Case Report

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Isolated superior striate vein thrombosis in adults

Bianca Mazini[1], Christophe Bonvin[2], Philippe Gailloud[3] and Diego San Millán[4]

Abstract

Background: Isolated cerebral deep medullary vein thrombosis has been described in the setting of hemorrhagic periventricular white matter lesions in preterm and full-term neonates, but to the best of our knowledge, has never been reported in adults. We present two cases of isolated thrombosis of the superior striate vein occurring in adults that could be analogous to deep medullary vein thrombosis in that they involve deep cerebral veins only without thrombosis of the subependymal or internal cerebral veins.

Case description: Two women aged 20 and 39, presented with transient neurological deficits and headache. Diagnosis of isolated superior striate vein thrombosis was based on CT and MRI findings with long term imaging follow-up. Both patients evolved favorably under conservative treatment without anticoagulation. Thrombophilia workup was negative and both patients were active smokers under oral contraception.

Conclusion: Isolated superior striate vein thrombosis is a rare form of intracranial venous thrombosis and should be considered in the differential diagnosis of stroke-like episodes with headache in adults. Isolated superior striate vein thrombosis presents with characteristic imaging features on CT and MRI.

Keywords

Deep medullary veins, superior striate veins thrombosis, venous infarction, stroke like episode, headache, thunderclap, SWI

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Background

Cerebral deep medullary vein (DMV) thrombosis is occasionally observed in association with deep cerebral venous thrombosis.[1,2] Isolated DMV thrombosis is known in the pediatric literature as a form of venous thrombosis leading of parenchymal hemorrhage in newborns.[3–6] To the best of our knowledge, isolated superior striate vein, which could be considered analogous to isolated DMV thrombosis, has not been reported. We describe here two cases occurring in adult women.

Cases presentations

Informed consent for publication of clinical data and images was obtained from both patients.

Patient 1

A 20-year-old normotensive woman with no past medical history presented with thunderclap headache associated with acute but transient left-sided hemiparesis and dysarthria resolving over a 30 min period (Figure 1). The patient was an active smoker under oral contraception. Non-contrast CT demonstrated linear hyperdensities oriented along the perivascular spaces, extending from the superior aspect of the putamen across the corona radiata towards the lateral subependymal region of the lateral ventricles, suggesting thrombosis of several superior striate veins (SSVs). On initial MRI, topographically matching linear hyperintensities showed restricted diffusion and susceptibility artifact on susceptibility-weighted imaging (SWI) consistent with thrombus. There was slight locoregional white matter T2 hyperintensity without restricted diffusion consistent with vasogenic edema. DSA was unremarkable. No other intracranial venous thrombosis was detected, in particular involving the deep cerebral

[1]Service of Diagnostic and intervention Radiology, Hospital of Sion, Centre Hospitalier du Valais Romand, Switzerland
[2]Neurology Service, Hospital of Sion, Centre Hospitalier du Valais Romand, Switzerland
[3]Division of Interventional Neuroradiology, The Johns Hopkins Hospital, Baltimore, MD, USA
[4]Unit of Neuroradiology, Service of Diagnostic and Intervention Radiology, Hospital of Sion, Centre Hospitalier du Valais Romand, Switzerland

Corresponding author:
Diego San Millán, Unité de neuroradiologie, Service d'imagerie diagnostique et interventionnelle, Hôpital de Sion, Centre Hospitalier du Valais Romand, Avenue du Grand-Champsec 80 1950 Sion, Switzerland.
Email: diego.sanmillan@hopitalvs.ch

