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Case Report Open Access

Moyamoya Disease: A Diagnostic Challenge

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Introduction

Takaku Suzuki and others described disease in Japan in the sixties. The term means a 'wavering puff of smoke' and was used to describe the abnormal vasculature at the base of the brain. Moyamoya disease is a cerebro vascular disease that features narrowing or stenosis, starting at the distal internal carotid and proximal portions of the anterior and middle cerebral arteries.

Moyamoya syndrome is a phenomenon caused by an olegemic state similar in presentation but caused by various disease entities [1]. In this condition, the carotid arteries that supply blood to the brain become thickened and narrowed gradually over time. As a result, the brain does not receive enough oxygen-rich blood.

To compensate for the reduced supply, the body grows new arteries around the head and brain. These new arteries, also called the 'Moyamoya vessels' try to compensate for the reduced blood flow. However, these new vessels never fully compensate for the reduced blood supply. They also have a propensity to bleed easily [2].

There are no community based studies from India. The prevalence of the disease ranges from 3.2 to 10.5 per 100,000 populations. In general, the disease has been found to be more prevalent among Asians and people of Asian origin. The exact cause of this disease is not known yet.

About 57% of the affected patients are Asian and 71% are female. Although the disease may be seen in any age group, it is more common in people from 5-15 years and 30-40 years of age. Family history is present in about 10%-15% of the patients [2]. There is sparse data on moyamoya disease from the Indian population [3].

Keywords CT angiography; Extra cranial vessels; Cerebral arteries

Clinical Features

The clinical presentations of Moyamoya disease include TIA, ischemic stroke, hemorrhagic stroke, seizures, headache, and cognitive impairment. The incidence of each symptom varies according to the age of the patients. An ischemic event is the most important clinical manifestation of MMD.

Cerebral hypo perfusion due to progressive major vessel occlusion results in repeated hemodynamic TIAs or ischemic strokes in children or young adults. Thus, Moyamoya disease is a classic example of a hemodynamic cause of stroke [4].

Here authors describe 2 cases with clinical and radiological findings consistent with moyamoya disease in young females aged 32 and 24 respectively.

Case 1

A 32-year-old female presenting with symptoms of headache and dizziness treated with anti-migraine medications, but as there was no improvement in her symptoms subjected for CT Angiogram as shown in Figure 1.

CT angiography of intracranial and extra cranial vessels

Neck vessels

- Arch of aorta is normal in caliber.
- The origin of innominate common carotid and subclavian arteries are normal.
- Both common carotids are normal in course and caliber.
- Bifurcation is normal. No evidence of splaying.
- Significant long segment stenosis noted in both internal carotid arteries (involving extra/intracranial segments) from its origin causing approximately more than 90% luminal narrowing.
- Both external carotid arteries are normal in course and caliber.
- Both vertebral arteries are normal in caliber.

Brain vessels

- Prominent PCOM noted bilaterally.
- Hypoplastic A1 segments of left anterior cerebral artery.
- Both anterior and middle cerebral arteries show significant luminal narrowing of more than 90%.
- Multiple collaterals noted joining the anterior and middle cerebral arteries.
- Hypoplastic P1 segment of left PCA.
- Hypoplastic right vertebral artery noted.
- Left vertebral artery is normal in course and caliber.
- No evidence of aneurysm/arteriovenous malformation.

Impression

- Significant steno-occlusion of both internal carotid, middle cerebral and anterior cerebral arteries.
- Multiple collaterals joining the middle and anterior cerebral arteries.

*Above features likely suggestive of a progressive arteriopathy and sequelae Moyamoya disease.

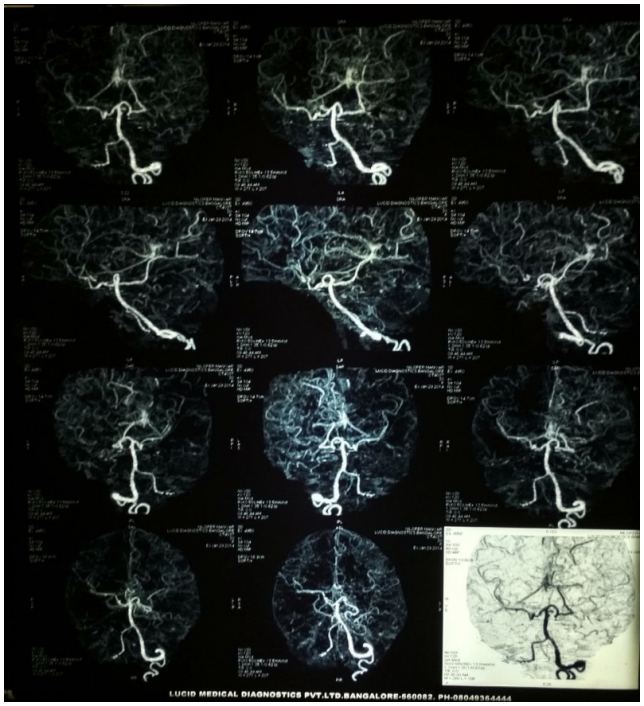

Figure 1: CT Angiography of intracranial and extra cranial vessels.

