Pearls & Oy-sters: Arteriovenous Malformation With Sinus Thrombosis and Thalamic Hemorrhage

Unusual Cause of Parkinsonism and Dementia

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Abstract

Deep cerebral venous thrombosis is an uncommon condition, which usually produces headache, altered consciousness, and ocular movement abnormality. Parkinsonism occasionally occurs when there is basal ganglia involvement. We report a case of a 78-year-old man who presented with a rapidly progressive parkinsonism with poor response to dopaminergic therapy. The patient had bilateral and symmetrical hypokinesia, rigidity, and marked gait impairment with festination. Brain MRI showed bilateral thalamic hyperintensity on T2-weighted and FLAIR sequences, with right thalamic and intraventricular hemorrhage due to straight sinus thrombosis. Angiography revealed an arteriovenous malformation in the quadrigeminal cistern with afferent supply from the posterior cerebral arteries, as well as partial thrombosis of the vein of Galen and half of the straight sinus. No predisposing factor for thrombosis was found. Given the location and size of the malformation, and the substantial amount of thalamic and intraventricular hemorrhage, conservative management was decided, with slow but progressive gait improvement. The presence of deep cerebral venous thrombosis should be suspected in cases of rapidly progressive parkinsonism with cognitive decline. As in this case, thrombosis may be secondary to a deep arteriovenous malformation, a very rare occurrence that may require specific therapy.

Pearls

- Rapidly progressive parkinsonism and dementia is a rare presentation of deep cerebral venous thrombosis (DCVT) with extensive basal ganglia involvement due to venous congestion.
- DCVT typically manifests with headache, blurred vision, seizures, abnormal consciousness, and may be associated with an underlying arteriovenous malformation (AVM).
- Hemorrhage can occur secondary to the DCVT, a complication that may result in rapid worsening. This association represents a management challenge in which the benefits of anticoagulation for thrombosis should be weighed against the risk of bleeding aggravation.

Oy-sters

- Rapidly progressive parkinsonism and dementia represents a clinical challenge with a broad differential diagnosis that includes DCVT, a potentially treatable condition that should be suspected in the presence of bilateral thalamic injury.
- Other treatable disorders encompass toxic, metabolic, infectious, and ischemic (i.e., Percheron artery occlusion) disorders.
- Brain degenerative disorders such as frontotemporal dementia, Lewy body dementia, and Creutzfeldt-Jakob disease may result in a similar picture with a slower progression.

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Go to Neurology.org/N for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

Case Report

A 78-year-old man with no relevant medical history presented with progressive unsteadiness for the last 3 months, with frequent falls (repeated early falls are a red flag for Parkinson disease). He was evaluated at the emergency department by a neurologist who observed a short-stepping, unsteady gait with anteflexion of the trunk, and festination. There was mild limb hypokinesia without other core features of Parkinson disease. He had also started a cognitive decline with false recognitions, spatial-temporal disorientation, and language fluency impairment. There was no history of visual hallucinations, involuntary movements, fever, head injury, or any other intercurrent conditions. Cranial CT scan was normal, and the patient was discharged.

Over the following 2 months, his cognitive and motor condition worsened, requiring help for daily living activities (Barthel index 40), and the patient was hospitalized.

The neurologic examination revealed axial and left limb bradykinesia. There was mild bilateral postural and kinetic tremor with no rest or reemergent component and moderate axial and appendicular rigidity without cogwheel phenomenon or modification with Froment maneuver. Severe hypomimia, hypophonia, and micrography were present. He was unable to stand without assistance, presented a short-stepping gait with severely blocked gait, and marked festination. Eye movements and saccades were normal; there were no pyramidal signs, dysphagia, dysarthria, myoclonus, cortical sensory abnormalities, or any other neurologic abnormalities. His UPDRS-III score was 54.

He was disoriented in time and place and showed impairment of short-term memory, semantic memory, and abstraction capacity. His language was well constructed with no repetition or comprehension impairment, but with poor verbal phonemic and semantic fluency. He scored 14 over 30 on Folstein mental status examination. Frontal release signs were present. Brain MRI showed right thalamic and intraventricular hemorrhage and thrombosis of the vein of Galen and straight sinus (Figure 1).

Digital subtraction angiography disclosed an arteriovenous malformation in the quadrigeminal cistern with afferent supply from the posterior circulation (Figure 2), corresponding to a Spetzler-Martin grade III (1.2 cm in diameter, adjacent to eloquent areas and with deep venous drainage). Partial thrombosis of the vein of Galen and half of the straight sinus was confirmed. A workup for hereditary and acquired prothrombotic states was negative, including a whole-body CT to rule out malignancy.

A neurovascular team (neurosurgery, neuroradiology, and neurology) opted for conservative management with no anticoagulant therapy considering the high risk of bleeding. None of the currently available options for AVM, such as surgery, stereotactic radiosurgery, or embolization, were performed given the deep location of the malformation and the concomitant acute thrombosis and hemorrhage.

