ARCHIVES OF HELLENIC MEDICINE: ISSN 11-05-3992

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Medical imaging Quiz – Case 61

ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2020, 37(3):422–424
ARCHIVES OF HELLENIC MEDICINE 2020, 37(3):422–424

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A 15-month-old female was admitted to the hospital, due to sudden onset of left hemiparesis. Parents reported an episode of transient paresis of the left leg one month ago. Past medical history was free of any health problem. Physical examination revealed neurological signs compatible with left hemiparesis. Examination of other systems was unremarkable.

Blood tests at admission were normal. Echocardiography did not detect any abnormalities. Computed tomography (CT) revealed hypodense areas in the frontoparietal and occipital regions in the right cerebral hemisphere, suggesting acute and chronic infarcts respectively.

Magnetic resonance imaging (MRI) using T1 and T2 weighted, spin echo, FLAIR, DWI and ADC sequences, revealed well-defined wedge-shaped hyperintensities in bilateral frontal, parietal, parieto-occipital, basal ganglia and subcortical white matter, suggestive of acute and chronic ischemic lesions. Basal ganglia collaterals were noted on T1W scans (fig. 1).

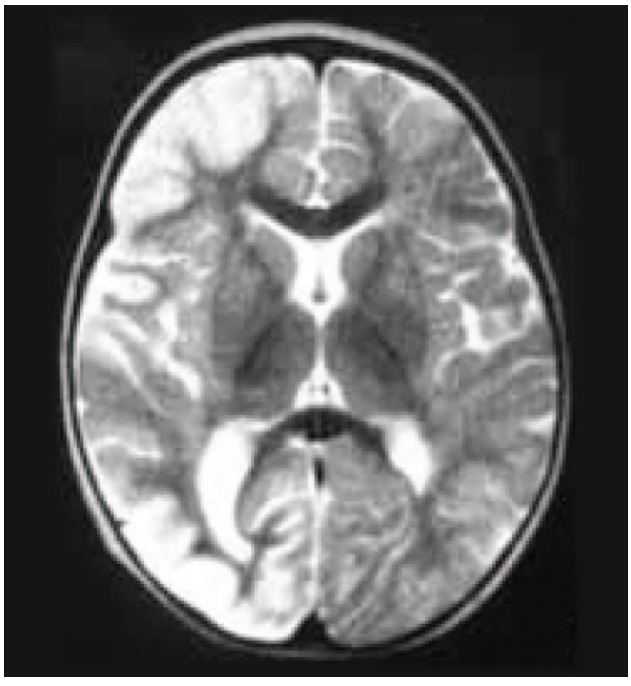

Figure 1. Axial T2 weighted image shows multiple hyperintense lesions in the right frontal, parietal, and paritooccipital lobe. A small high signal lesion is also noted at the left parietal lobe.

MRA images through the circle of Willis revealed near-complete occlusion of both supraclinoid internal carotid arteries (ICAs), middle, anterior and right posterior artery. The collateral "moyamoya vessels" in the basal ganglia resembled a "puff of smoke" (fig. 2). After the MRA findings a conventional cerebral angiography (CCA) was performed. CCA revealed near-complete

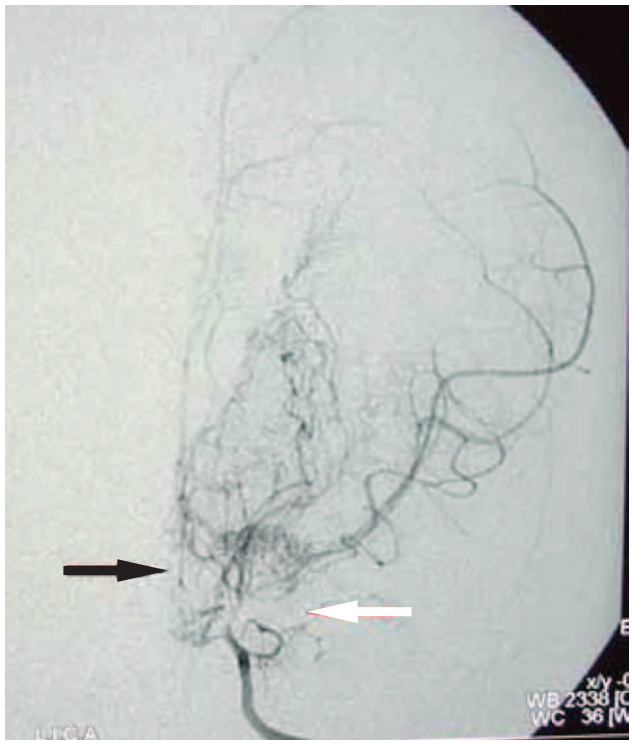

Figure 2. Left internal carotid angiogram confirms (a) subclinoid occlusion of the left internal carotid (black arrow) and reveals vascular collateral network called puff of smoke (white arrow), and (b) occlusion of the left middle cerebral artery (white arrow) and stenosis of the anterior cerebral artery (black arrow).

occlusion of both supraclinoid ICAs, anterior cerebral arteries and of right posterior artery. Both anterior cerebral arteries seemed to fill from the collaterals. Vascular channels in the skull-base region (leptomeningeal collaterals) and basal ganglia resembled a "puff of smoke". External carotid arteries (ECA), common carotid arteries and the basilar system were normal (fig. 2).