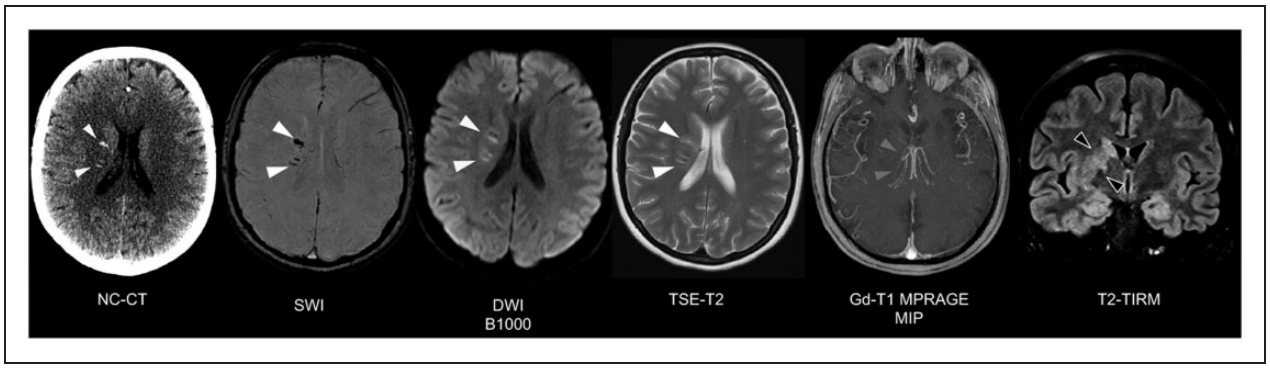

Figure 1. Non-contrast CT and MRI of Patient 1 at presentation. Thrombosed SSVs (white arrowheads) appear as spontaneous hyperdense streaks in the right corona radiata, extending from the right putamen to the superior-lateral angle of the right lateral ventricle. MRI documents blooming artifact on SWI and restricted diffusion on DWI in the thrombosed SSVs. Vasogenic edema is visible on T2-TIRM images around the thrombosed veins (embossed black arrowheads). The right thalamostriate vein (gray arrowheads) is patent (confirmed by DSA – not shown).

venous system. The patient was discharged with persistent headache but no residual neurological deficit. The headache resolved over a few weeks.

Follow-up CT (11 days) and MRI (1 and 3 months) confirmed the absence of thrombus progression. The thrombus was less conspicuous on SWI but linear susceptibility artifacts remained visible on all follow-up studies (1, 3 and 15 months), a finding consistent with the absence of recanalization. Edema was markedly reduced at one month. Follow-up MRI at one year showed residual foci of T2 hypersignal in the right corona radiata compatible with gliosis.

Patient 2

A 39-year-old normotensive woman with no past medical history presented with severe headache, transient dysarthria, left leg and face weakness, and left arm paresthesia (Figure 2). Cardiovascular risk factors included active smoking, oral contraception and dyslipidemia. Initial non-contrast CT revealed spontaneous linear hyperdensities in the right corona radiata similar to Patient 1, consistent with acute SSV thrombosis. In addition, a punctate acute hemorrhage was visible in the right putamen. MRI performed one day after onset showed slight diffusion restriction and a susceptibility artifact on echo-gradient T2 consistent with SSV thrombosis. The right putaminal hemorrhagic infarct was surrounded by a halo of cytotoxic edema itself encircled by vasogenic edema. The patient was discharged three days later with no residual neurological deficit but with a headache that persisted several weeks.

Follow-up MRI (10 days) showed a stable punctate hemorrhage, increased vasogenic edema in the right putamen and corona radiata, and new vasogenic edema in the right caudate nucleus. The two-year follow-up MRI demonstrated persistent susceptibility artifact within the thrombosed SSV and residual T2 hyperintensity in the upper portion of the corona radiata, consistent with gliosis and dilated perivascular spaces.

Given the uncomplicated clinical course, the small size of the thrombus, the presence of treatable risk factors (both patients ceased smoking and oral contraception after the acute episode), the absence of thrombus progression, and the lack of literature evidence in favor of anticoagulation in cases of isolated DMV thrombosis, no anticoagulation was initiated. Both patients remained symptom free without recurrence of intracranial thrombosis during an eight-year follow-up period.

Discussion and conclusion

We present imaging findings suggestive of isolated SSV thrombosis in two women admitted with stroke-like episodes associated with severe acute headache. The SSVs are part of the deep cerebral veins that drain the superior portion of the lentiform nucleus and the internal capsule, and contribute to the drainage of the head and body of the caudate nucleus.[7–9] They join the subependymal veins and in particular the thalamostriate vein. Acute SSV thrombosis was suspected on non-contrast CT upon visualization of linear hyperdense streaks extending from the superior aspect of the putamen towards the lateral subependymal region of the lateral ventricles, oriented along the perivascular spaces and the course of the SSVs. On MRI performed in the acute phase, these streaks showed susceptibility artifacts on GRE-T2 and SWI as well as restricted diffusion consistent with acute thrombosis.[10,11] Thrombosis affected right SSVs in both cases, with virtually identical CT and MRI findings, with the exception of a punctate putaminal hemorrhage with surrounding cytotoxic edema present in Patient 2.

CT and MRI showed the lack of involvement of other deep cerebral veins and in particular the subependymal veins or the DMVs in both patients. DSA was unremarkable in Patient 1 (not performed in Patient 2). Parenchymal abnormalities seen on initial MRIs – vasogenic edema in both cases and a punctate putaminal hemorrhage in one – were consistent with venous hypertension secondary to SSV thrombosis.

