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SHORT REPORT

Extensive Carotid Fibromuscular Dysplasia with Dissections and Aneurysm of the Ascending Aorta

Zafer KOÇ[1], A. Muhteşem AĞILDERE[2], Ayşe Filiz KOÇ[3], Osman KIZILKILIÇ[1], Özlem YALÇIN[1]

[1]Department of Radiology, Faculty of Medicine, Adana Teaching and Medical Research Center, Başkent University, Adana - Turkey

[2]Department of Radiology, Faculty of Medicine, Başkent University, Ankara - Turkey

[3]Department of Neurology, Faculty of Medicine, Cukurova University, Adana - Turkey

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Fibromuscular dysplasia (FMD) is an uncommon vascular disease characterized by increased muscular and fibrous tissue in the arterial wall, which results in alternating dilation and narrowing of vessel segments. The reported prevalence in adults ranges from 0.6% (based on angiographic findings) to 1.1% (based on autopsy findings), and the condition is more frequent in females (1-4). In patients with FMD, abnormalities of the fibromuscular lining of the artery tend to form webs that restrict blood flow, and affected vessels often have a "string-of-beads" appearance on angiography (1,3-5).

The renal arteries are most frequently affected in FMD, but this disease may also involve the carotid, cerebral, mesenteric, coronary and iliac arteries. Patients whose carotid arteries are affected may exhibit disorders related to blood supply from these vessels, such as transient ishemic attacks and stroke (1,4-8). In this report, we briefly summarize a case of FMD with extensive carotid involvement, dissections, aortic aneurysm and stroke. We also discuss the angiographic findings and review the relevant literature. The interesting feature of this case is aneurysm in the ascending aorta and aortic dissections, which may be possible signs of the involvement of these arteries in FMD. We report this case because of this interesting and rare feature.

