Fibromuscular Dysplasia and Intravenous Thrombolytic Treatment

Fibromusküler Displazi ve İntravenöz Trombolitik Tedavi

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ABSTRACT

Fibromuscular dysplasia (FMD), which usually affects middle-aged and older women, is a non-atheromatous and non-inflammatory angiopathy. Definitive diagnosis is made only by angiography showing classic string-of-beads appearance. In this article, we present a patient with acute ischemic stroke due to FMD who was successfully treated with thrombolytic therapy as well as to revise the approach to FMD in the light of the literature. (*Archives of Neuropsychiatry 2014; 51: 175-177*)

Key words: Fibromuscular dysplasia, stroke, thrombolytic treatment

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ÖZET

Fibromusküler displazi (FMD) ateromatöz ve inflamatuvar özellikte olmayan, genellikle orta ve yaşlı kadınları etkileyen, segmental tutulum gösteren bir anjiopatidir. Kesin tanı klinik özelliklerin varlığı ve dijital subtraksiyon anjiyografide ipe dizilmiş tesbih tanesi görünümünün saptanması ile konur. Bu makalede akut gelişimli iskemik inme nedeniyle değerlendirilerek trombolitik tedavi uygulanan ve daha sonraki etiyolojik tarama sırasında FMD saptanan bir olgu ve bu olgu bağlamında FMD'ye bağlı iskemik inme geçiren olgularda trombolitik tedaviye bakış sunulmaktadır. (*Archives of Neuropsychiatry 2014; 51: 175-177*)

Anahtar kelimeler: Fibromusküler displazi, inme, trombolitik tedavi

Çıkar Çatışması: Yazarlar bu makale ile ilgili olarak herhangi bir çıkar çatışması bildirmemişlerdir.

Introduction

Fibromuscular dysplasia (FMD) is a non-atheromatous and non-inflammatory angiopathy with segmental involvement affecting women aged 40-50 years. Involvement of some arteries is observed more frequently in FMD. Renal arter involvement is observed in 60-75% of all patients, carotid and vertebral artery involvement is observed in 25-30% and multiple arterial involvement is observed in 28%, whereas isolated mesenteric artery or iliac artery involvement is observed very rarely. Cervicocephalic FMD is observed in less than 1% of the population and at the C1-2 vertebral corpus level. FMD of the intracranial arteries is less common and is limited to the intrapetrous part of the internal carotid artery or to the syphon of the carotid artery. Intracranial aneurysm may accompany in 7-51% (1,2,3). The patients usually present with headache, tinnitus, vertigo, transient retinal and cerebral ischemia, cerebral infarction or subarachnoid hemorrhagia. Although the cause is not known exactly in the etiopathogenesis, it has been proposed that immunological or

estrogenic action on the arterial wall, trauma or presence of underlying collagen tissue disease may have a role (4,5). Cerebral ischemia has been associated with arterial sclerosis or arterial thromboembolism. The definite diagnosis in FMD is made by digital subtraction angiography (DSA) showing classic string-of-beads appearance. This finding which is obtained in 90% of the cases is considered characteristic for FMD (6,7).

Here, the experience obtained from a patient who presented to our unit because of acute ischemic stroke, received IV thrombolytic treatment in the acute period and diagnosed subsequently with FMD following investigations performed to determine the etiology is shared.

Case

A 44-year old female patient was evaluated in the emergency outpatient clinic because of acutely developed right side weakness and speech difficulty. She had no history of systemic disease. The initial neurological examination revealed mixed

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type aphasia and right hemiparesia (3/5) with predominant motor component and hypoactive deep tendon reflexes on the right side (National Institute of Health Stroke Scale (NIHSS):13). In the acute period, there was no hemorrhage on computerized brain tomography (CBT). The ASPECT score was found to be 8 (Figure 1). ECG was found to be normal. There was no pathology in the patient's personal and familial history. The blood pressure measurements, blood biochemistry tests and complete blood count performed in the emergency department were found to be normal. All contraindications in terms of IV thrombolytic treatment were questioned and it was decided to administer IV thrombolytic treatment after obtaining consent from her relatives. Thrombolytic treatment was administered approximately 2,5 hours after the onset of symptoms. Headache, hypertension, peripheral hemorrhage and alternation in consciousness were not observed during the follow-up. On follow-up CBT performed

