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Intracranial dural arteriovenous fistula as a cause for symptomatic superficial siderosis: A report of two cases and review of the literature

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Abstract

Background: Superficial siderosis (SS) is the occult deposition of hemosiderin within the cerebral cortex due to repeat microhemorrhages within the central nervous system. The collection of hemosiderin within the pia and superficial cortical surface can lead to injury to the nervous tissue. The most common presentation is occult sensorineural hearing loss although many patients have been misdiagnosed with diseases such as multiple sclerosis and amyotrophic lateral sclerosis before being diagnosed with SS. Only one case report exists in the literature describing an intracranial dural arteriovenous fistula (dAVF) as the putative cause for SS.

Case Description: We describe two cases of SS caused by a dAVF. Both patients had a supratentorial, cortical lesion supplied by the middle meningeal artery with venous drainage into the superior sagittal sinus. In both patients, symptoms improved after endovascular embolization. The similar anatomic relationship of both dAVFs reported presents an interesting question about the pathogenesis of SS. Similar to the pathologic changes seen in the formation of intracranial arterial aneurysms; it would be possible that changes in the blood vessel lining and wall might predispose a patient to chronic, microhemorrhage resulting in SS.

Conclusions: We describe the second and third cases of a dAVF as the cause of SS, and the first cases of successful treatment of SS-associated dAVF with endovascular embolization. As noninvasive imaging techniques become more sensitive and easily obtained, one must consider their limitations in detecting occult intracranial vascular malformations such as dAVF as a possible etiology for SS.

Key Words: Dural arteriovenous fistula, embolization, superficial siderosis

INTRODUCTION

Superficial siderosis (SS) occurs as a result of hemosiderin deposition along leptomeninges, pial, subpial, and subependymal tissues due to recurrent microhemorrhages within the central nervous system (CNS). Intracellular uptake of iron and possibly neuroinflammation causes neuronal injury leading to various symptoms of SS such as sensorineural hearing loss, ataxia, transient focal
neurological episodes similar to transient ischemic attack, and cognitive impairment.\[^{2,4,19,24,36,40,42-44}\]

SS is seen in nearly half of the patients after single episode of high grade aneurysmal subarachnoid hemorrhage.\[^{23}\] In the setting of nontraumatic and nonaneurysmal cortical SS, cerebral amyloid angiopathy has been described as the most common etiology of SS in large population studies.\[^{24,26}\] Vascular lesions such as cavernous malformations and neoplastic lesions such as ependymoma, hemorrhagic giant pro lactinoma, meningeal melanocytoma, and childhood cerebellar tumors, have also been reported to induce SS.\[^{1,9,21,33,34,39}\]

Before the advent of magnetic resonance imaging (MRI), SS was diagnosed in postmortem studies. However, with the wide availability of MRI in current practice, the SS is more frequently being diagnosed. Pathognomic finding of hemosiderin deposition can be characterized as hypointensity on T2-weighted imaging (T2WI) MRI and gradient recalled echo T2WI (GRE T2*WI) MRI.\[^{11,13,18,20}\] There is accumulating evidence that three-dimensional T2 star-weighted angiography\[^{12}\] may be superior to GRE T2*WI for the diagnosis of SS.\[^{8,46}\]

Previously, a case of intracranial dural arteriovenous fistula (dAVF) treated with open surgery and a spinal dAVF as a cause of symptomatic SS have been reported as the putative cause for SS of CNS.\[^{6,38}\] We will describe the second and third cases of intracranial dAVF as the cause of symptomatic intracranial SS and successful treatment of SS-associated dAVF with endovascular embolization.

**CASE DESCRIPTIONS**

**Case 1**

An 88-year-old male referred from outside center with the suspicion of subarachnoid hemorrhage, presented with bilateral upper extremity paresthesias and concern for stroke-like symptoms. The physical examination was unremarkable. His past medical history was significant for the previous stroke with residual right upper extremity paresthesias. A 1.5-tesla MRI revealed susceptibility artifact without associated fluid-attenuated inversion recovery nonsuppression along sulci predominately at the vertex [Figure 1a]. MR-angiography did not reveal susceptibility artifact without associated fluid-attenuated inversion recovery nonsuppression along sulci predominately at the vertex [Figure 1a]. MR-angiography did not reveal any possible source for the findings. The diagnostic cerebral angiogram revealed a left parasagittal Borden Grade I dAVF supplied by arterial feeders from the frontoparietal trunk of the left middle meningeal artery with early venous drainage into the superior sagittal sinus (SSS) [Figure 1b]. The left parasagittal dAVF lesion was treated successfully using transarterial Onyx × 34 and Onyx × 18 embolization with no residual filling and no off-target embolization [Figure 1c-d]. After the procedure, the left sided paresthesias had completely resolved, and the right-sided paresthesias were decreased in frequency. The patient was discharged without complication on postprocedure day 1.

**Case 2**

A 73-year-old male presented with a history of progressive cognitive impairment, gait ataxia, and sensorineural hearing loss. MRI of the brain showed classic findings of SS. Conventional angiography showed a Borden III splenic AVF, which was supplied by the right middle meningeal artery and drained into the SSS with cortical venous reflux into the vein of Trolard [Figure 2a-c]. Embolization was carried out from the proximal portion of the pedicle with optimal penetration into the fistula resulting in angiographic cure and return of antegrade flow in the vein of Trolard [Figure 2d]. A follow-up angiogram was performed at 12 months which demonstrated stable cure of the dAVF.

