Paradoxical Embolism to the Basilar Apex Associated With May-Thurner Syndrome

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Background: Embolic occlusion of intracranial vessels can be caused by material arising proximally, most commonly from the heart, the aorta, or the carotid or vertebral arteries, and rarely from systemic veins. May-Thurner syndrome is an uncommon condition in which there is impaired venous return because of compression of the left common iliac vein by the overlying right common iliac artery, resulting in iliofemoral deep venous thrombosis.

Objective: To describe a young patient with presumed paradoxical embolism to the basilar apex associated with a patent foramen ovale and May-Thurner syndrome.

Design: Single case report.

Results: A 16-year-old girl with a history of bulimia and oral contraceptive use had a “top of the basilar” syndrome. She was found to have a patent foramen ovale on transthoracic and transesophageal echocardiography. Magnetic resonance venography of the lower extremities revealed May-Thurner syndrome. Antiphospholipid antibodies (antiphosphatidylserine, anticardiolipin, and antiphosphatidyl-ethanolamine), factor V Leiden mutation by polymerase chain reaction, and homocyst(e)ine levels were normal. Anticoagulation with intravenous unfractionated heparin sodium followed by warfarin sodium was used, resulting in resolution of her neurologic deficits.

Conclusions: Deep venous thrombosis is notorious for its variable clinical manifestations and the potential dire consequences of a missed diagnosis. Physicians caring for patients with presumed paradoxical embolism should assess for May-Thurner syndrome.

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Paradoxical Embolism

EMBOLISM of cardiac origin accounts for approximately 15% of all ischemic strokes. However, among younger patients with cerebral ischemia, one fifth to one third of these episodes are presumed to be caused by emboli of cardiac origin. Many times, the site of the original embolus goes undetected, and unusual disorders need to be more seriously considered. May-Thurner syndrome is a possible cause of paradoxical emboli, which can go undetected if the patient is assessed for venous thrombosis by lower-extremity venous ultrasound alone. The syndrome is caused by a mechanical obstruction of the left common iliac vein and is associated with recurrent deep venous thromboses.

REPORT OF A CASE

A 16-year-old girl with a history of bulimia arrived at the emergency department after the acute onset of slurred speech, blurred vision, horizontal diplopia, right-sided numbness, and incoordination accompanied by an unsteady gait. Her symptoms had started approximately 2 to 3 hours before her arrival and had been preceded by an episode of vomiting on the morning of admission. She had no history of alcohol, tobacco, or other drug abuse. The patient’s medications included fluoxetine hydrochloride, oral contraceptives, and an over-the-counter appetite suppressant consisting of 12 mg of ephedrine and 40 mg of caffeine per tablet. On admission, she was hypertensive and somnolent. She had a right eyelid ptosis and decreased pupillary reflex, a left abducens palsy, bilateral facial paresis, right arm dysmetria, and bilateral Babinski signs. Initial laboratory examination showed leukocytosis (13.9 × 10³/mm³) and results from a toxicology screening were negative. Unenhanced computed tomography of the head showed no abnormalities.

The patient was admitted to the hospital and treatment was started with aspir-
leptomeningeal inflammatory disease with an embolus and irregularity of the left P1 segment was demonstrated by magnetic resonance imaging. Cerebral angiography showed a filling defect of the distal basilar artery consistent with a "top of the basilar" syndrome (Figure 2).

During the next 2 days, the patient's anarthria, tongue weakness, abducens palsy, hemiparesis, and dysmetria improved. A second brain magnetic resonance image showed progressive posterior circulation infarcts involving the midbrain, bilateral cerebral peduncles, pons, both thalami, and both cerebellar hemispheres, consistent with a "top of the basilar" syndrome (Figure 3 and Figure 4).

With improvement of her anarthria, the patient disclosed a history of intermittent left leg swelling and pain with exertion before her admission. A vascular surgeon was consulted, who recommended against surgery during the current hospitalization. Arrangements were made for future closure of the patient's PFO. At discharge, the only residual deficit was a slight left abducens palsy. Results of testing for antiphospholipid antibodies (anti- phosphatidylserine, anticardiolipin, and antiphosphati- dylethanolamine), factor V Leiden mutation, protein C, protein S, and homocyst(e)ine levels were unremarkable.

