Moyamoya disease and aortic coarctation in a patient with common brachiocephalic trunk

Moyamoya hastalığı ve aort koarktasyonunun eşlik ettiği bir brakiyosefalik kütük olgusu

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Introduction

Moyamoya disease is characterized by a slowly progressive stenosis and obliteration of the large vessels at the base of the brain, affecting mainly the supraclinoid segment of the internal carotid artery and the initial portion of the anterior or middle cerebral arteries and the posterior cerebral arteries (1). Due to the slow progression of the disease and in response to progressive cerebral ischemia, a large network of collateral vessels is formed from the external carotid arteries, the vertebra-basilar system and other vessels (2).

The idiopathic or primary form of Moyamoya disease, which is sometimes familial, has to be distinguished from the secondary form, referred to as Moyamoya syndrome, which can be associated with certain systemic conditions such as sickle cell disease, chronic basilar meningitidis, neurofibromatosis, X-ray irradiation, homocystinuria and the syndromes of Down, Turner, Alagille and Williams. The association of Moyamoya disease and congenital heart defect such as coarctation of the aorta has been infrequently reported. To our knowledge, there have been only three previous reports of the association of Moyamoya disease with coarctation of the aorta (3-5). However common brachiocephalic trunk (CBT) associated Moyamoya disease has not previously been reported.

We report a case with Moyamoya disease with aortic coarctation and CBT presented with acute convulsion and right hemiparesis.

Case report

The patient is a 7 -year-old boy with previously diagnosed mental-motor retardation and known aortic coarctation. He had been admitted to hospital at 16-month of age because of right sided hemiparesis and convulsion. He had no family history of inherited or cerebrovascular diseases. He was the third child of non-consanguineous parents. The work up that time included a magnetic resonance imaging of the brain revealing multiple areas of infarcts of varying ages involving the left frontal, parietal, temporal, occipital, right frontal lobes, centrum semiovale and right basal ganglia (Fig. 1A-B). Magnetic resonance angiography showed normal internal carotid artery on both sides up to the level of bifurcation, beyond which severe stenosis of the both carotid arteries were seen. The network of collateral vessels was demonstrated at the diencephalic region (Fig. 2). A child had been treated with anticonvulsant and subsequently had been advised to have physiotherapy. At the last control, upper limb hypertension was established. The systolic pressure gradient between the right upper and lower limbs was 30 mmHg. Echocardiography revealed a high systolic gradient (45 mmHg) at the aortic isthmus. Cardiac catheterization and angiography revealed CBT, coarctation of the aorta and stenosis at the origin of left subclavian artery (Fig. 3). A cerebral angiography was done at the same time and confirmed the diagnosis of Moyamoya disease. Cerebral angiography demonstrated bilateral stenosis of the supraclinoid segments of internal carotid arteries and multiple collateral perforating vessels (Fig. 4A-B).

This typical radiological and angiographic appearance, together with the clinical picture, allowed the diagnosis of Moyamoya disease to be made. Extensive investigation in order to exclude biochemical abnormalities (cholesterol, triglycerides and lipoproteins), haematological abnormalities (including antithrombin III, proteins C and S, plasminogen, factors VII and VIII, folic acid, B6 and B12 and factor V Leiden mutation, lupus anticoagulant and anticardiolipin antibodies) and metabolic disorders (including plasma lactate and ammonia, blood gases, plasma homocysteine, serum and urine-aminoacids) did not reveal underlying diagnosis. Narrowed coarctation segment was excised with direct end-to-end circumferential anastomosis of the aorta. Surgical therapy was planned directed toward revascularization of intracranial vessels for the Moyamoya disease.

Discussion

Moyamoya is accepted as primary if it is isolated (Moyamoya disease), or as secondary (Moyamoya syndrome) if the anomaly is associated with an acquired condition or congenital disorder. In the Moyamoya syndrome, the reasons for the obliterative process are clear, whereas in primary Moyamoya, the pathogenetic factors are unknown. For our case, it is unclear whether he had Moyamoya disease or syndrome because cerebrovascular anomaly was not isolated and was associated with aortic coarctation and CBT.

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In this disorder, pathogenetic factors are unknown. Recently there have been increased reports of Moyamoya associated with other developmental abnormalities of the cerebral and systemic vasculature such as aneurysm, arteriovenous malformation, renal artery stenosis (6). Voros et al reported five patients with Moyamoya disease and accompanying vascular anomalies, suggesting a prenatally determined origin of Moyamoya disease (6).