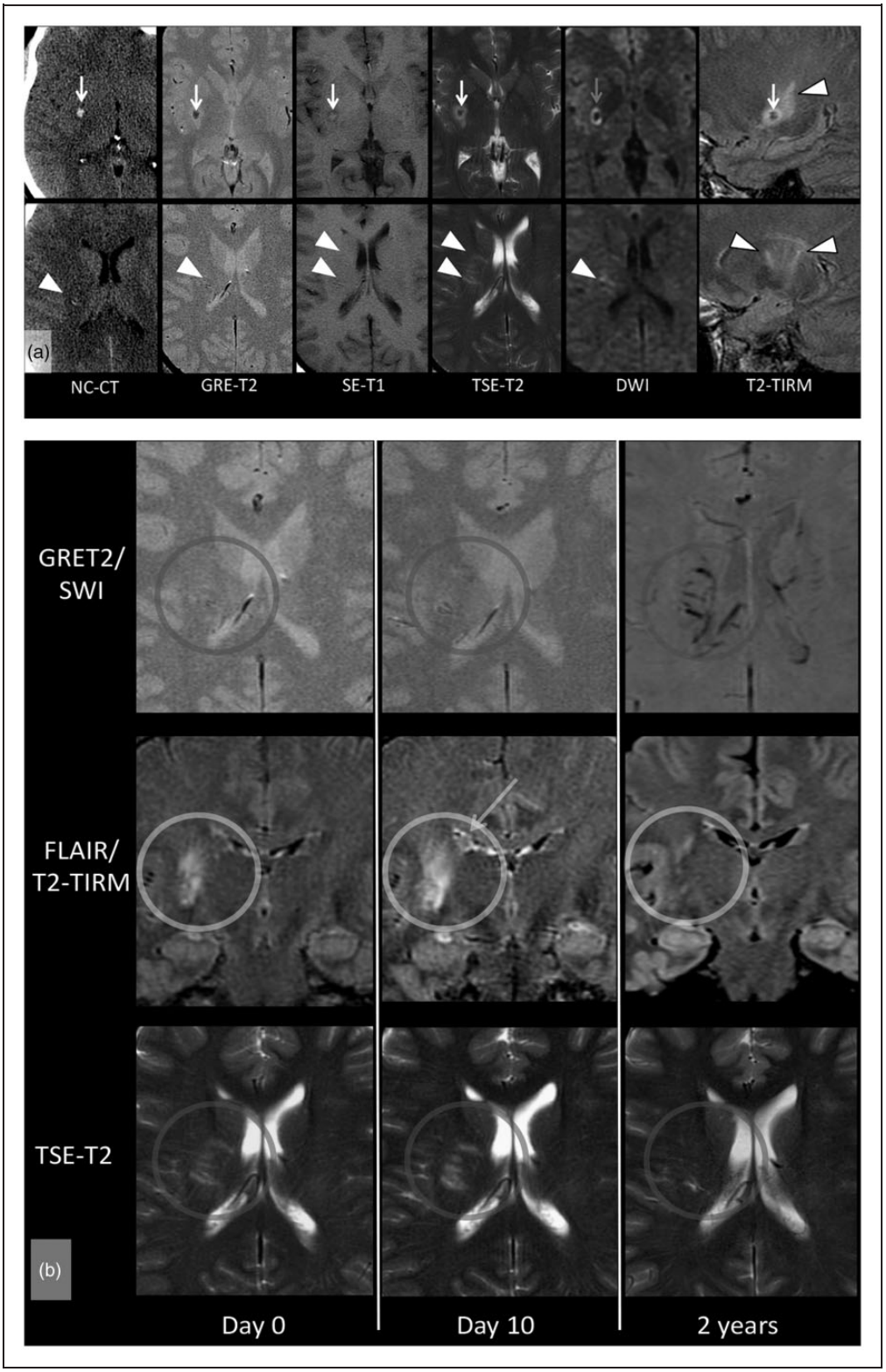

Figure 2. Non-contrast CT and MRI of Patient 2. (a) At presentation, CT shows hyperdense streaks consistent with right SSV thrombosis (white arrowhead), with slightly restricted diffusion on DWI and susceptibility artifact on echo-gradient T2 (EG-T2); this finding is less pronounced than in Patient 1 due to the lower sensitivity of EG-T2 compared to SWI. A millimetric right posterior-superior putaminal hemorrhage (white arrow) appears as a hyperdense lesion on CT and displays susceptibility artefact on EG-T2 and peripheral restricted diffusion on DWI. Sagittal T2-weighted images show vasogenic edema surrounding the thrombosed SSVs in the corona radiata. (b) Comparison of MRI findings at presentation, 10 days and 2 years. The edema has increased at day 10 but the EG-T2 appearance of the SSV thrombosis is unchanged. SWI sequences have replaced EG-T2 for the two-year follow-up study. SSV susceptibility artifacts are more conspicuous due to higher sensitivity of SWI and consistent with persistent thrombotic occlusion. The edema has evolved into smaller areas of gliosis and slightly enlarged perivascular spaces within the right corona radiata.