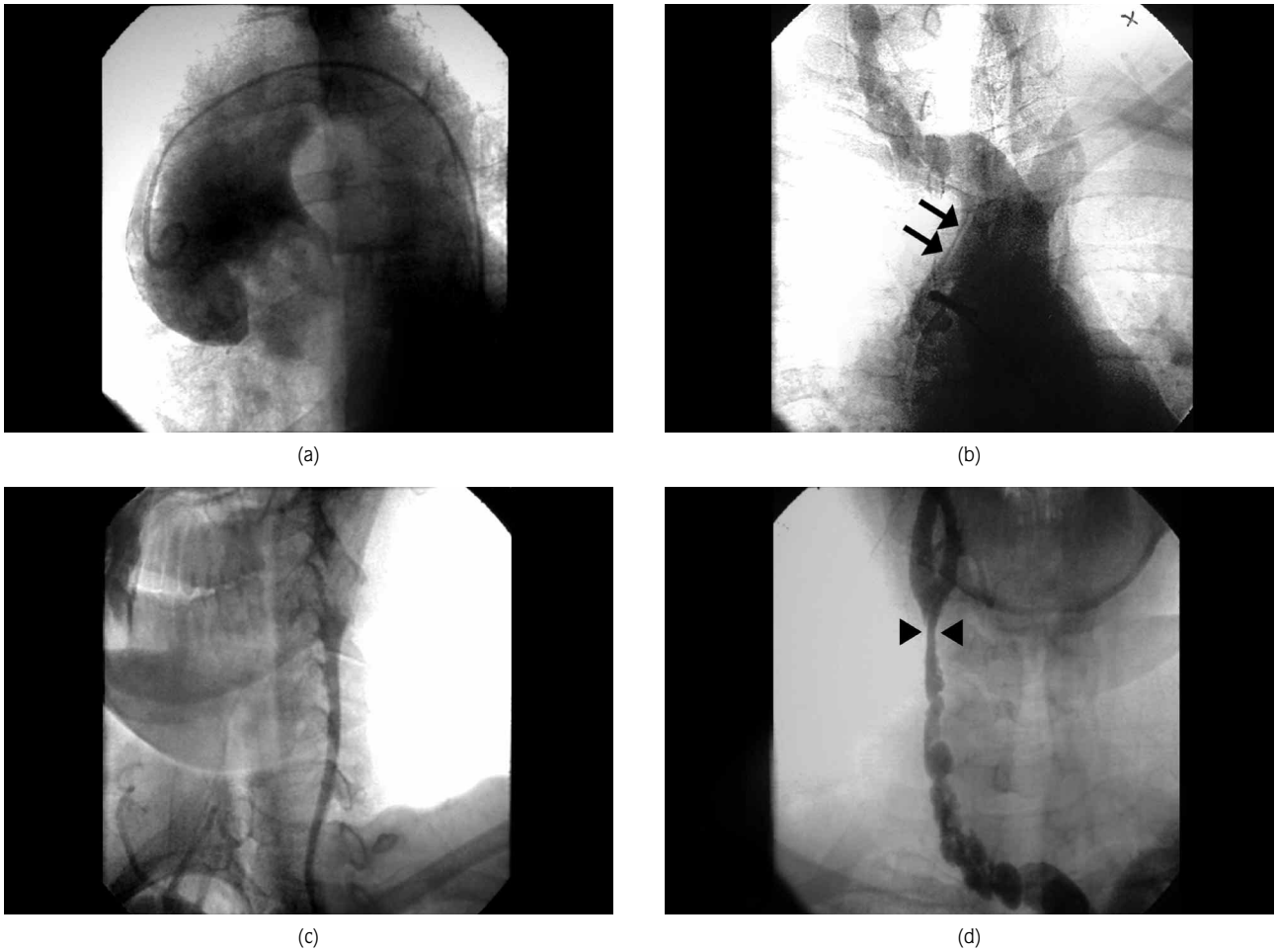
Case

A 35-year-old female was brought to our Emergency Department in a state of acute-onset unconsciousness. There was no history of menstrual irregularity, hypertension, systemic illness or use of oral contraceptives or other drugs. On initial assessment, her blood pressure was 220/120 mmHg, pulse 96/min, and temperature 37.5 ºC. Neurological examination revealed a comatose patient with normal brainstem responses, including light, oculocephalic, corneal and gag reflexes. She exhibited decerebrate posture to painful stimuli. Babinski reflexes were bilaterally indifferent and deep tendon reflexes were brisk.

Her complete blood count was normal except for an elevated white blood cell count of $14x10^9$/l (normal range $4.5\text{-}11x10^9$/l). All blood biochemistry analysis, including ammonium and uric acid, was normal. Lumbar puncture revealed an opening cerebrospinal fluid (CSF) pressure of 270 mmH_2O (normal range 50-180 mmH_2O). The results of CSF analysis were unremarkable. CSF tests for immunoglobulin G and immunoglobulin M for herpes simplex virus types I and II were negative. Anti-streptolysin-O, rhemmatoid factor, anti-nuclear antibody, anti-DNA and lupus erythematosus cell were negative. Additionally, sex hormones including estrogen, progestron, prolactin, free T3 and T4, and TSH levels were normal.

Computed tomography of the brain revealed moderate brain edema and electrocardiography revealed sinus tachycardia. Electroencephalography revealed background activity composed of slow theta and delta waves, and the echocardiographic examination demonstrated an aneurysmal dilatation in the ascending aorta. Angiography performed 3 h after the patient lost consciousness demonstrated a 5.5-cm in diameter aneurysmal dilatation of the ascending aorta (Figure 1a) and dissections in the aortic arch (Figure 1b), the left subclavian artery, the left common carotid artery (Figure 1c), and the distal segment of the right common carotid artery (Figure 1d). The brachiocephalic and right common carotid arteries had a string-of-beads appearance (Figure 1d). Abdominal ultrasonography and renal angiography were normal. Based on these findings, the patient was diagnosed with FMD. Eighteen hour after hospitalisation, O2 saturation and PaO2 decreased and PaCO2 increased. Blood pressure started to decrease. She developed a Cheyne-Stoke's type breathing pattern. Later she required mechanical ventilation and Dopamin infusion was started. The follow-up cerebral CT and/or MRI were not possible because of the patient's condition.

To address the elevated CSF pressure due to brain edema, intravenous treatment with mannitol (600 cc/d, qid) and dexamethasone (32 mg/d, qid) was administered. Antihypertensive medicines that had been applied in a previous medical center (Isosorbide dinitrate, 5 mg/d and Captopril, 75 mg/d tid) were not continued. The patient died due to transtentorial herniation on the third day of hospitalization.



(a) (b) (c) (d)

Figure 1. The patient's angiogram showed an aneurysm in the ascending aorta (a), and dissections in the arcus aorta (arrows in b), and left common carotid artery (c). The string-of-beads appearance in the brachiocephalic and right common carotid arteries (d), and 70% stenosis in the distal right common carotid artery (arrowheads in d) are seen.

