Thalamic Stroke and Congenital Factor V Deficiency

To the Editor:

Since Owren’s first description in 1947,1 there have been several studies of congenital factor V deficiency associated with hemorrhagic tendencies. This deficiency has also been associated six times with thrombotic complications.2-4 We report here what we believe to be the first case of cerebral thrombotic stroke associated with this deficiency.

A 20-year-old man was referred to our hospital because of an acute behavioral disorder. He previously had had an appendectomy and a traumatic lumbar hematoma. A congenital factor V deficiency was diagnosed before a tooth extraction.

The patient suddenly presented with somnolence and confusion. Neurological examination showed a right-sided facial weakness, apathetic behavior, hypophonia, memory impairment, and moderate apraxia. Cerebral computed tomography scan and magnetic resonance imaging showed a thalamic lacunar infarction in the territory of the left thalamothalamic artery. All cardiac investigations were normal, and the cerebrospinal fluid was normal. Doppler ultrasound and cerebral vascular exploration by magnetic resonance imaging showed no abnormalities of the cervical arteries. The routine hematologic and biologic studies showed no inflammatory syndrome, but screening coagulation tests revealed a prolonged activated partial thromboplastin time and marked prolongation of prothrombin time. Determination of coagulation factor activities yielded normal results, while factor V activity was 18% (normal value >60%). The plasma concentrations of antithrombin III, protein C, protein S, and plasminegen were found to be normal. There were no antiphospholipid antibodies and no lupus anticoagulant. The patient was treated with antiplatelet agents, without complications.

We investigated available members of the family and found three other relatives with the same deficiency: the patient’s father (factor V activity, 45%), his brother (11%), and his sister (51%), none of whom had ever suffered from thromboembolic or hemorrhagic events.

The congenital factor V deficiency is usually responsible for hemorrhagic disorders.1,3 The mode of inheritance seems to be autosomal recessive; only homozygous patients have hemophiliac-like hemorrhagic or thrombotic manifestations, while heterozygotes are usually asymptomatic. In 1965, Miller7 reported the first thrombotic complication associated with a factor V activity under 20%. Since then, Reich et al8 and Manotti et al9 described similar cases with recurrent systemic venous thrombosis. Only one case, a brother of the propositus, with the same defect, had suffered from arterial thrombosis (myocardial infarction).

Incidence of thromboembolism in factor V deficiency is between 5%4 and 8.7%.4 This association is paradoxical because thrombotic complications in subjects presenting with this deficiency usually predispose to hemorrhagic manifestations. However, when we analyze the reported observations,2-4 the thrombotic complications seem to appear preferentially in young subjects, are usually recurrent, and are found in other relatives of the family. Thus, we hypothesize that this disorder could be, in some cases, the etiologic cause of thrombosis. Because factor V participates in the prothrombinase complex, which is composed of factor Xa, factor Va, factor II, calcium ions, and phospholipids, an abnormal factor V could promote thrombus formation. We suggest that another unknown deficit of hemostasis could be involved in the pathogenesis of these thrombotic complications.

References


Magnetic Resonance Imaging of Acute Spontaneous Dissection of the Vertebral Artery

To the Editor:

Previous reports of magnetic resonance imaging (MRI) of vertebral artery occlusion have been made.1-5 but none of the studies were enhanced with gadolinium (Gd-DTPA). We report a patient with clinical and angiographic findings suggestive of spontaneous dissection of the left vertebral artery (VA), in which, in the acute stage, Gd-DTPA–enhanced MRI is shown to be clinically useful for the early diagnosis of VA dissection.

This previously healthy 38-year-old man presented with a 2-week history of pain in the occipital and posterior neck region, with irradiation to the left temple. Before admission he had no nausea, vomiting, or other neurological symptoms. His head pain was nearly constant, worsened with head movement, awoke him at night, and was refractory to common analgesics. One week before admission, he was seen by a physician who detected blood pressure of 170/110 mm Hg and who prescribed atenolol 50 mg/day. The morning of his admission he was unsteady, light-headed, and had blurred vision, increased intensity of the occipitotemporal pain, nausea and vomiting, and progressive weakness in the right
Three months after admission, the patient was well and only minor neurological signs could be detected. A repeat cranial MRI examination with and without Gd-DPTA was normal. A digital subtraction angiography was performed and showed resolution of the previous angiographic abnormalities.

Magnetic resonance imaging is more sensitive than CT for detection of strokes, particularly in the vertebrobasilar system. Only a few reports of MRI imaging of vertebrobasilar artery occlusions have been made. In two of them, nonenhanced MRI was performed in the acute stage, within 24 hours of the ictal event, and showed absence of a flow void in the affected vessel on axial T2-weighted images and areas of iso- or mild hyperintensity signal relative to brain parenchyma on T1-weighted or proton density images. None of them used a contrast-enhanced MRI scan.

In our patient, gadolinium demonstrated contrast enhancement of the abnormal signal over the left VA, suggesting slow flow within this artery, and thus aided the diagnosis of dissection. Although changes in signal intensity within the vessel may be attributed to thrombus formation, altered (i.e., slow) flow dynamics is the more likely possibility in this case because of the hyperintense signal on T1-weighted imaging enhanced with gadolinium. This phenomenon occurs more frequently in vessels with slow flow, usually in the venous system, probably because of the enhancement of the hydrogen nuclei within the slowly flowing vertebral artery.

This case supports the usefulness of enhanced MRI as a noninvasive method for early diagnosis of patients with suspected acute intracranial vertebral artery dissection, a situation in which plain MRI may not show conclusive abnormalities.

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References

Platelet Aggregation in Patients With Parkinson’s Disease

To the Editor:

In their article, Struck et al commented on the lower incidence of ischemic stroke in patients with Parkinson’s disease as compared with controls and attributed it to decreased tobacco intake, decrease in generalized atherosclerosis, and dopamine deficiency.
Magnetic resonance imaging of acute spontaneous dissection of the vertebral artery.
S Arroyo, A Muñoz, A Vázquez, A Hernández and E Varela de Seijas

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