Patient foramen ovale is present in 35% of people aged 1 to 29 years and in 25% of people aged 30 to 79 years. A PFO provides opportunity for right-to-left shunting during transient increases in the right atrial pressure above the left atrial pressure, reversing the interatrial gradient. Patients with PFO are more likely to have an unknown cause of stroke. Lechat et al studied the prevalence of PFO as detected by contrast echocardiography in 60 adults younger than 55 years, with ischemic strokes and normal cardiac examination. A PFO was present in 40% of patients with stroke, compared with 10% in a control group. In patients with no identifiable cause of stroke, the prevalence was 54%. Stroke recurrence is uncommon in these patients.

May and Thurner first described "iliac compression syndrome" in 1956. The syndrome consists of impendence in venous flow of the left common iliac vein by the overlying right common iliac artery. The chronic pulsatile force of the overlying artery results in intimal hypertrophy with further diminution of venous flow. May-Thurner syndrome is estimated to be present in 2% to 5% of all patients undergoing evaluation for lower extremity venous disorders. Iliocaval compression may be completely asymptomatic, as revealed at autopsy by May and Thurner, or it may manifest itself clinically in 2 phases as described by Cockett and Thomas. The acute phase consists of sudden swelling of the left leg coinciding with iliofemoral venous thrombosis. The average age at onset is between 18 and 30 years. The chronic phase consists of venous claudication, leg swelling, venous stasis ulcers, and a tendency for recurrent venous thromboses. Varicosities and pigmentation changes of the ankle...
can be found many times as well. The morbidity associated with the chronic phase is cumulative and is seen approximately 10 to 15 years after the onset of the acute phase. The possibility of recurrent venous thromboses is of paramount importance in patients with a history of paradoxical embolus as long as the right-to-left cardiac shunt remains.

Tests commonly used for detection of May-Thurner syndrome include strain gauge venous plethysmography during rest and exercise, magnetic resonance venography, and contrast venography. In a healthy patient, venous outflow increases with exercise. In the patient with iliacaval compression, exercise will cause the venous outflow to actually decrease because of the venous obstruction. This difference in venous outflow can be found by means of strain gauge venous plethysmography during rest and exercise. The gold standard for diagnosis of May-Thurner syndrome has been contrast venography. Ascending contrast venography defines the anatomic characteristics with a sensitivity of approximately 90%. The remaining 10% can be verified by transbrachial descending contrast venography. Compared with ascending contrast venography, magnetic resonance venography has been found to have a 100% sensitivity and 96% specificity in the detection of deep venous thrombosis. Doppler ultrasonography of the lower extremities is less accurate in the assessment of the pelvic and intra-abdominal veins and is highly operator-dependent, whereas reconstructed 3-dimensional intravascular ultrasonography has provided useful and complementary information to that obtained with 2-dimensional imaging.

This rare cause of venous thrombosis does not respond well to conservative therapy. Over time, there are recurrent deep venous thromboses as well as the onset of the chronic-phase symptoms. Studies comparing anticoagulation alone with surgical intervention are not available, although surgical intervention to correct the underlying mechanical obstruction is favored currently. This consists of elevating the right common iliac artery off of the left common iliac vein with excision of intimal spurs and vein patch angioplasty. Newer endovascular techniques including stents have also been developed.

Although our patient had other possible causes for her posterior circulation infarcts (oral contraceptives, ephedrine-containing compound, and the possible non-occlusive thrombus of the right common iliac vein), the
presence of a May-Thurner syndrome brings to light an interesting possible cause of some cases of paradoxical emboli. The association with recurrent deep venous thromboses as well as the poor response to nonsurgical therapy make screening for this syndrome of great importance in the examination of the patient with presumed paradoxical central nervous system embolism.

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REFERENCES