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CASE NOTE

Aorta coarctation presenting with intracranial aneurysm rupture

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Abstract: Most vascular diseases have a tendency to affect both heart and the brain. Intracranial aneurysms are more often found in patients with aorta coarctation than in general population, and aneurysm rupture occurs much earlier in these patients. Here, we report a case of aorta coarctation which was diagnosed with its cerebrovascular complications. Before presenting with cerebrovascular complications, the disease can easily be diagnosed with physical examination and non-invasive radiological investigations like echocardiography or cardiac magnetic resonance imaging.

Key words: aorta coarctation; intracranial aneurysm; subarachnoid haemorrhage.

Aorta coarctation is seen in one person out of every 1000 people.[1] It is observed two times more frequently in men than in women. The weakness or lack of lower extremity pulses, arterial pressure difference between arms and legs, the existence of hypertension and cardiac murmur are findings indicating aorta coartation.[2] In untreated cases with severe coarctation, death may occur before age of 40 years owing to congestive heart failure, aorta rupture, aneurysm rupture and infective endocarditis. Although the disease can be diagnosed with routine physical examination, occasionally it may be diagnosed subsequent to the development of cerebrovascular complications.[2]

Case Report

A 15-year-old girl was admitted to our Paediatric Neurology Department with complaints including headache and disability to open her right eyelid. The headache was localised at the right side of the head and around the right eye and started 1 month ago. The pain was unresponsive to analgesics. Two weeks later, diplopia and ptosis of the right eyelid were added to the overall symptomatology.

Key Points

1 Congenital heart disease is a significant risk factor for intercranial aneurysm development.
2 All patients with aorta coartation should be evaluated for intracranial aneurysm at the time of diagnosis.
3 When an underlying intercranial aneurysm is found in patients with aorta coartation, they should be followed up regularly to prevent catastrophic results.

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Her prenatal, natal and family histories were unremarkable. On physical examination, her blood pressure was measured as 130/80 mmHg on the right arm and 120/70 mmHg on the left arm. Measurement on the both legs revealed 85 mmHg systolic pressure. Peripheral pulses were ++/++ in upper, and +/+ in lower extremities. Cardiac auscultation revealed 2/6° systolic murmur at the left 3rd intercostal space expanding towards below left scapula. On neurological examination, the right eye was ptotic, right pupile was mydriatic with weak direct and indirect light reflexes and the movement of the right eye was restricted in all directions except the right outward. There were right central facial paralysis owing to the compressive effect of aneurysm on motor nucleus of seventh cranial nerve at the level of brainstem and hyperaesthesia on the right facial side.

Initial cranial computerised tomography demonstrated subarachnoid haemorrhage within the basal cisterns especially in the right perimesencephalic and the pericavernous regions. Brain magnetic resonance imaging and magnetic resonance angiography revealed a right-sided posterior communicating artery aneurysm with 1.5 cm extending partially into the cavernous sinus which was also ruptured into the same space. There was significant vasospasm in the right proximal middle and anterior cerebral artery distribution. Conventional cerebral angiography revealed the location of the aneurysm clearly (Fig. 1). Contrast enhanced thoracic aorta magnetic resonance imaging showed coarctation of the aorta (Fig. 2). The coarctation segment was short tubular type which localised after the origin the left subclavian artery. Post-stenotic dilatation was seen after the severe coarctation.

The haemorrhage in the cavernous sinus space explained the present clinical symptomatology but the presence of significant vasospasm revealed a contraindication for an acute neurosurgical operation. Acute therapy with high dose nimodipine to prevent an ischemic stroke secondary to the cerebral vasospasm was started. The left-sided hemiparesis which started on the third day of the follow-up progressed to a complete hemiplegia and the patient died because of extensive haemorrhage and ischemic infarction in the right median artery distribution.

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Fig. 1 Conventional cerebral angiography revealing the location of the aneurysm (arrows).



Fig. 2 Contrast enhanced magnetic resonance angiography shows the coarctation of the aorta (arrows).