All imaging findings were characteristic of the diagnosis. Laboratory workup excluded cardiovascular, infectious, autoimmune, hematologic, prothrombotic and metabolic disorders.

Comments

Moyamoya disease was first described in Japan in 1957. Many similar cases have been reported, mainly in Japan and other Asian countries and less frequently in North America and Europe. The annual incidence of moyamoya is 0.35 to 0.94 per 100,000 population. There is a female predominance, with a male-to-female ratio of 1:2.2. Age distribution of MD varies with a higher peak reported in the 45–49 year old group and a lower peak in the 5–9 year old group.

The etiology of moyamoya disease is unknown; nevertheless the high incidence among the Japanese and Asian population, together with a familial occurrence of approximately 10 percent of cases, suggests a genetic etiology. Familial moyamoya disease has been linked to chromosomes 3p24.2–p26, 6q25, 8q23, 12p12, and 17q25.

Classic angiographic findings of moyamoya vessels are described as "moyamoya phenomenon". Patients with the angiographic appearance of moyamoya and no known risk factors are considered to have moyamoya disease whereas those with underlying disease "moyamoya syndrome".

Some of the conditions associated with moyamoya syndrome include: Congenital heart disease, atherosclerosis, infectious diseases, hematologic conditions such as sickle cell diseases and beta thalassemia, factor XII deficiency, metabolic diseases, homocystinuria, vasculitis and autoimmune diseases, systemic lupus erythematosus, Grave's disease, antiphospholipid antibody syndrome, connective tissue disorders and neurocutaneous syndromes, neurofibromatosis type 1 (NF1), chromosomal disorders, vasospasm after subarachnoid hemorrhage, cranial trauma, and brain tumors.

The clinical manifestations of moyamoya are variable and include transient ischemic attack, ischemic stroke, hemorrhagic stroke, and epilepsy. Ischemic cerebrovascular events, either transient ischemic attack (TIA) or infarction, are more prevalent than hemorrhagic events in children.

Head CT and brain MRI are important studies for the detection of brain infarction and hemorrhage in patients with moyamoya. Noninvasive and conventional angiographic studies can demonstrate stenosis or occlusion of the circle of Willis vessels. Transcranial Doppler ultrasonography is a noninvasive way to evaluate intracranial hemodynamics and large artery stenosis. A number of methods may be useful to estimate resting brain perfusion and blood flow reserve. However, the gold standard for the diagnosis of moyamoya disease remains the conventional cerebral angiography.

The diagnosis of moyamoya disease is based upon the characteristic angiographic appearance of bilateral stenosis affecting the distal internal carotid arteries and proximal circle of Willis vessels, along with the presence of prominent basal collateral vessels. Moyamoya disease is one of the differential diagnoses of stroke in children or young adults.

Diagnostic criteria for idiopathic moyamoya disease proposed by a Japanese research committee include the following major requirements: (a) Stenosis or occlusion at the terminal portion of the internal carotid artery and at the proximal portion of the anterior and middle cerebral arteries on MRA, (b) abnormal vascular networks in the basal ganglia on MRA, and (c) exclusion of conditions such as arteriosclerosis, autoimmune disease, brain neoplasm, Down syndrome, head trauma, and neurofibromatosis.

There is no curative treatment for moyamoya disease. In patients with moyamoya syndrome, it is also important to search for and treat the underlying condition. Secondary prevention for patients with symptomatic moyamoya is largely centered on surgical revascularization techniques. The goal of surgical treatment for moyamoya disease is to reduce the risk of ischemic stroke by improving the cerebral circulation. Surgical techniques can be divided into direct and indirect revascularization procedures and their combinations. Superficial temporal artery to middle cerebral artery (MCA) bypass or middle meningeal artery to MCA bypass are the most common direct techniques. Direct methods are technically difficult to perform in children because of the small size of donor and recipient vessels. Indirect techniques include encephaloduroarteriosynangiosis, encephalomyosynangiosis, encephaloarteriosynangiosis and omentum transplantation

Antiplatelet agents, usually aspirin, have been used to treat some patients with moyamoya disease or moyamoya syndrome, particularly those who are asymptomatic or have mildly symptomatic ischemic disease, those considered to have a high risk for poor surgical outcome or even patients after surgical revascularization. However, evidence of benefit with antiplatelets in these settings is limited and equivocal.

Natural history of moyamoya disease tends to be progressive.

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