Superficial Siderosis as a Manifestation of a Dural Arteriovenous Fistula

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Dural arteriovenous malformations (DAVF) represent 10-15% of cerebral vascular malformations[1]. When symptomatic, their manifestations are directly related to the DAVF's location and its pattern of venous drainage. Neurological symptoms may present acutely or progressively, however intracranial hemorrhages occur spontaneously. No case of repeated *intermittent* hemorrhage due to an intracranial DAVF has been reported. Continuous or recurrent microhemorrhages into subarachnoid spaces are the proposed pathogenesis of superficial siderosis (SS), an uncommon and often unrecognized condition[2-6]. Resultant hemosiderin deposits in the leptomeninges, pial and subpial layers progressively induce neuronal damage and are responsible for SS manifestations[2]. Although other vascular pathologies have been reported in association with SS such as *cavernous malformations*, aneurysms and arteriovenous malformations[2,7,8], intracranial DAVF has not yet been described.

We describe here, the first case, to our knowledge, of intracranial DAVF that presented with progressive SS symptoms. In the literature, the only other DAVF associated with SS was located in the thoracic spine[8]. Pathogenic mechanisms of SS due to an intracranial DAVF are discussed.

Case Report

A 69-year-old woman in previous good health presented with a four year history of progressive headaches, gait disturbance, hearing loss and urinary incontinence. Neurological examination on admission showed cerebellar ataxia, bilateral neurosensorial hearing impairment and urinary retention. Cerebral and spinal 1.5 Tesla magnetic resonance imaging (MRI) scanner with routine T1, T2 and Flair, and gradient echo sequences revealed

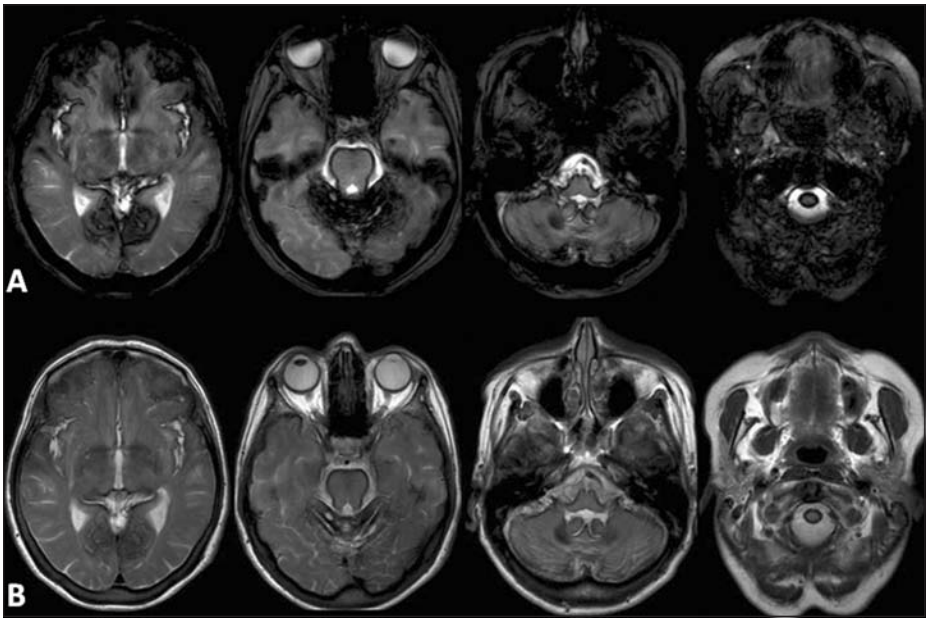

Figure 1: *MRI scanner with gradient-echo sequences (A) and axial T2 weighted images (B) showing a low signal intensity rim over the cerebellar vermis and folio, cerebral cortex, brainstem, cranial nerves and cervical spinal cord. Note the maximum hemosiderin deposit at the tentorium level. The gradient echo sequences show more extensive abnormality.*

