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BRIEF COMMUNICATION **KISA RAPOR**

THREE CASES OF STROKE ASSOCIATED WITH TAKAYASU ARTERITIS

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ABSTRACT

Takayasu arteritis (TA) is a form of large vessel vasculitis characterized by granulomatous inflammation of the vessel wall. It tends to be more prevalent at a younger age and among females. Stroke is a common complication of TA, occurring in 10-20% of cases. However, it is exceedingly rare for stroke to be the initial clinical manifestation. Here, we present three cases of stroke associated with TA. In the first case, a 16-year-old female patient presented with acute stroke symptoms and received thrombolytic treatment. Despite no prior medical history of disease, the patient was diagnosed with Takayasu through subsequent examination and imaging after the stroke. In the second case, a 45-year-old female patient, previously diagnosed with TA, presented with ischemic stroke. Vascular imaging revealed large vessel occlusion believed to be secondary to vasculitis. In the last case, an elderly female patient with TA and additional comorbid diseases experienced a stroke presentation. It is crucial to recognize that stroke can indeed be the initial clinical presentation of Takayasu arteritis. Therefore, clinicians must remain vigilant and consider the risk of stroke in the follow-up of TA patients, as it significantly impacts the disease's course and prognosis.
Keywords: Stroke, Takayasu, vasculitis, thrombolytic.

TAKAYASU ARTERİTİ İLİŞKİLİ ÜÇ İNME VAKASI

ÖZ

Takayasu arteriti (TA), damar duvarının granülomatöz inflamasyonu ile karakterize, erken yaşta ve kadın cinsiyette daha sık görülen bir büyük damar vaskülitidir. İnme, TA'nin %10-20 oranında görülen sık bir komplikasyonu olup, TA'nin ilk klinik prezentasyonunun inme ile olması ise çok nadirdir. TA ile ilişkili üç inme vakası sunulmaktadır. İlk olguda akut inme kliniği ile başvuran ve trombolitik tedavi uygulanan 16 yaşında kadın hasta sunuldu. Özgeçmişinde bilinen tanısı olmayan hasta, inme sonrası yapılan muayene ve görüntülemelerle TA tanısı aldı. İkinci olguda TA dışı tanısı olmayan ve iskemik inme ile başvuran 45 yaşında kadın hasta sunuldu. Hastanin vasküler görüntülemelerinde vaskülite sekonder geliştiği düşünülen büyük damar okluzyonu izlendi. Son olguda ise TA ve ek komorbid hastalıklara sahip olan ve inme ile başvuran ileri yaşta kadın hasta sunuldu. Takayasu arteritinin ilk klinik prezentasyonunun inme olabileceğinin akılda tutulması, TA tanısı mevcut hastaların takibinde ise inme riskinin göz ardı edilmemesi hastalığın seyri ve prognozu açısından önemlidir.
Anahtar Sözcükler: İnme, Takayasu, vaskülit, trombolitik.

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INTRODUCTION

Takayasu arteritis (TA) is a large vessel vasculitis characterized by granulomatous inflammation of the vessel wall. İt is more common in early years and in females (1,2). Stroke is a common complication of TA, occurring in 10-20% of cases. It is very rare for the first clinical presentation to be a stroke (3,4). Here, we present three cases of stroke associated with TA. Written consents have been obtained from all patients for this report.

CASE 1

No significant history was noted for a 16-year-old female patient who presented with complaints of right-sided weakness and inability to speak. The patient was conscious but unresponsive to commands, with no verbal output. Nasolabial sulcus was obscured on the right side, and muscle strength was graded as 1/5 in the upper right and 3/5 in the lower right limbs. Bilateral radial pulses were not palpable, while proximal and distal pulses of the lower extremities were normal. At 90 minutes post-symptom onset, the NIH score was 15. Intravenous tissue plasminogen activator (IV-tPA) was administered due to acute ischemic infarction detected in the left MCA territory on diffusion MR (DWI) images (Figure 1). Cranial and carotid CT angiography revealed occlusion of the left ICA, along with multiple stenoses in the brachiocephalic trunk (Figure 2). The case presenting in the acute phase was evaluated together with the interventional neuroradiology unit regarding the indication for thrombectomy. Thrombectomy was not planned due to the risk of dissection in the patient with findings suggestive of vasculitis on CT angiography imaging.

Routine laboratory tests including biochemistry, hemogram, and sedimentation rate were within normal limits. Despite negative vasculitis markers, TA was diagnosed based on imaging findings. The patient received 1000 mg/day pulse steroids for 3 days and a single dose of cyclophosphamide. Subsequently, the NIH score decreased to 11 during follow-up, and the patient was discharged on ASA 300 mg/day. The patient was given monthly cyclophosphamide therapy and oral prednisolone therapy. Following the suppression of inflammation, the patient underwent revascularization surgery. Unfortunately, the patient was lost due to sepsis developed after a successful operation.