Fibromuscular dysplasia is a disease of small and medium-sized arteries that most often affects the primary aortic branches (4). The precise etiology of this condition is unknown, but multiple factors are thought to be involved. The proposed contributors include genetic predisposition, vasculitic and connective tissue disease, hormonal disturbances, and alpha-1 antitrypsin deficiency (5, 9-11). In our patient collagen tests were negative and hormone levels were normal. Alpha-1 antitrypsin deficiency predominantly affects the lungs and the liver. In the lungs, this deficiency produces a chronic progressive lung disease such as emphysema or asthma. The symptoms of liver disease due to alpha-1 antitrypsin deficiency in adults are those seen in liver cirrhosis: jaundice, abdominal swelling, bleeding into the gut and, ultimately, coma. Additionally, it is associated with an increased risk of hepato-cellular carcinoma. This cancer may be the first sign of disease. In our patient, liver function tests, ammonium level, and abdominal US were normal. Her past history revealed no lung or liver disease findings.

The cephalocervical vessels are affected in 29% of FMD cases (4,5), and spontaneous artery dissection occurs in 10-20% of these patients (3,8,10). The distal internal carotid artery is the most frequent site of cephalocervical artery involvement (5,6), and common carotid artery involvement is uncommon. Involvement of the carotids and/or the intracranial arteries may cause amaurosis fugax, transient ischemic attacks and thromboembolic stroke, and may change the level of consciousness from lethargy to coma (5,7,9). Our patient suffered a stroke because of common carotid, brachiocephalic and left subclavian artery involvement. Renal angiography should always be considered in cases of carotid FMD, because of possible renal artery involvement. If FMD is encountered anywhere in the peripheral circulation, the carotid arteries should be evaluated by angiography.

Fibromuscular dysplasia is categorized into 3 different types based on histological and angiographic findings in the arteries. Type 1 is the most common and is characterized by medial fibroplasia. These cases exhibit a string-of-beads appearance on angiography (1,4,6). This appearance is produced by sequential occurrence of luminal stenosis and aneurysmal outpouchings, and by lengthened carotid arteries that are tortuous, coiled and kinked (7,8). Type 2 is much less common than Type 1. In these cases, the primary abnormality is intimal fibroplasia, and affected arteries show long segments of concentric cylindrical narrowing (1,8). Type 3 is rare. It features subadventitial fibroplasia, and is characterized by involvement of only one side of an artery wall. Diverticulation occurs at these weakened sites, and these lesions are visualized on angiograms (8). The typical string of beads appearance seen in the right common carotid artery on angiography is consistent with type 1 FMD in our case.

This case was evaluated as young stroke based on admission of the patient with the complaint of sudden loss of consciousness, detection of high blood pressure upon physical examination at first admission, the absence of any abnormal blood biochemistry result, the fact that there was no history of intoxication or infection, the presence of decerebrate rigidity upon neurological examination and the acute progression of the clinical course of the disease. The absence of computed tomography evidence of intracranial hemorrhage and intracranial mass lesions or findings suggestive of central nervous system infection explaining such a severe clinical appearance also supported the diagnosis. The presence of the aneurysm in the ascending aorta and its dissection in echocardiography also suggested stroke. The absence of any cells in CSF obtained by lumbar puncture excluded CNS infection as a cause of this presentation. Angiography demonstrated dissections in the aortic arch, left subclavian artery, left common carotid artery, and the distal segment of the right common carotid artery, as well as the string-of-beads appearance in the brachiocephalic and right common carotid arteries (Figure 1a-d). All these findings suggested FMD. Additionally, these findings confirmed the diagnosis of stroke on the basis of FMD considering a young female patient.

Fibromuscular dysplasia generally affects mainly primary aortic branches (4). Studies have shown that patients with FMD may have multiple aneurysms (5,6). The most common sites for these aneurysms are the small and medium-sized arteries (5,6). Diffuse, thoracic, and abdominal aortic involvement has been reported in rare cases (12-16). Thoracic descending aortic involvement has been reported as atypical coarctation (13,15,16), and ascending aortic aneurysm and dissection are rare findings in cases of FMD (12). In our case, we suspected that the proximal aortic aneurysm and the aortic dissection were due to FMD.

Fibromuscular dysplasia should be included in the differential diagnosis for any young patient who presents with sudden-onset stroke and no previous warning signs of the illness. Although aneurysm and dissections of thoracic aorta are rare findings in cases of FMD, it should be considered a potential cause of thoracic and abdominal aortic aneurysm and dissection in female patients with no atherosclerosis or other risk factors.

Corresponding author:

Zafer KOÇ

Başkent University, Faculty of Medicine,

Adana Teaching and Medical Research Center,

Department of Radiology, Adana - Turkey

E-mail: zaferkoc@superonline.com

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