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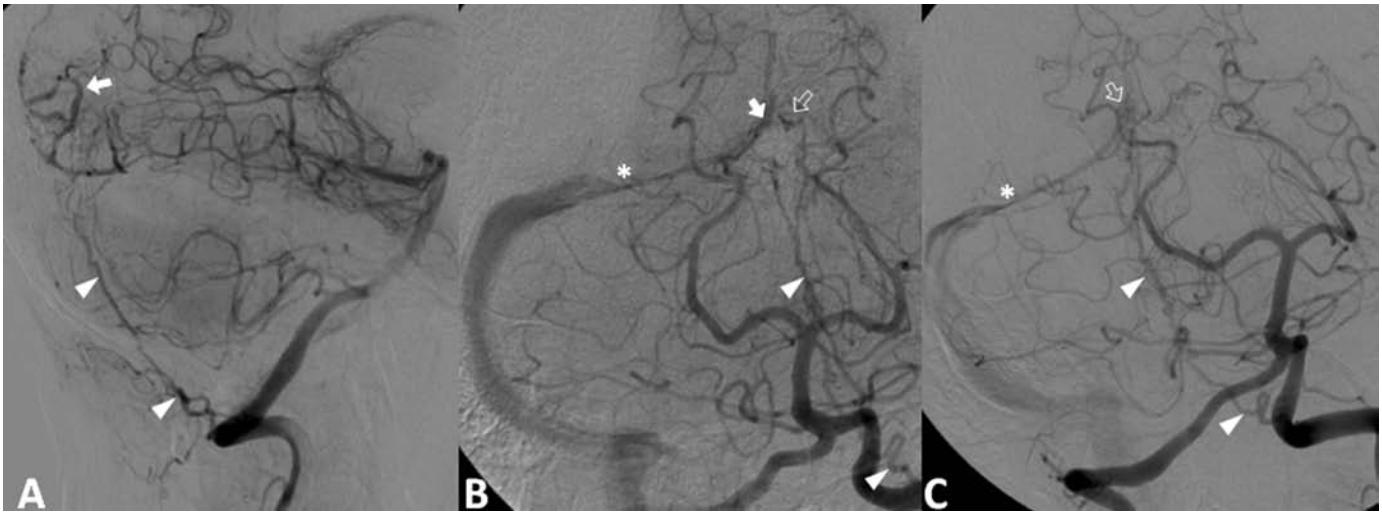

Figure 2: *Cerebral Angiography, left vertebral injection, arterial phase in lateral (A), anteroposterior (B) and oblique views (C). The high-flow DAVF is located in proximity to the torcula herofili on the right side of the tentorium (open arrowhead). The DAVF is supplied by the left posterior meningeal artery (white arrowhead) and drains towards the contralateral transverse sinus through an occipital cortical vein (white arrow). Note the proximal stenosis of the right transverse sinus (asterisk).*

characteristic findings of SS with a superficial hypo-intense rim along the pial surface of cortical gyri, cerebellum, brainstem, and cervical spinal cord (Figure 1). These exams did not show any possible source of SS. However, angiography (Figure 2) revealed a tentorial DAVF mainly supplied by a posterior meningeal branch of the left vertebral artery and accessorily by a posterior branch of the left middle meningeal artery. There was no supply from the internal carotid artery. The venous outflow was into an occipital cortical vein tributary of the the right transverse sinus, which was stenotic on its proximal segment. There was a retrograde filling of the occipital cortical vein. This qualifies as a Cognard grade 3 or a Borden grade 2 dural fistula[9,10]. Attempts to occlude the DAVF by embolization were unsuccessful. Therefore, we proceeded with a surgical intervention via a left sub-occipital approach. The yellowish discoloration of the exposed occipital lobe due to the superficial siderosis was minimal. After having exposed the tentorium, the venous drainage from the fistula was identified and coagulated using a bipolar. No attempt was made to remove the dura at the location of the fistula. Post-operatively, the patient did not experience any new neurological deficit. The follow-up angiography performed two weeks after surgery showed no residual fistula (Figure 3). At a follow-up evaluation six months after discharge the patient's gait velocity, stride length, and posture were improved.

Discussion

Dural arteriovenous fistulas constitute abnormal connections between a dural arterial feeder and a dural leptomeningeal vein. Neurological symptoms are predicted by the DAVF's location and its pattern of venous drainage. When symptomatic, DAVFs may manifest acutely or progressively with a variety of neurological symptoms related to venous hypertension. Intracranial hemorrhages due to DAVF occur spontaneously with acute clinical presentations[11,12]. Only 4% of all intracranial fistulae are located in the tentorium[1,11]. Typically, tentorial DAVFs present with intracranial hemorrhage because of their exclusive leptomeningeal venous drainage[13]. If left untreated, the risk of hemorrhage from an anterior fossa or tentorial DAVF may reach 91%[11]. Despite this high rate of clinically manifest acute hemorrhages, *repeated* microhemorrhages from a DAVF have not been reported.