Although the association between coarctation of the aorta and cerebral aneurysms is not uncommon, the association of Moyamoya disease and coarctation of the aorta has been infrequently reported. Lutterman et al described five patients with Moyamoya syndrome and structural congenital heart disease. In their study, coarctation of the aorta was in three patients (4). In addition to this report two case reports have been published. Shuester

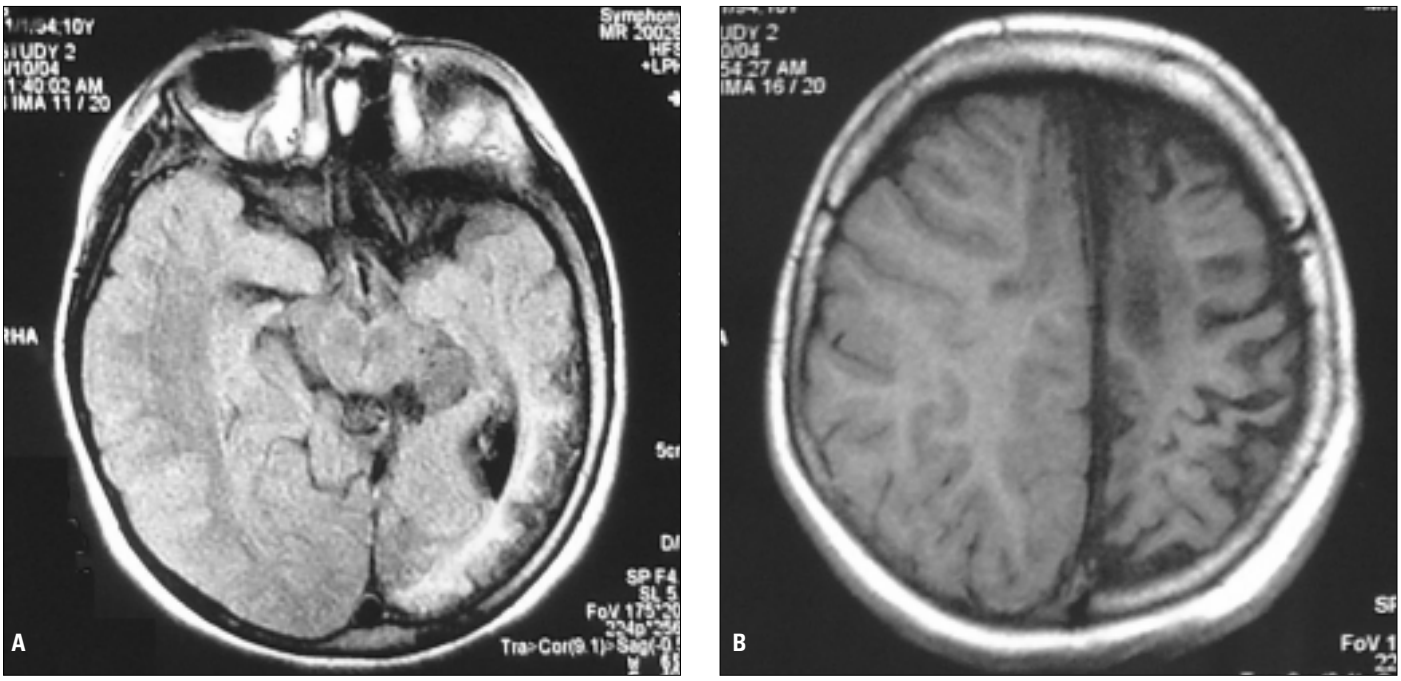

Figure 1. Consecutive axial proton density image at the level of basal ganglia shows bilateral multiple flow voids image (A) and T1-weighted image reveals multiple areas of infarcts of varying ages involving the left frontal, parietal, temporal, occipital, right frontal lobes (B)

