The Evaluation of Vertebrobasilar Artery System in Neuro-Behçet and Behçet Disease using Magnetic Resonance Angiography

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ABSTRACT

The aim of this study is the evaluation of the vertebrobasilar artery system in patients with Behçet's and Neuro-Behçet's disease. For this aim; 20 adults with clinically diagnosed Behcet's disease, 20 adults with Neuro-Behçet's disease, and 19 age- and gender-matched controls were examined by magnetic resonance angiography (MRA). During MRA, diameters of left vertebral artery (LVA), right vertebral artery (RVA), basilar artery (BA), and proximal segment (P1) of posterior cerebral artery between origin and junction with the posterior communicating artery were measured. In all groups, LVA was dominant than RVA ($P < 0.05$). The diameters of BA and right P1 of Neuro-Behçet's disease were larger than the other groups ($P < 0.05$). In addition, the diameters of left P1 of Neuro-Behçet's disease were larger but not statistically significant. There is no difference between the groups in terms of gender. Behçet's disease can affect vascular structures; therefore vertebrobasilar artery system should be examined in patients with Behçet's and Neuro-Behçet's disease. Anat Rec, 297:1302–1305, 2014.

Key words: Behçet's disease; Neuro-Behçet's disease; vertebrobasilar artery system

Behçet's disease (BD) is a systemic inflammatory disorder of unknown etiology and has a wide distribution with higher incidence rates in Mediterranean countries and the Far East (Benamour et al., 1990; Chaillou et al., 1992; Davatchi et al., 2010.). It is characterized by recurrent oral and genital aphthous ulcers and uveitis. The pathological lesion is a vasculitis, involving veins, venules, capillaries, and less frequently, arteries. It can affect the skin, joints, intestines, lungs, heart, and central nervous system (Al Kawi et al., 1991; Neudorfer et al., 1993; Colak et al., 2011). Neurological involvement of BD is caused by paranchymal or vascular injury and range from 5 to 30% (Öktem-Tanör et al., 1999; Joseph and Scolding, 2007).

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Neuro-Behçet's disease (NBD) usually shows a brainstem or a meningomyelitic syndrome and an organic confusional syndrome (Lee et al., 2001). Radiologically, NBD involves the white matter, brainstem, basal ganglia and thalamus; neurological symptoms are generally correlated with radiological findings (Wechsler et al., 1993).

The vertebrobasilar artery system (posterior circulation) supplies blood to the posterior part of the cerebral hemispheres, the cerebellum, and the brainstem (Pai et al., 2007; Davim et al., 2010). The vertebral arteries converge medially as they ascend the medulla and unite to form the basilar artery (BA) approximately at the level of the junction between medulla and pons. BA terminates by dividing into two posterior cerebral arteries at a variable level; however most frequently in the interpedincular cistern (Crossman, 2008; Wake-Buck et al., 2012).

The serious clinical consequences can occur by arteriopathy; so in this study, we aimed to determine the structure of vertebrobasilar circulation in patients with BD and NBD by magnetic resonance angiography (MRA).

MATERIALS AND METHODS

Subjects

This study was conducted on MRA scans of 20 adults with clinically diagnosed BD, 20 adults with NBD and 19 control subjects matched by age and gender. MRA scans of consecutive BD and NBD patients admitted to the Department of Radiology, Faculty of Medicine, İnönü University, were taken. All patients satisfied the criteria for Behçet's disease recommended by the International Study Group (ISG) (ISG for Behçet's Disease, 1990). The patients have no any other systemic disorders and were stable during MRA. Our study was conducted according to the guidelines of the Declaration of Helsinki and approved by the Ethics of Research Committee of İnönü University Faculty of Medicine (2011-27) and, were obtained from all the participants.

All of the patients with BD were taking colchicine (0.5–1 mg daily) and had inactive disease at the time of the study. Exclusion criteria were accepted as the presence of cervical pain or stiffness, any disease with neurological involvement, carotid artery involvement, chronic liver and kidney failure, body mass index of 30 and above, hypertension, heart failure, diabetes mellitus, vasculitis, a systemic disease such as rheumatoid arthritis, systemic lupus erythematosus, and so forth.

MRI Examinations

MRI scans were performed by 1.5 TMRI system (Gyroscan Intera Master, Philips) using a standard head coil for our study. We obtained axial T2-weighted images by turbo spine echo sequence (slice thickness: 6 mm), FLAIR and T1-weighted conventional MRI images. Being given 0.2 mmol/kg contrast agent (gadolinium) using a 18G IV catheter with automatic injection pump, T1A axial and sagittal images were obtained.