In Patient 1, the edema was markedly reduced on the one-month follow-up MRI. In Patient 2, MRI performed 10 days after onset due to persistent severe headache revealed a slight increase of the vasogenic edema. MRI obtained at one year (Patient 1) and two years (Patient 2) showed residual hyperintense T2 foci in the upper portion of the right internal capsule consistent with gliosis and perivascular space enlargement. The persistence of SSV susceptibility artifacts in all follow-up MRIs indicated a lack of recanalization of the thrombosed SSV that can be explained by their small size.

The absence of significant venous infarction was in keeping with the favorable clinical outcome in both cases: the neurological deficits resolved rapidly (less than 24 h in Patient 1, 3 days in Patient 2) while the headaches persisted for a few weeks. Vascular risk factors included active cigarette smoking and oral contraception. Screenings for inflammatory conditions (e.g., Behçet's disease, disseminated erythematous lupus) and thrombophilia remained negative. Given the presence of identifiable risk factors (both patients ceased smoking and oral contraception after the acute episode), a favorable immediate clinical evolution, the absence of thrombus progression, the small clot burden restricted to the SSV, the absence of previous episodes of intracranial or systemic venous thrombosis and the lack of cases of isolated SSV thrombosis in the literature with treatment recommendation, no systemic anticoagulation was initiated. The patients have remained symptom free since the presenting events, without recurrence of intracranial thrombosis (eight-year follow-up in both cases).

No previous reports of isolated SSV were found in the literature. Isolated SSV could be analogous to isolated DMV thrombosis given that they both are tributaries of the subependymal veins of the lateral ventricles, exhibit very small calibers and correspond to the drainage territory of the middle cerebral arteries. Isolated DMV thrombosis has been described in the setting of hemorrhagic periventricular white matter lesions in preterm and full-term neonates. Histopathological studies have documented the association of DMV thrombosis with hemorrhagic infarction in neonates with periventricular leukomalacia.[3] In 2011, Arrigoni et al. diagnosed DMV congestion and thrombosis by MRI in 21 neonates and demonstrated white matter anomalies within the drainage territory of the affected DMVs.[4] DMV abnormalities were isolated in nine of these cases and associated with germinal matrix hemorrhage and/or periventricular hematoma in 10 instances, suggesting different stages of a common pathological process. Fetal brain damage due to DMV involvement has also been demonstrated prenatally in three fetuses by Doneda et al.[5]: in-utero MRI showed fan-shaped white matter abnormalities consistent with DMV territory venous infarcts secondary to acute intracranial venous hypertension in relation to severe cardiac failure that led to in-utero demise in all cases. A recent clinical vignette described in-utero sonography in a 31-week fetus with a left cerebral white matter lesion consistent with intracerebral hemorrhage or neoplasm[6]; the baby was delivered seven days later and postnatal MRI showed progression of the lesion to bilateral extensive hemorrhagic white matter lesions consistent with DMV thrombosis.

Finally, in an MRI study of patients with pathologically proven neuro-Behçet disease, Albayram and co-authors demonstrated edema and hemorrhagic foci in the striatum and corona radiata as well as linear susceptibility on SWI artifacts described as "prominent venous structures" consistent with isolated SSV thrombosis, very similar to the findings in our two patients.[12]

In conclusion, isolated SSV thrombosis is a rare subset of DVST that needs to be included in the differential diagnosis of stroke-like episodes with headache (including thunderclap headache). Although histological proof could not be provided in the present cases, the diagnosis of isolated SSV thrombosis can be strongly suspected on CT and MRI based on recognition of linear modifications along the course of SSVs consistent with clot. Our experience, although limited to two cases, suggests that, unlike DVST, isolated ISS thrombosis with favorable early evolution may not necessitate systemic anticoagulation.

Authors' contribution

The corresponding author certifies that all authors have participated and been involved in the cases presented and/or in the elaboration of the present manuscript.

Availability of data and material

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Declaration of conflicting interests

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Ethics approval

All procedures performed in the studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

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ORCID iDs

Philippe Gailloud https://orcid.org/0000-0003-0768-3273
Diego San Millán https://orcid.org/0000-0003-4945-812X

Patient consent

Consent was obtained for both patients included in the study.

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