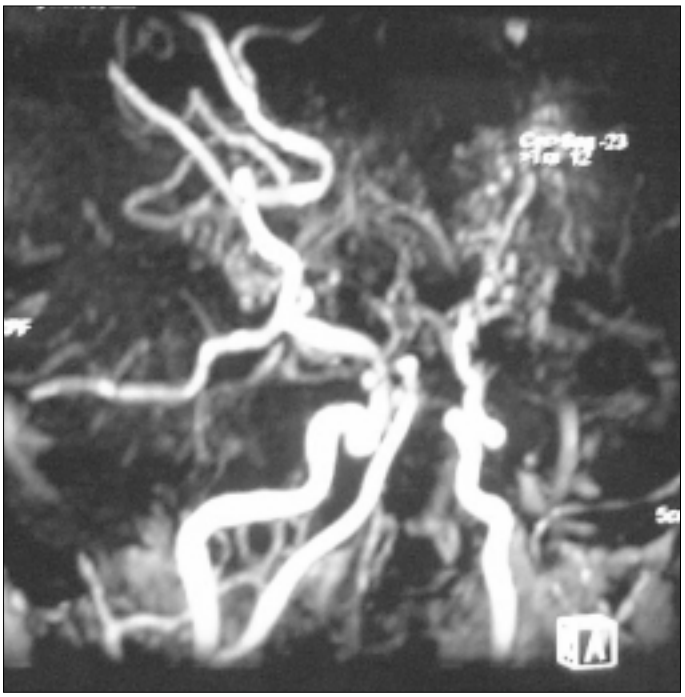

Figure 2. Magnetic resonance angiography shows normal internal carotid artery on both sides up to the level of bifurcation, beyond which severe stenosis of the both carotid arteries are seen. The network of collateral vessels at the diencephalic region is demonstrated

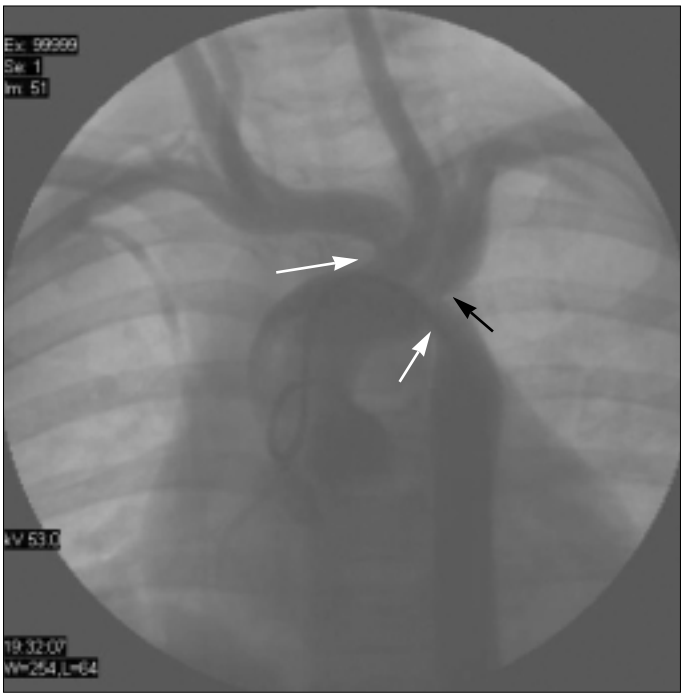

Figure 3. Cardiac angiography shows common brachiocephalic trunk (right white arrow), aortic coarctation (white upper arrow) and stenosis of the left subclavian artery origin (black arrow)