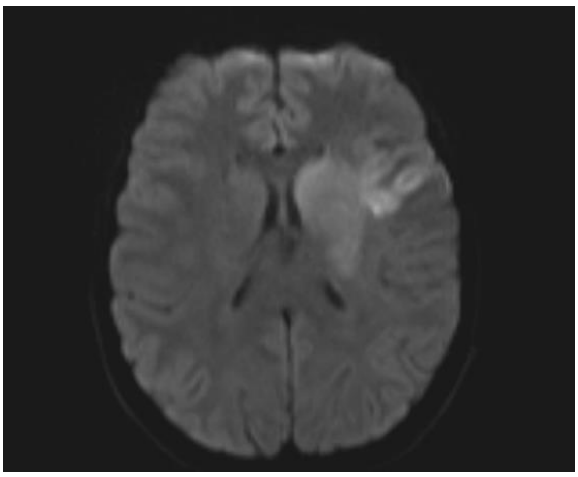

Figure 1. Acute infarction in the left MCA area.

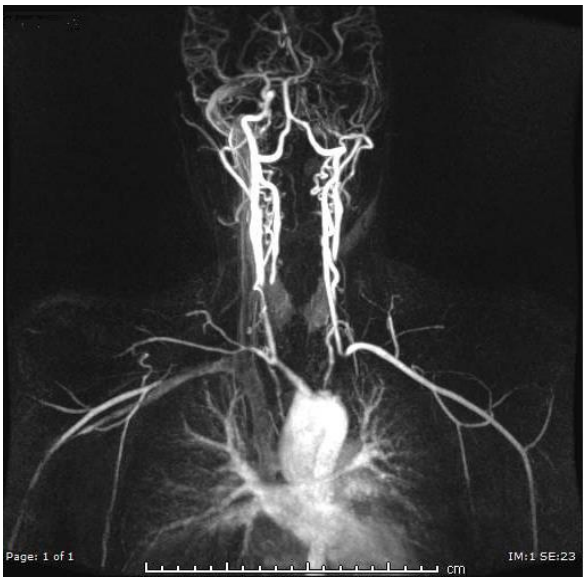

Figure 2. Occlusion on left ICA.

CASE 2

A 45-year-old female with known TA presented with left-sided numbness. Neurological examination revealed full consciousness, cooperation, and orientation. Muscle strength was graded as 5/5 in the upper and lower extremities, with left hemihypoesthesia. The patient was on regular methotrexate therapy. DW-MRI revealed hyperintensity in the territory of the right MCA, consistent with an acute ischemic infarction. Additionally, CT angiography of the head and neck

showed total occlusion of the left ICA and stenosis in the right ICA (Figure 3, 4). With an NIH score of 3, IV-tPA was not administered. Routine laboratory tests were unremarkable, and the patient was discharged on dual antiplatelet therapy without complications.

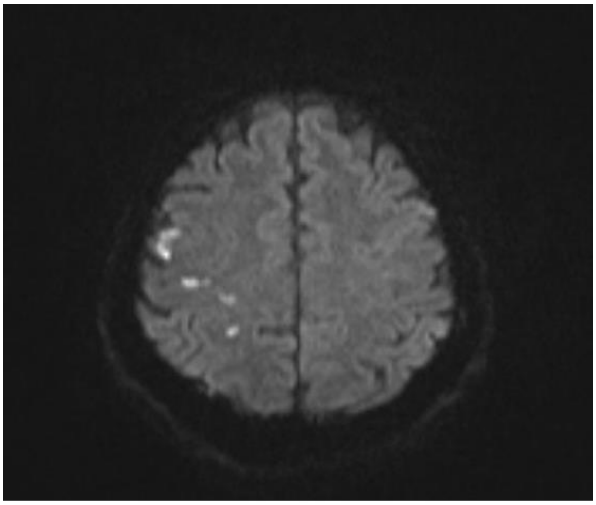

Figure 3. Acute infarction in the right MCA area.

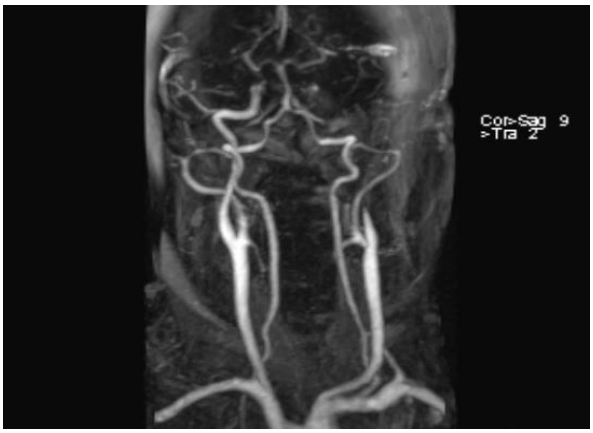

Figure 4. Total occlusion of the left ICA and stenosis in the right ICA.