**DISCUSSION**

Iron and ferritin are found in many types of cells in the brain including neurons, microglia, and oligodendroglia in a normal state.\[^{45}\] In pathological states where blood extravasates into the brain, brain converts the iron in heme to hemosiderin, which appears 6 days after extravasation of blood into the brain parenchyma.\[^{16}\] Moreover, SS of the CNS is thought to be a result of hemosiderin deposition within the external surface of the brain due to repeat microhemorrhages. Macroscopically, SS leads to dark brown discoloration of the leptomeninges and superficial CNS parenchyma.\[^{10}\] Microscopically, SS is characterized by hemosiderin deposition in the brain.
With increasing numbers of vascular lesion
the SS has been
Since small
This patient required open surgical
Cortical SS, on the
Cerebrospinal
SS, and nonvascular intracranial pathologies, including
tumors or the brain and spinal cord (e.g., myxopapillary
ependymoma), ventricular shunts, chronic subdural
hematomas, previous CNS surgery, cavernous, and
tartovenous malformations (AVMs).[12,17,22,25,27,35,37,39,41,42]

The most common presentation of SS of CNS is
reported to be occult sensorineural hearing loss followed
ataxia/gait imbalance. Corticospinal, cognitive, and
olfactory dysfunctions are also frequent, and many
patients have been misdiagnosed with diseases such as
multiple sclerosis and amyotrophic lateral sclerosis before
being diagnosed with SS.[2,4,19,36,40,42] Cortical SS, on the
other, is associated with permanent focal neurologic
deficits such as hemiparesis or hemianopia of abrupt
onset transient focal neurologic deficits mimicking
transient ischemic attacks such as spreading pallor
or transient aphasia, and less commonly cognitive
impairment, seizures, and headache.[24] Cerebrospinal
fluid analysis may reveal xanthochromia and elevated
iron, ferritin, and red blood cell count.[5]

Dural arteriovenous fistulas (dAVF) represents 10–15% of
cerebral vascular malformations and are not commonly
associated with SS. There has only been one other
case of SS due to an intracranial dAVF described in
the literature.[18] This patient required open surgical
obliteration of the fistula, which resulted in resolution
of the patient’s symptoms. We reported two additional
cases of SS due to an intracranial dAVF which were
treated using intra-arterial Onyx embolization. Rapid
resolution of symptoms after embolization in the first
case suggests that hemodynamic effects of the dAVF may
partly responsible for the symptoms besides SS.

With the increased availability of MRI and concomitant
reductions in cost to obtain these studies, it is presumed
that prevalence of SS will increase. The majority of
these cases would presumably be asymptomatic and
incidental findings, but given the early identification
of this potentially debilitating neurologic disease. The
combined use of sensitive MR-angiographic techniques
such as (four-dimensional time of flight) combined
with diagnostic cerebral angiography in a select group of
patients will conceivably increase the numbers of patients
with SS due to occult intracranial vascular malformations
such as dAVF.

While SS due to dAVF should be included in the
differential diagnosis of an atypical presentation of
neurologic disease, the likelihood of this being the
etiology of the patient’s disease process is low. An
increasing number of reports associate atrumatic
cortical SS with cerebral amyloid angiopathy and less
commonly with reversible cerebral vasocnstriction
syndrome, primary angiitis of the CNS, and reperfusion
injury.[24,26,28–30] With increasing numbers of vascular lesion
induced SS cases reported in the literature, however,
and potentially minimally invasive and highly effective
treatment modalities such as endovascular embolization
as treatment options, the well-informed clinician may
be rewarded by considering a vascular lesions such as
dAVFs as a possible etiology for SS.[33] Since small
AVMs and dAVFs may be missed even with the best
noninvasive imaging techniques, when considering the
potentially devastating consequences of SS, conventional
angiographic evaluation can be considered for patients
with symptomatic SS. Symptomatic SS with recurrent
episodic encephalopathy due to a spinal dAVF has also
previously been reported as a cause of symptomatic SS.[6]

The two additional cases reported in the present case
as well as the previous case reports highlight the need
for a through cerebrovascular workup in symptomatic
SS patients and consideration of intracranial and spinal
dAVFs in the differential diagnosis of SS.

The similar anatomic relationship of both dAVFs
reported in the present report as well as the previous case
report presents an interesting question about the likely
pathogenesis of dAVF-induced SS. The middle meningeal
artery supplied the arterial feeders in both lesions as well
as in the previous case report, and the drainage pattern was directly into the SSS with cortical venous reflux, whereas it was into transverse sinus in the previous case report. It is plausible that the cortical venous reflux supplied by a high flow venous structure such as the SSS could create sufficient turbulent vascular flow patterns to cause pathologic changes in the walls of the delicate and fragile blood vessels of dAVF’s. In addition, similar to the pathologic changes seen in the formation of intracranial arterial aneurysms, it would be possible that changes in the blood vessels themselves might predispose a patient to chronic microhemorrhages resulting in SS.

CONCLUSION

We report the second and third cases of SS due to intracranial dAVF, which were treated with intra-arterial Onyx embolization. As noninvasive imaging techniques become more sensitive and easily obtained, it is crucial to keep in mind occult intracranial vascular malformations such as dAVF as a possible etiology for incidental SS in the appropriate patient population.

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Conflicts of interest

There are no conflicts of interest.

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