A dopamine transporter–SPECT imaging (DaTscan) showed a questionable bilateral putaminal hypofunction (eFigure 1, links.lww.com/WNL/B797). Levodopa therapy (up to levodopa/carbidopa 100/25 mg qid) was instituted and withheld after 8 weeks with no benefit on clinical grounds.

Brain MRI and angiography 3 months after hospital discharge showed marked radiologic improvement with no signs of thalamic or intraventricular hemorrhage, reduction of thalamic edema, partial recanalization of the straight sinus, and flow decrease in the malformed nidus (eFigure 2, links.lww.com/WNL/B797).

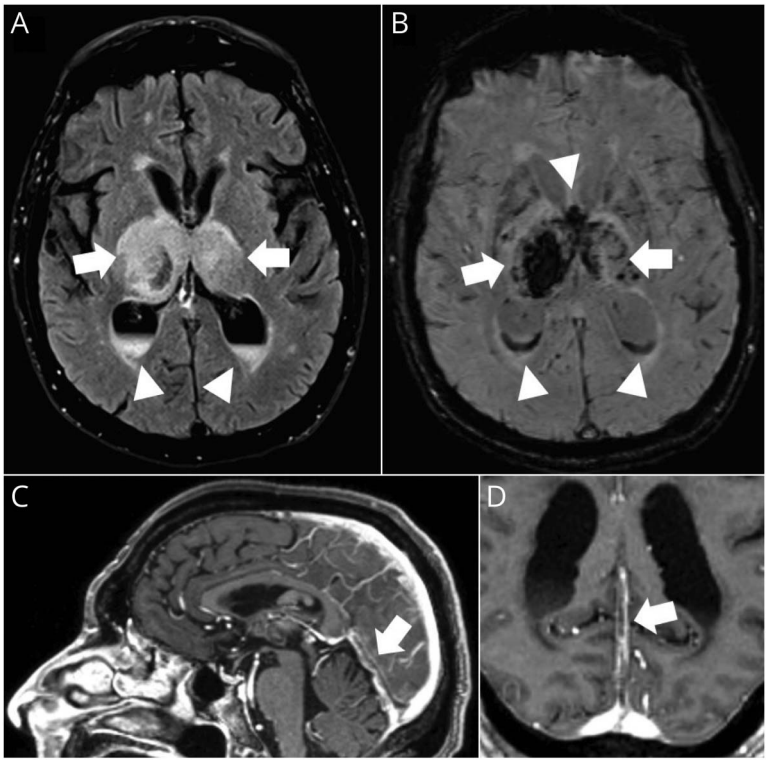
The patient showed partial spontaneous improvement in his rigid-akinetic syndrome. Three months after discharge, he had an autonomous gait and only mild rigidity in his left limbs. Cognition also improved, but the patient showed fluctuating episodes of aggressiveness that were controlled with a low dose of quetiapine without exacerbating his parkinsonian features. Being aware of the possible motor worsening, we selected an atypical neuroleptic in a low dose.

Discussion

This patient presented with recent-onset parkinsonism and cognitive decline, a rare occurrence that could be explained by the extensive basal ganglia involvement secondary to DCVST.[1-6] The rapidly evolving parkinsonian features without significant DaTscan abnormalities together with the lack of response to levodopa led to the suspicion of secondary parkinsonism. The differential diagnosis in these cases includes frontotemporal dementia, Lewy body dementia, Creutzfeldt-Jakob disease, Percheron artery occlusion, infectious encephalitis, toxic-metabolic encephalopathy, Behçet disease, and thalamic glioma or lymphoma.

In this case, given the basal ganglia involvement shown on MRI, primary degenerative causes were discarded, and gadolinium-enhanced sequence confirmed the diagnosis of DCVST. The presence of thalamic bleeding, an unusual complication of DCVST, was likely responsible for our patient's rapid worsening in the days before admission. Half of the cases of venous thrombosis with parkinsonian features significantly improved or disappeared 2–6 months after the initiation of anticoagulant therapy.[6,7] In contrast to idiopathic Parkinson disease, extrapyramidal features in patients with DCVST are bilateral and of acute onset. The disruption of frontosubcortical circuits by thalamic lesions likely represents the basis for his cognitive decline, resulting in frontal-type behavioral disturbances, with executive and attentional dysfunction, and impaired semantic and language fluency.[8-10]

Figure 1 Brain MRI Showing Axial Fluid-attenuated Inversion Recovery (A), Axial Susceptibility (B), and Sagittal (C) and Amplified Coronal (D) Gadolinium-Enhanced T1 VIBE-Weighted Images



Fluid-attenuated inversion recovery (FLAIR) hyperintensity and swelling (A, arrows) associated with confluent low susceptibility signal foci in both thalami (B, arrows) can be noted, consistent with bithalamic hemorrhage and edema. There is also an intraventricular bleeding in both lateral and third ventricles (arrowheads). Following gadolinium administration (C and D), a partial filling defect (arrows) in straight sinus and vein of Galen can be identified, consistent with thrombosis.