Figure 1. Initial CT is normal

Figure 2. Follow-up CT shows left hemispheric acute infarct and callosal hemorrhage

after the initial 24 hours, hemorrhage was present in the corpus callosum splenium and pericallosal space which developed secondary to thrombolytic treatment in addition to acute ischemic lesion at the left temporal lobe level (Figure 2). On neurological examination performed 48 hours after treatment, hemiparesia on the right side at a level of 4+/5 and sensorial aphasia with predominance of difficulty in naming were found (NIHSS:4). Considering the age of the patient, lipid profile, lipid electrophoresis, protein, antithrombin III, factor Leiden mutation, ANA, anti DNA, HgA1c, Homocysteine, B12, thyroid function tests and tests directed to chronic infections were performed. No significant pathology was found. Transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) were found to be normal. DSA revealed focal stenosis in both internal carotid arteries, vertebral artery and vertebral artery V3, V4 segments and string-of-beads appearance in the left renal artery (Figure 3,4).

The case was considered thromboembolic stroke which developed on the background of FMD. 24 hours after thrombolytic treatment acetylsalicylic acid was initiated at a dose of 150 mg/day and a rehabilitation program was started. In the clinical fol-

Figure 3. DSA shows string of beads in the left renal artery due to stenosis

Figure 4. DSA shows stenosis in the left ICA

low-up, marked improvement occurred in aphasia findings. The patient was discharged with findings of sensorial aphasia and right hemiparesia at a level of 4+/5 (NIHSS:3). After one year, a very mild difficulty in naming was continuing and she returned back to active working life.

Discussion

FMD can be defined as a clinical process in which difficulties are experienced in diagnosis and treatment. It is mostly found during the investigations performed after development of cerebrovascular events in young individuals with stroke. In these patients who are generally asymptomatic, involvement is found primarily in the renal arteries and secondarily in the carotid arteries. Renovascular hypertension related with renal artery involvement has been reported with a rate of 4/1000 and cerebrovascular diseases related with involvement of the carotid arteries has been reported with a rate of 2/1000 (8,9). Ischemic stroke is reported with a rate of 3.6% in patients with FMD. Aneurism and related intracranial or subarachnoid hemorrhage are found with a 3-fold higher rate compared to the normal population (8,9). Since both ischemic and hemorrhagic stroke can be observed in these patient, treatment should be adjusted to target the primary event.

Systemic thrombolytic treatment with TPA is considered gold standard in adult patients with ischemic stroke. After exclusion of treatment complications and difficulties in specifying indications, thrombolytic treatment has become a mandatory treatment method for appropriate patients and under appropriate hospital conditions which is currently preferred frequently rather than being a limited treatment method. This treatment method made a breakthrough in the prognosis of the disease and the morbidity decreased markedly (11,12).

Thrombolytic treatment has been used in a limited number of cases for FMD patients (13). Thrombolytic treatment was tried for the first time in a 12-year old girl who was evaluated in the emergency outpatient clinic with a picture of right hemiparesia and global aphasia which developed acutely and diagnosed with ischemic stroke in Mannheim Heidelberg University (12). The NIHSS was found to be 19 initially, 15 after thrombolytic treatment, 8 at the time of discharge and 5 at the follow-up visit performed 3 months later. In investigations performed to find the etiology after thrombolytic treatment, the patient was identified to have FMD and no difference was found on the follow-up cerebral MRI and angiography compared to the previous investigations. After our patient was diagnosed with acute ischemic stroke attack, she received thrombolytic treatment and a diagnosis of FMD was made after investigations. Her findings improved one year later such that she could go back to active working life.

Our patient is the second patient who received IV thrombolytic treatment for acute ischemic stroke related with FMD in

the literature. As a result of a long-term follow-up, the desired objective was obtained in treatment. The most important problem here is the fact that different types of stroke may occur in patents with FMD and this may create contraindication in terms of thrombolytic treatment. However, this possibility is valid for all patients with stroke caused by different etiologies. Since no definite judgement can be made with only two patients, more precise views will arise as the experiences in this are increase.

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