CASE 3

A 72-year-old female with a history of hypertension, diabetes mellitus, and TA presented with speech impairment and left-sided weakness. Despite regular use of antihypertensive, antidiabetic, and azathioprine medications, the patient demonstrated dysarthric speech and weakness in the left extremities. Imaging revealed acute ischemic infarction in the right MCA territory on DWI, with no stenosis observed on cranial and carotid CT angiography. Routine laboratory tests were normal, and the patient was discharged on dual antiplatelet therapy.

DISCUSSION AND CONCLUSION

Takayasu arteritis (TA) is characterized by an inflammatory process affecting the aorta, its main branches, and pulmonary arteries, often resulting in arterial wall thickening, stenosis, occlusion, or dilation (1,2). These lesions frequently lead to organ dysfunction secondary to ischemia (5).

The clinical manifestations of TA vary throughout the course of the disease. In the early stages, predominant nonspecific systemic symptoms include headaches (31%), fever (29%), shortness of breath (23%), weight loss (22%), vomiting (20%), and musculoskeletal complaints such as myalgia, arthralgia, or arthritis (14%) (6). Later, symptoms vary based on the localization of the affected artery. Symptoms such as transient ischemic attack, dizziness, fainting, headache, and visual impairment may occur due to carotid and vertebral artery involvement (3,6). While stroke is a common complication of TA, it is rare for it to occur solely with acute ischemic stroke, as in our first case (6).

In 1990, the American College of Rheumatology established diagnostic criteria for TA, requiring the presence of 3 out of 6 criteria: being under 40 years of age, claudication of the extremities, decreased brachial artery pressure, blood pressure difference between the extremities greater than 10 mmHg, murmur on the subclavian arteries or aorta, and aortogram abnormalities (7). Our first newly diagnosed case met the criteria of being under 40 years of age, having decreased brachial artery pressure, and exhibiting a blood pressure difference between the extremities.

Vascular imaging serves as a primary marker for diagnosis and treatment. While digital subtraction angiography (DSA) is the gold standard diagnostic method, its invasiveness limits its utility (8). Magnetic resonance (MR) and computed tomography (CT) angiography typically show increased contrast uptake suggestive of inflammation, arterial wall thickening, and occlusion (8,9). Both of our cases exhibited similar findings of arterial wall thickening, narrowing, and occlusion.

Acute phase reactants or inflammatory markers are not reliable guides for diagnosis as their levels may rise early in the disease but return

to normal during its course (1,2). In our first case, normal levels of inflammatory markers did not exclude the diagnosis of TA, and these markers cannot be used to monitor treatment response.

Treatment options for TA include medical treatment, endovascular, or surgical intervention. In cases of active disease, high-dose prednisolone or equivalents are the first-line treatment options (10). Other options include methotrexate, azathioprine, cyclophosphamide, and anti-TNF agents. Antiaggregant should be initiated in cases of concomitant ischemia, but prophylactic use is not recommended.

In cases of stenotic or obstructive vascular lesions with hemodynamic significance, revascularization procedures are warranted. These procedures are ideally performed during periods of disease remission and in specialized centers equipped to manage the complexities of TA-related vascular pathology (10).

Percutaneous transluminal angioplasty (PTA) with balloon angioplasty or stenting is a preferred approach due to its minimally invasive nature. However, the risk of restenosis and stent thrombosis remains a concern, particularly in the setting of chronic inflammatory conditions like TA.

Surgical revascularization techniques, such as bypass grafting, may be necessary for complex or extensive lesions not amenable to endovascular intervention. Total CCA bypass, commonly utilized in cases of extensive carotid artery involvement, can effectively restore blood flow and alleviate symptoms.

The choice between surgical and endovascular approaches depends on various factors, including lesion characteristics, patient comorbidities, and institutional expertise. In cases where endovascular intervention poses a higher risk of complications, surgical revascularization may be the preferred option.

It is essential to carefully evaluate each patient's individual circumstances and tailor the revascularization strategy accordingly. Close multidisciplinary collaboration between rheumatologists, vascular surgeons, and interventional radiologists is paramount to ensure optimal outcomes in TA patients undergoing revascularization procedures (11,12).

The use of intravenous tissue plasminogen activator (IV-tPA) in TA patients with hyperacute ischemic stroke is limited. However, IV-tPA may contribute to functional improvement if administered within the first 4.5 hours of symptom onset, as seen in our first case (13,14).

Although thrombectomy for acutely developing large vessel occlusions in TA patients remains controversial due to the risk of re-occlusion and dissection, there have been cases showing significant neurological improvement after thrombectomy (15-17). In our first case, despite imaging showing proximal vessel occlusion, the patient was under 18 years of age, and thrombectomy was not performed due to the risk of dissection.

In addition to the direct effects of TA, the presence of comorbidities such as diabetes mellitus may precipitate stroke in elderly patients evaluated with a diagnosis of TA.

As a conclusion, early detection of Takayasu arteritis plays a crucial role in preventing the progression of vascular involvement and the development of severe clinical manifestations. By recognizing Takayasu arteritis as a potential cause of ischemic stroke, particularly in young female patients, clinicians can implement appropriate management strategies that may significantly influence the treatment outcomes and prognosis of the disease.

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Ethics

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