MRA Examinations

MRA was performed during conventional MR using neck coil after injected 0.2 mmol/kg i.v. contrast agent. The images from aortic arch to circle of Willis obtained by TE 50 ms, TR 500 ms FFE method were evaluated by Maximum Intensity Projection. During MRA, diameters of left vertebral artery (LVA), right vertebral artery (RVA), BA and proximal segment (P1) of posterior cerebral artery between origin and junction with the posterior communicating artery were measured.

Statistical Analysis

Data were tested for normal distribution using the Kolmogorov-Smirnov test. For normally distributed data, group comparisons were made using one-way ANOVA followed by Tukey post-hoc test. The results were expressed as means ± standard deviation (SD). For nonnormal distributed data, the results were compared with Kruskal–Wallis test. Multiple comparisons of group means were compared using the Mann–Whitney *U*-test. The results were expressed as median (min–max). The level of significance was set at $P < 0.05$.

RESULTS

Of 20 Behçet's patients in the study, 13 (65%) were female and 7(35%) were male. Among 20 Neuro-Behçet patients, 11 (55%) were female and 9 (45%) were male. In all groups, LVA was dominant than RVA ($P < 0.05$). The diameters of BA and right P1 of Neuro-Behçet's disease were larger than the other groups ($P < 0.05$) (Fig. 1). In addition, the diameters of left P1 of Neuro-Behçet's disease were larger; but not statistically significant (Table 1). There were no differences among the groups in terms of gender (Table 2). The neurological involvement of Neuro-Behçet's disease was also confirmed by MRI.

DISCUSSION

BD is a multisystem disorder first described by Hulusi Behçet, a Turkish dermatologist, and consists of a triad of recurrent ulcers of the oral and genital mucosa with relapsing uveitis (Behcet, 1937; Akpolat et al., 2000). In the current study, we evaluated vertebrobasilar artery system in BD and NBD using MRA. Neurological involvement develops more than 10 years after onset BD (Akman-Demir et al., 1999; Matsuo et al., 2005; Chae et al., 2008). Parenchymal involvement primarily affects the brainstem, spinal cord, and cerebral hemispheres. Nonparenchymal involvement includes intracranial hypertension, aseptic meningitis, cranial neuropathy, and cerebrovascular disorders such as dural sinus thrombosis, arterial dissection, occlusion, and aneurysm (Kidd et al., 1999; Matsuo et al., 2005; Chae et al., 2008).

Vascular lesions are arterial occlusion, arterial aneurysm, venous occlusion, and venous varicosities in BD (Matsumato et al., 1991; Koçak et al., 2004). Aneurysm formation in neuro-vasculo-BD usually happens due to rupture of the internal and external elastic laminae, with thickening of the tunica intima, degeneration of the tunica media, and vasculitis of the vasa vasorum, with perivascular infiltration, predominantly lymphocytes. One hypothesis suggests that autoimmune complexes are accumulated, causing aseptic embolization of the vasa vasorum or lumen of the artery, with resultant focal arteritis and necrosis, which erodes the blood