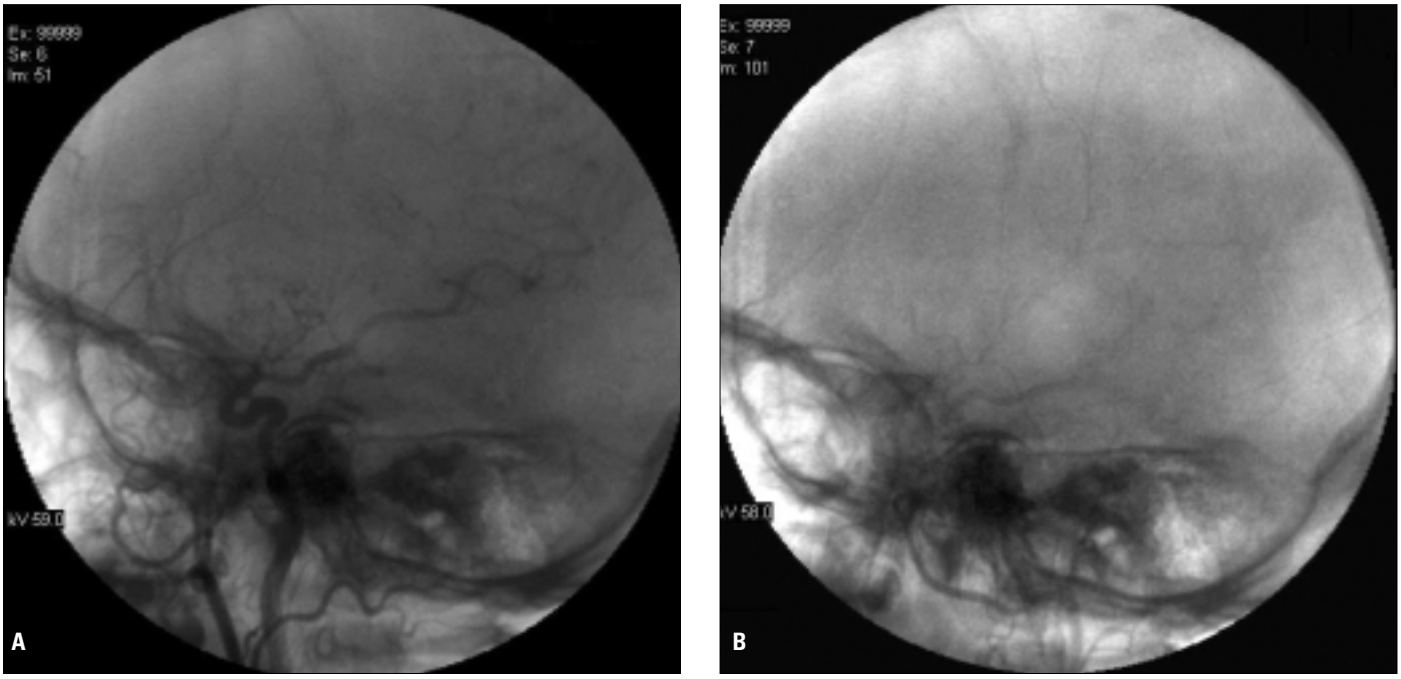

Figure 4. Right (A) and Left (B) carotid artery angiography reveals bilateral stenosis of internal carotid arteries and abnormal vascular collateral networks "puff of smoke" that develop adjacent to the stenosis

and Roberts described a 7 year-old girl with a previous Noonan syndrome (5). Other case of a 4-year-old boy, presented with acute right sided hemiparesis was reported by Christiaens et al (3). To our knowledge our case is the third report.

Common brachiocephalic trunk is an anatomic vascular variant in which both common carotid arteries, together with the right subclavian artery, originate from the aortic arch via a single trunk. Moskowitz and Topaz performed total 1480 cardiac catheterizations in children over a period of 10 years and discovered 48 patients (3.2%) to have a CBT, of whom 98% had associated congenital cardiac malformations (7). In their study the majority of patients with obstructive left heart defects had hypoplasia of the aortic arch, including all patients with coarctation of the aorta, and those with interruption of the aortic arch at the isthmus. Coronary artery abnormalities were seen in 21% of the patients. Moyamoya disease was not seen in their study. On the other hand, an association of CBT and Moyamoya disease has not been described previously to the best of our knowledge.

There may be different explanations for this association. One explanation is that this combination is incidental. Another explanation is that both findings could be the expression of a systemic congenital malformation of blood vessels, as systemic vasculopathies have been described in some cases of Moyamoya (2,8). Intimal thickening and fibroelastosis of the vascular wall are frequent histological findings in Moyamoya disease (9). Some autopsy cases with Moyamoya disease show non-inflammatory fibrodysplasia in extra-cerebral arteries suggesting systemic vasculopathy (10). Vörs et al reported that the small vessel walls displayed hyaline degeneration and intimal and medial hyperplasia with no signs of arteritis (6). In our patient, pathological study of the resected segment of the aorta showed an increase in intimal thickening, but no signs of vasculitis. Thus, we think that systemic vasculopathy may be a pathogenic factor in such cases, and further investigation of etiology and mechanism of Moyamoya syndrome and aortic coarctation is needed.

We believe that careful evaluation of children with congenital heart disease especially coarctation of aorta and symptoms of cerebral ischemia is warranted to detect the presence of Moyamoya disease. Prompt diagnosis and treatment of Moyamoya disease is important in these children to prevent progressive neurological deterioration.

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