TÜBİTAK

CASE REPORT

Turk J Med Sci
2006; 36 (4): 251-253

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Early Onset Stroke Due To Fibromuscular Dysplasia in a Child

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Abstract: Fibromuscular dysplasia (FMD) is a rare, idiopathic and nonatheromatous disease. Definitive diagnosis is made only by angiography, which is characterized by a "string of beads" appearance. In this article, we present the rare entity of early-onset thromboembolic stroke due to FMD in a case who was found to have a carotid artery lesion in angiography. Antiaggregant therapy was given (300 mg/oral/day) and partial improvement was observed on follow-up.

Key Words: Fibromuscular dysplasia, carotid artery, child, stroke, angiography

Çocukta Fibromüsküler Displaziye Bağlı Erken Başlangıçlı Stroke Olgusu

Özet: Fibromüsküler displazi (FMD) nadir görülen, idiopatik, ve nonatheramatöz bir hastalıktır. Kesin tanı anjiografi ile dizilmiş tesbih tanesi şeklindeki görünümle konur. Bu makalede çocukluk döneminde nadir görülen, tromboembolik strokla prezente, karotid arter lezyonlu olgu sunulmaktadır. Antiagregan tedavi 300mg/oral/gün verildi. Kontrolde kısmi iyileşme gözlendi.

Anahtar Sözcükler: Fibromüsküler displazi, korotis, çocuk, inme, anjiyografi

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Fibromuscular dysplasia (FMD) is a nonatheromatous angiopathy primarily affecting medium-sized arteries (1-13). The etiology of the disease is not known, but various opinions have been put forward about the genesis of FMD, such as genetic predisposition, trauma, and underlying connective tissue disease (1,2). The symptoms vary in correlation to the affected arterial region. In patients whose carotids have been involved, most frequently ischemic symptoms, and rarely bleeding, have been reported (4). Definitive diagnosis is made by angiography, which is characterized by a "string of beads" appearance (4,5). In this article, we present a FMD case who had early-onset ischemic stroke due to involvement of the carotid artery.

Case Report

A 12-year-old female patient was admitted to the hospital with the complaint of sudden-onset left-side weakness and speech disorder. Her past and family history was normal and there was no previous systemic disease, transient ischemic attack (TIA) or drug abuse. On physical examination, her blood pressure, heart rate and systemic functions were all within normal limits. On neurologic examination, the patient was fully conscious and cooperative; left central facial paralysis, dysarthric speech and left hemiparesis (3/5) were present. In laboratory investigations, routine blood, urine and biochemical tests were normal. A hypodense infarct area in a right basal ganglia and capsula interna was detected in computed tomography (CT). She had been investigated for the young stroke etiology: hemostatic parameters such as prothrombin time, partial thromboplastin time, bleeding time associated with protein C and S, antithrombin III, and fibrinogen levels; and collagen markers such as rheumatoid factor, antinuclear antibody (ANA), and antiDNA were normal, as were cardiac tests including electrocardiography and echocardiography. In the cerebral angiography, there was a

Received: September 19, 2005
Accepted: June 15, 2006

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"string of beads" appearance in the extracranial carotid artery, which is characteristic of FMD (Figures 1, 2). Renal angiography was normal.

Operation was not recommended by the neurosurgery clinic. We started antiaggregant therapy (acetylsalicylic acid 300 mg/oral/day) and rehabilitation program. On follow-up, her symptoms were partially improved.

FMD is seen mostly in the 4th and 5th decades and in adult women, but rarely affects children (2,4,7,8,12). Ischemic cerebrovascular symptoms, such as vertebrobasilar insufficiency, amaurosis fugax, TIA or cerebral infarcts are observed in the majority of cases with cerebral artery lesion (3,4,7,9,11-13). In asymptomatic cases, the presence of a murmur on