This unusual presentation of a DAVF might be related to its specific tentorial location and its proposed pathogenesis. Thrombosis of veins draining into tentorial sinuses, and not one of the major sinuses themselves, is the proposed pathogenesis of acquired tentorial fistulas[12]. The local venous hypertension generated by the acute thrombosis may open latent arteriovenous communications, leading to the DAVF[12]. Normal venous drainage is redirected in a retrograde fashion through leptomeningeal veins. The lumen of these veins travelling within the leptomeninges is separated from the adjacent subarachnoid spaces only by dural collagen and a layer of endothelium[14]. This is in contrast with the walls of major sinuses which also include a layer of elastic fibers[14]. We propose that episodes of venous thrombosis within the dural leaves with subsequent local venous hypertension might have lead to seepage of blood elements through the leptomeninges. The existence of a fragile venous architecture in close proximity to the subarachnoid and subpial spaces may render DAVFs susceptible to develop SS.

Superficial siderosis is a rare disorder resulting from subclinical recurrent or continuous microhemorrhages into the subarachnoid spaces[2-6]. Deposition of hemosiderin in pial and subpial layers along the superior vermis and crests of the cerebellar folia, basal frontal lobe, temporal lobe, brainstem, spinal cord, nerve roots, and cranial nerves results in neural tissue damage[2-4]. Once the leptomeningeal ferritin biosynthesis capacity is exhausted, free radical damage and lipid peroxidation may induce neuronal injury[2-4]. When SS becomes symptomatic, the most frequent clinical findings are neuro-sensorial hearing loss (95%), cerebellar ataxia (88%), and pyramidal signs

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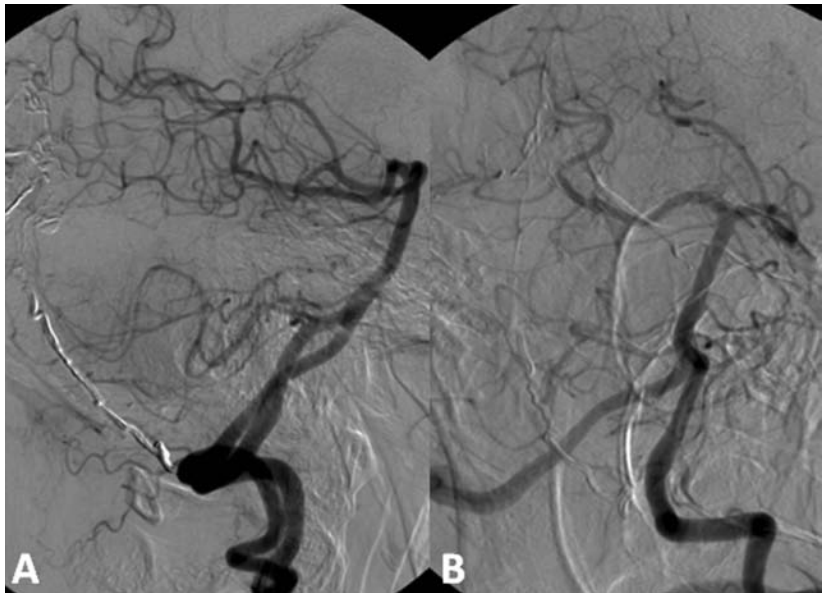

Figure 3: *Three-month post-operative cerebral angiography, left vertebral injection, arterial phase in anteroposterior (A) and oblique views (B), demonstrating the complete exclusion of the DAVF, with no early right transverse sinus opacification.*

(74%)[2,3,5,6]. Most often, as in our patient, symptoms appear progressively over many years, demonstrating the insidious nature of SS[2,15].

As opposed to idiopathic SS, in secondary SS, the source of microhemorrhages is identified[2-4,6]. Vascular pathologies have rarely been reported with SS, comprising 18% of secondary SS[2,6]. These include cavernous malformations, AVMs, cerebral aneurysms, and venous malformations[2,7]. Whatever the cause, SS is due to intermittent bleeding. In our case, we hypothesize that the particular location of this fistula may favor the occurrence of intermittent bleeding because of its drainage into weaker veins in this area. Only one other case in the literature describes an association between SS and a DAVF located at the thoracic dura but the diagnosis was made intra-operatively[8].

To the authors' knowledge, this is the first report of an intracranial DAVF presenting with progressive symptoms most likely attributable to SS. In this particular case, we ruled out other possible causes for SS. There is always the possibility that the discovery of the fistula be fortuitous, however, as described earlier, the tentorial location of this DAVF may have contributed to the intermittent bleeding. In addition, the maximum of hemosiderin deposit is at the level of the tentorium.

Elimination of the source of the bleeding should be sought whenever possible in order to stabilize and possibly improve the patient's symptoms.[3-5,7].

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