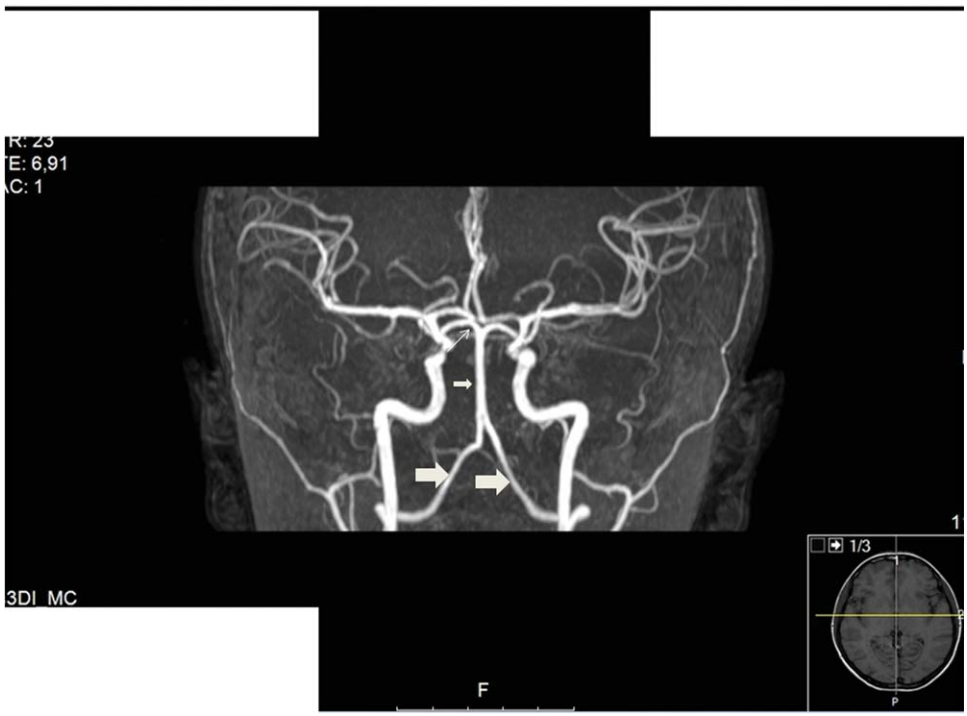

Fig. 1. Left vertebral artery (LVA) (➡), right vertebral artery (RVA) (➡), basilar artery (BA) (➡), left proximal segment (P1) of posterior cerebral artery, and right proximal segment (P1) of posterior cerebral artery (↗) in patient with neurobehçet disease are shown.

TABLE 1. Diameters of left vertebral artery (LVA), right vertebral artery (RVA), basilar artery (BA), left-right proximal segment (P1) of posterior cerebral artery of all groups are shown in the table.

Groups	LVA	RVA	BA	Left P1	Right P1
Control	2.60 ± 0.18	2.20 (0.7–3.6)	2.71 ± 0.08	1.30 ± 0.10	1.25 ± 0.07
BD	2,31 ± 0.11	2.10 (1.1–3.2)	2.76 ± 0.10	1.37 ± 0.06	1.44 ± 0.08
NBD	2.62 ± 0.14	2.15 (1.0–3.8)	3.16 ± 0.11[a]	1.52 ± 0.07	1.67 ± 0.05[b]

[a]$P < 0.05$ compared with other groups.
[b]$P < 0.05$ compared with control group.

TABLE 2. The differences among the groups in terms of gender and mean of ages of all groups

Groups	Female	Male	Age
Control	11 (%59.9)	8 (%42.1)	39.8 ± 10.2
BD	13 (%65)	7 (%35)	42.6 ± 12.0
NBD	11 (%55)	9 (%45)	42.5 ± 9.0

$P = 0.47$ (Pearson chi-square test).

vessel from the endothelial surface outward, leading to aneurysm formation (Benamour et al., 1990; Ho and Deruytter, 2005; Aktaş et al., 2008).

Dilatative arteriopaty is defined by an increased length and diameter of arteries. The intracranial VA and BA are preferentially involved. According to the study of Lou and Caplan (2010), four different clinical situations may occur during the dilatative posterior circulation arteriopathy: (1) Asymptomatic; (2) acute vertebrobasilar arterial territory ischemia; (3) chronic progressive course related to compression of cranial nerves, the brainstem, or the third ventricle; and (4) catastrophic outcome caused by vascular rupture.

In the literature, there are a few case reports about BD and aneurysm of intracranial arteries (Nakasu et al., 2001; Kizilkilic et al., 2003; Ho and Deruytter, 2005; Aktaş et al., 2008). In our study, the diameters of BA and right P1of NBD were larger than the control. It can be concluded that fatal aneurysm may occur in patients of our study. Because of clinical importance of intracranial aneurysm, the patients with BD should be evaluated about intracranial circulation. In a case, Aktas et al. (2008) reported fatal subarachnoid bleeding caused by posterior cerebral circulation aneurysm with BD. They discussed that these aneurysms could be related to BD. Therefore, early diagnosis and treatment of aneurysm might prevent complications.

Agrawal et al. (2007) reported that there was no consensus on the treatment of asymptomatic intracranial aneurysms in BD. However, assuming vasculitis process

leading to formation of aneurysm, a trial of immunosuppressive therapy was assured against a possibility of endovascular intervention as shown in this case. Similarly, Ogata et al. (2013) advised that patients with BD should be examined for the aneurysm and an aggressive medical treatment be planned for them. Immunosupresive therapy induces regression of vascular changes (Koçak et al., 2004).

Mohammed et al. (2012) reported arterial flow abnormalities with NBD in 100% of the affected population. Radiographic findings and flow abnormalities were significantly less in patients on immunesuppressants and antiplatelet drugs. So these treatments might retard structural cerebral vascular disease progression and improve flow patterns (Mohammed et al., 2012).

In conclusion, Magnetic Resonance Angiography studies with Behçet's Disease/Neuro-Behçet's Disease have been limited only cases or small series of cases. Based on all these studies, immunsupresive therapy could prevent arterial complications. Since the rate of intracranial dilatative arteriopathy is probably low, the complications of serious diseases should not be underestimated. Therefore patients with Behçet's Disease should be evaluated for intracranial vascular structure.

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