DCVST was not suspected initially in this patient in the absence of headache, seizures, focal neurologic deficits, or abnormal consciousness, the typical features of brain sinus thrombosis.[11]

Our patient had a deep arteriovenous malformation in the quadrigeminal plate; the association of straight sinus and vein of Galen thrombosis with a deep arteriovenous malformation complicated with thalamic hemorrhage has not been described. The vascular malformation was the only risk factor for venous thrombosis in the absence of other predisposing condition, such as infection, trauma, cancer, inflammatory bowel disease, radiation, nephrotic syndrome, prothrombotic drugs, or recent surgery. Hypercoagulable states were ruled out, including factor V Leiden, prothrombin gene mutation, protein C and S deficiency, hyperhomocysteinemia, or dysfibrinogenemia. Alternatively, it could be argued that venous thrombosis could have contributed to generate the arteriovenous malformation. It has been suggested that precipitants such as trauma or thrombotic events may facilitate AVM formation by triggering angiogenic responses.[12]

Reversible parkinsonism and rapidly progressive dementia have been described in a series of 21 patients with high-grade dural arteriovenous fistulae involving the transverse or superior sagittal sinus.[13] Successful treatment of the fistulae resulted in clinical improvement. It was suggested that venous congestion in the basal ganglia, even in the absence of MRI signal changes, explains parkinsonism in these cases of superficial sinus thrombosis.[13] In our patient, no other typical symptoms of deep AVM were present, such as seizures, tinnitus, or cranial neuropathies.

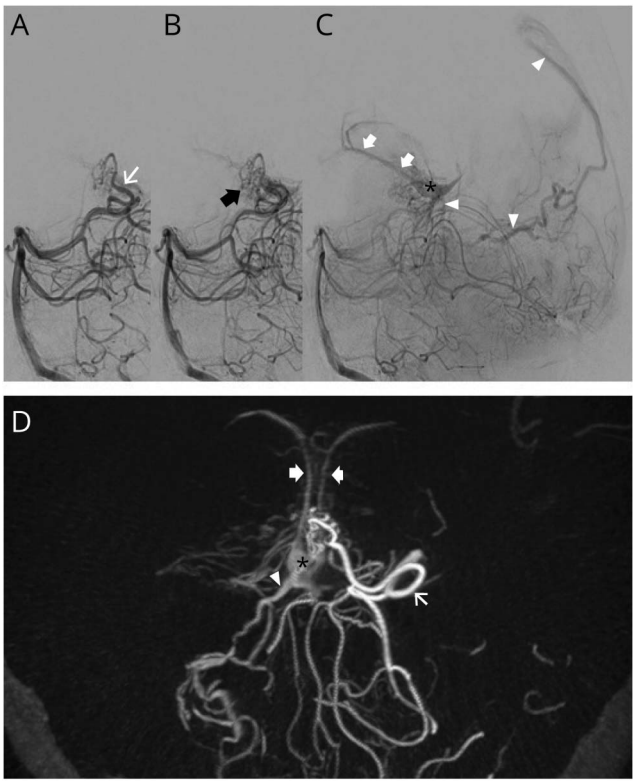
The management of DCVST complicated with thalamic hemorrhage represents a difficult challenge. The benefit of anticoagulation, indicated in brain venous thrombosis even in the presence of hemorrhage, needs to be weighed against the risk of bleeding worsening.[14] There are no clear recommendations on antiplatelet therapy.[11] The added bleeding risk of having an AVM must also be considered,[15] but there are no recommendations on drug therapy in this context. In our case, given the malformation location and the presence of recent thalamic bleeding, and the lack of supportive literature, conservative management without anticoagulation was decided.

In conclusion, DCVST secondary to a deep arteriovenous malformation is a rare, yet treatable condition to be suspected in the presence of rapidly progressive cognitive decline with parkinsonism. The presence of thalamic and intraventricular hemorrhage complicates its management due to the risk of bleeding worsening once anticoagulation therapy is started.

Study Funding

No targeted funding reported.

Figure 2 Digital Subtraction Angiography



Sequential images in lateral projection (A, B, and C) and axial MIP (maximum intensity projection) of multiplanar reconstruction from 3D rotational acquisition (D). A prominent left posterior choroidal artery (branch of the posterior cerebral artery) is identified (white thin arrow), which feeds a 9-mm nidus of an arteriovenous malformation (AVM) located in the quadrigeminal cistern (black thick arrow), with drainage toward the vein of Galen (black asterisk) and reflux from there to both internal cerebral veins (white thick arrows) and to a small cortical vein (white arrowheads) due to straight sinus thrombosis (lack of filling).

Disclosure

The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

Appendix Authors

Name	Location	Contribution
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Juan Carlos Garcia-Monco, MD, PhD, FAAN	Department of Neurology, Osakidetza Basque Health Service, Basurto University Hospital, Bilbao, Spain; University of the Basque Country	Drafting/revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data; Study concept or design; Analysis or interpretation of data

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