Case 2

A 24-year-old female presented with symptoms of recurrent hemiparesis, expressive aphasia preceding with headache having waxing and waning of symptoms, CT angiogram was done as shown in Figure 2.

Angiography study shows

- Arch of aorta and major arteries arising from it are normal in caliber with good contrast opacification.
- Both the common carotid arteries are normal in caliber with adequate contrast opacification.
- Vertebral arteries show normal in caliber and good contrast opacification.
- The cervical and intracranial portion of right internal carotid artery shows smooth concentric luminal narrowing.
- The left internal carotid artery is within normal limits.
- There is severe luminal narrowing of the M1, M2 and M3 segments of right middle cerebral artery with poor contrast opacification.
- Multiple collaterals are seen adjacent to M1 segment.
- There is paucity of cortical branches of right middle cerebral artery.
- There is hypoplasia of A1 segment of right anterior cerebral artery.
- The left A1 segment and A2 as well as A3 segment of bilateral anterior cerebral arteries show adequate contrast opacification.
- Bilateral posterior cerebral arteries are normal in caliber and with good contrast opacification.
- There is hypoplasia of posterior communicating artery. The right posterior communicating artery is normal.
- Anterior communicating arteries are normal.
- Basilar artery is normal.
- The aortic arch is normal in course and caliber.
- Normal three vessel origin is seen from the arch of the aorta.
- The brachiocephalic artery is normal in course and caliber.

Impression

- Long segment concentric luminal narrowing of cervical and intracranial right internal carotid artery.
- There is also severe narrowing of right middle cerebral artery in its entire extent with multiple small collaterals adjacent to right M1 segment.

These changes possibly represent recanalization of a long segment thrombus involving the right internal carotid artery and middle cerebral artery with residual luminal stenosis, more severe in the right middle cerebral artery. This needs correlation with clinical and laboratory findings to rule out primary thrombotic disease. However, isolated involvement of single medium sized vessel though uncommon cannot be entirely rule out the possibility of Vasculitis and Moyamoya disease.

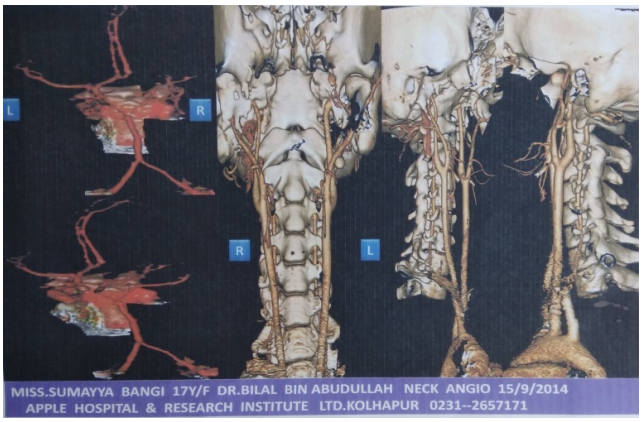

Figure 2: Shows CT angiography study having expressive aphasia preceding with headache and waning symptoms.

Discussion

We report 2 cases, and describe an approach towards the diagnosis of Moyamoya Disease based on clinical and radiological findings. A discrete analysis with reference to CT Angiogram clinches to its diagnosis in adult variety, However involvement of extra cranial arteries and formation of a large thrombus and sequelae of a thrombus makes a differential diagnosis of vasculitis *vs* sequelae of Moyamoya Disease.

All the possible laboratory investigations reveal negativity towards vasculitis and strokes. The possible differential diagnosis in this scenario is Vasculitis, Fibro muscular dysplasia of a vessel, Takayasu arteritis, Giant cell arteritis and Temporal arteritis. Moyamoya disease is not well studied in the Indian Population. The overall annual stoke rate was 4.5% per person-year [5].

Diagnostic criteria include stenosis or occlusion at the terminal portion of the internal carotid artery, abnormal vascular networks in the basal and presence of these angiographic findings bilaterally.

Regarding screening, there is insufficient evidence to screen the asymptomatic individuals or in the relatives of the patients with moyamoya disease in the absence of strong family history or medical conditions predisposing to the disease [4].

Diagnostic criteria previously required that steno-occlusive change at the ICA should be evident bilaterally for the definitive diagnosis of moyamoya disease [6]. Recently, however, diagnostic criteria for definitive moyamoya disease were revised to include patients with unilateral terminal ICA steno-occlusion as well [7].

Extra cranial–intracranial arterial bypass, including anastomosis of the superficial temporal artery to the middle cerebral artery and indirect bypass, can help prevent further ischemic attacks, although the beneficial effect on hemorrhagic stroke is still not clear [8]. The natural history of moyamoya disease tends to be progressive. Untreated patients often suffer cognitive and neurologic decline due to repeated ischemic stroke or hemorrhage [3].

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