Denouement and Comment

Moyamoya Disease

Moyamoya disease is a chronic cerebrovascular condition in which internal carotid artery or its terminal branches occlude progressively. New vessels develop, which bypass the stenotic arteries of the circle of Willis, creating a rich network of small fragile arteries. The dense angiographic appearance of newly formed collaterals is the hallmark of moyamoya, a Japanese term meaning "hazy, like a puff of smoke."

Moyamoya disease is rare, with an incidence of 0.35 to 0.54 per 100 000 people in Asia,[1-3] whereas its incidence is much lower in Western countries.[3] It has a female predominance of 1.8:1,[1] which is less evident at younger ages.[4]

Secondary moyamoya disease has been associated with neurofibromatosis type 1, Down syndrome, fibromuscular dysplasia, congenital heart disease, cranial radiotherapy, intrathecal chemotherapy, sickle cell disease,[5] and others. However, most cases are idiopathic, and a genetic origin may be present, as indicated by a higher prevalence in people of Asian descent[3] and by the occurrence of familial cases in approximately 10% of patients.

Clinical presentation is age dependent. Children usually present with an ischemic stroke and seizures rather than with hemorrhagic stroke. Adults typically present with hemorrhagic stroke more often than ischemic stroke and only rarely with seizures.[5,6] In children and adults, other symptoms can include recurrent transient ischemic events and migrainelike headaches.

The diagnosis of moyamoya disease is a clinicoradiologic one; a history of acute neurologic deficits with the characteristic angiographic image is diagnostic. Magnetic resonance imaging reveals acute and chronic strokes. Although the magnetic resonance angiogram may suggest moyamoya disease, as in our case, the criterion standard for the diagnosis remains conventional arterial angiography.

Several medical treatments, such as corticosteroids, calcium channel blockers, anticoagulants, and antiplatelet drugs, have been used with only limited benefit, reported in individual cases, whereas no controlled studies have been performed.[7,8] The most promising treatment of moyamoya disease is surgical, consisting of a bypass between the external and internal carotid systems. This can be achieved by a direct revascularization technique, such as superficial temporal artery to middle cerebral artery anastomosis, or by an indirect bypass, such as a pial synangiosis. Data are scant comparing results after different surgical approaches, so there is no universally accepted technique,[4,5,9] but indirect revascularization is generally preferred in children.

The prognosis of this disease is age related; the younger the patient, the worse the outcome. A favorable prognosis is found in 53% to 58% of patients 3 years or younger[4,5] compared with an 86% favorable prognosis in children older than 6 years.[4] As noted by Scott et al,[5] prognosis is also related to functional status at the time of surgery.

Our patient had a common presentation of moyamoya disease in children, with abrupt-onset seizures and recurrent ischemic strokes, but at an exceptionally young age. Our diagnostic assumption was confirmed by the angiogram (Figure 2). The classic "puff of smoke" can be seen at the end of the internal carotid artery (center of image), which is totally occluded without blood flow in the anterior cerebral artery and middle cerebral artery. An extensive collateral vessel network is noted. Of interest is the ophthalmic artery, the vessel on the far left of the image, which is recruited in the collateral circulation.

Our patient had poor functional status before surgery at age 13 months, with difficult-to-control seizures and the neurologic sequelae of 2 strokes. Pial synangiosis was performed without perioperative complications. However, on day 1 after surgery, the patient had a new catastrophic ischemic stroke, this time involving the entire right middle cerebral artery territory with resultant midline shift and uncal herniation. His prognosis was guarded, and in accordance with the family's directives, palliative care was initiated.

The interest of this case lies in 3 aspects. First, the young age on first presentation of our patient is rare and supplements previous case reports with onset of the disease at 4[10] and 2½[11] months of age. Second, it reinforces that young age at onset and poor functional status before surgery are risk factors for poor outcome. Third, it underlines the importance of considering vessel imaging studies in patients of Asian descent who present with acute symptomatic seizures and stroke.

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