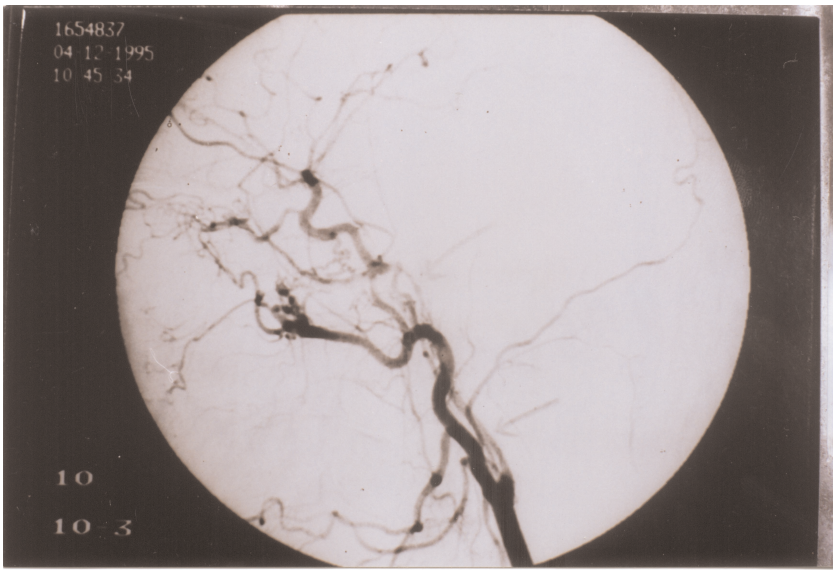

Figure 1. Posterior-anterior carotid angiogram demonstrating the typical "string of beads".

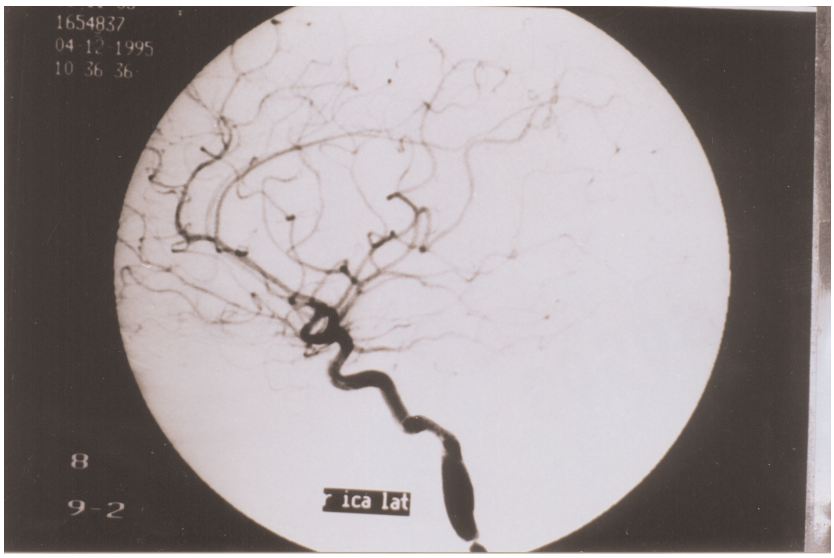

Figure 2. Lateral view of the arteriogram.

auscultation of the carotid artery can be revealed, but in our case, no pathology on carotid auscultation was detected. It is reported that the frequency of aneurysms in FMD is three times higher and subarachnoid hemorrhages more frequent than in the normal population. However, the majority of stroke cases consist of ischemic cerebrovascular events (1,3,4,6,11).

In histopathologic examinations, three different types of disorder in vessel walls are described: intimal fibroplasia, medial fibroplasia and subadventitial hyperplasia. Medial fibroplasia is the most frequent and it is accepted as a characteristic feature of FMD (2,4).

FMD diagnosis can be made only by the characteristic angiographic appearance of a "string of beads" (1-11). Angiographic appearance in our case was also in harmony with FMD. Although the true incidence of FMD is not known, in patients in whom carotid angiography was performed, FMD has been reported at a rate of approximately 0.3-0.9% and bilateral involvement as 60%.

Most of the patients can be effectively treated with antiplatelet agents. However, if the patients have persistent or progressive symptoms, dilatation of the carotid artery may be indicated (1,3,4,6). Surgical intervention is recommended for FMD of the carotid arteries associated with intracranial aneurysms because antiplatelet medication is contraindicated (1,3,4). We preferred the medical approach for our case, and observed partial improvement in her follow-up.

In this article, we emphasize that fibromuscular dysplasia should also be considered in the differential diagnosis of childhood stroke. For this purpose, angiography must be planned for diagnosis. Additionally, patients with FMD must be followed carefully for progression of the central vascular pathology and hypertension.

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