this combined approach was considered less invasive than neurosurgical occlusion. During the localizing DSA, the DAVF was found occluded with partial recanalization of the sinus (Fig. 1d–e). Follow-up MRI after 1 year showed no signs of recurrence of the fistula, a patent superior sagittal sinus, and obvious reduction of the dilated vessels (Fig. 1f).

DAVF s are one or more direct arteriovenous connections without intervening capillary bed. They are associated with trauma, cerebral venous sinus thrombosis (CVST), and thrombophilia [4, 8, 14]. Clinical outcome correlates with angiographic features. The retrograde flow into cortical veins in our patient is associated with a hemorrhage risk of 10% according to the Cognard classification [3]. Borden type II en III fistulas are associated with either progressive neurological deficits or hemorrhage [1].

Our case demonstrates that spontaneous obliteration of a DAVF can occur after treatment of polycythemia by venesections. The combination of polycythemia and heterozygosity for FVL had resulted in a prothrombotic state that probably caused gradual development of CVST and subsequent DAVF formation.

CVST commonly occurs in patients with a DAVF [14]. One hypothesis proposes opening of physiological arterial-venous shunts between meningeal arteries and venous sinuses due to increased venous pressure [7, 14]. A rat-model suggests that venous hypertension may decrease cerebral perfusion which induces angiogenesis with subsequent development of a DAVF [9, 14]. In our patient venous pressure probably decreased after normalization of the blood viscosity. Finally, organization of a thrombus in a
dural sinus may lead to “growth” of dural arteries that are normally present in sinus walls, which may result in a direct communication between artery and sinus [2].

FVL is a risk factor for CVST [12], but is also implicated in the pathogenesis of DAVFs [4, 8, 15]. Moreover, polycythemia is a risk factor for CVST [12], and these patients are prone to ischemic events probably because increased blood viscosity leads to decreased cerebral blood flow [5, 11]. Venesections may increase flow in patients with polycythemia [6, 13].

Spontaneous obliteration of DAVFs has been described [4, 10]. Possible mechanisms include recanalization of CVST, intracranial bleeding (with mass effect or vasospasm leading to obliteration), or intrinsic compression of the DAVF in the sinus wall [10]. Based on the observation in our patient, we suggest that in patients with polycythemia and DAVFs that have a relatively low risk of hemorrhage, polycythemia should be treated before interventional treatment, because venesections may lead to spontaneous closure of the DAVF.

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Conflict of interest statement The authors report no conflict of interest.

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