Discussion

It is widely known that congenital heart disease is a significant risk factor for intracranial aneurysm development, but the pathophysiological basis of this relation is still not known. Generally, the association of aorta coartation, intracranial aneurysm and intracranial aneurysm rupture has been related to arterial hypertension. Arterial hypertension is not a satisfactory explanation for intracranial aneurysm rupture, because intracranial aneurysm ruptures may occur years after surgical repair of the aorta coartation, in the absence of hypertension.[2] Embriologically, cardiac muscular arteries, aortic arc, and cervicocephalic arteries are originated from the neural ridge tissue. Therefore, the association between cardiac pathologies and intracranial aneurysm can be explained with developmental abnormalities originated from the neural ridge.[3]

There are many reasons for the long-term follow-up of patients with coarctation, including re-coarctation, systemic hypertension, aortic aneurysm formation, heart failure, infective endocarditis and cerebrovascular disease. Since the frequency of intracranial aneurysm varies between 2.5% and 50% in aorta coartation cases,[4] all patients with aorta coartation should be evaluated for intracranial aneurysm at the time of diagnosis.

There is controversy about the proper timing of the surgical treatment of an unruptured intracranial aneurysm or an aorta coartation. The incidence of intracranial aneurysm rupture as a cause of death in patients with aorta coartation is higher compared with the estimates of general population. The risk of rupture also exists after surgical repair of aorta coartation, in the absence of hypertension.[2] There are not evidences or suggestions in the literature describing the time of surgery of unruptured intracranial aneurysm in patients with aorta coartation. Only Orsi *et al*.[5] reported repair of two unruptured intracranial aneurysm in the presence of asymptomatic aorta coartation. Therefore, cerebral circulation should undergo examination in every case of aorta coartation, and when an underlying intracranial aneurysm is found, these patients should be followed up regularly.

Magnetic resonance angiography is widely used for the diagnosis of intracranial aneurysms which is the most frequent cause of nontraumatic subarachnoid haemorrhage. The major advantage of this technique is its non-invasive nature, but its lower sensitivity and specificity (varying between 81–95% and 80–100% respectively) with failing to detect the aneurysms smaller than 3 mm[6] makes cerebral angiography more preferable in special circumstances.

Recently, a magnetic resonance angiography study with 100 aorta coarctation patients showed the frequency of intracranial aneurysms with mean largeness of 3.5 mm as 10% whereas nine of them were asymptomatic.[7] This seems to be higher than the normal prevalence in the community (1–2%) which is suggested by arteriographic and autopsy studies.

The reported mean age of intracranial aneurysm rupture in patients with aorta coartation is 25 years old whereas this varies between 50 and 54 years of age in patients without aorta coartation. Also the mortality incidence related to the intracranial aneurysm rupture among the normal population elevates from 0.46–0.59% to 4.8% in cases with aorta coartation.[8] The present case had serious aorta coartation and her intracranial aneurysm ruptured at young age.

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Prognosis in aneurysm rupture is dependent on the size of aneurysm, the presence of previous subarachnoid haemorrhage and diagnosis time. A recent multicentric study showed that patients are considered to have a low risk of rupture that have no history of subarachnoid haemorrhage and have aneurysms located in the anterior circulatory area with the diameter of lower than 7 mm.[9] Owing to present case's age, size and location of the aneurysm, our patient was categorised in the risky group in terms of rupture.

In conclusion, this case indicated the significance of early diagnosis of aorta coarctation that can be detected in routine paediatric physical examination. Because aorta coartation is very easy to diagnose by routine paediatric examination, it should not to be allowed to complicate this cardiac pathology with distressing cerebrovascular complications which can have catastrophical results as in our patient. As the risk of rupture of intracranial aneurysm is higher in patients with repaired or unrepaired aorta coartaion, and there are not evidences or suggestions regarding the time of surgery for intracranial aneurysm in patients with aorta coartation, cerebral circulation should undergo examination in every case of aorta coartation, and when an underlying intracranial aneurysm is found, these patients should be followed up regularly.

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