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Case Report / Olgu Sunumu

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ANCA-associated vasculitis with temporal artery involvement: A report of two cases

Temporal arter tutulumu olan ANCA ilişkili vaskülit: Olgu sunumu

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Abstract

Antineutrophil cytoplasmic antibodies (ANCA)-associated vasculitis and giant cell arteritis are two different vasculitides that differ in their clinical presentations, treatments and prognoses. Temporal artery involvement occurs rarely in ANCA-associated vasculitis. We present two patients with ANCA-associated vasculitis with temporal artery involvement.

Keywords: Temporal artery, vasculitis, anca-associated vasculitis

Öz

Antinötrofil sitoplazmik antikor (ANCA) ilişkili vaskülit ve dev hücreli arterit, klinik görünümleri, tedavileri ve prognozları açısından farklılık gösteren iki farklı vaskülittir. ANCA ilişkili vaskülitlerde temporal arter tutulumu nadiren görülür. Bu yazıda, temporal arter tutulumu olan iki ANCA ilişkili vaskülit hastasını sunacağız.

Anahtar Kelimeler: Temporal arter, vaskülit, anca ilişkili vaskülit

Introduction

Antineutrophil cytoplasmic antibodies associated (ANCA-associated) vasculitis consists of granulomatosis with polyangiitis, microscopic polyangiitis, and eosinophilic granulomatosis with polyangiitis and causes necrotizing pauci-immune inflammation in small to medium blood vessels.[1]

Giant cell arteritis is a vasculitis of medium to large blood vessels.[2] The involvement of the branches of the extracranial carotid artery leads to the classic symptoms.[2] Giant cell arteritis can also involve the aorta and its branches. Superficial temporal artery biopsy result is the gold standard for diagnosis.[2] Typically, a granulomatous reaction is seen with mononuclear infiltration along the elastic lamina.[3]

ANCA-associated vasculitis and giant cell arteritis are two different vasculitides that differ in their clinical presentations, treatments and prognoses. The prognosis of ANCA-associated vasculitis is worse and requires more intensive immunosuppression.[3,4] Temporal artery involvement in ANCA-associated vasculitis is rare.[5-7] However, its distinction from giant cell arteritis is crucial for effective therapy. Therefore, ANCA-associated vasculitis should be considered in patients with atypical presentations of giant cell arteritis. Here, we present two patients with ANCA-associated vasculitis with temporal artery involvement.

Case Reports

Case 1

A 73-year-old man was admitted with complaints of widespread pain. He had asthma and he complained of numbness of the feet and leg weakness. He also had jaw

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claudication. There was no tenderness or induration in the temporal region. The upper and lower extremity motor strength was 3/5. Laboratory testing showed C-reactive protein (CRP) 78 mg/L, erythrocyte sedimentation rate (ESR) 55 mm/h and eosinophils 25.6×10^3/L. The 24-hour urine protein was negative and there was no hematuria. Antinuclear antibodies indirect immunofluorescence assay (ANA-IFA) was negative, while p-ANCA was positive. Doppler ultrasonography and magnetic resonance angiography (MRA) of temporal arteries were normal. Thorax and abdomen computed tomography (CT) of thorax and abdomen were normal. Electromyography (EMG) revealed sensorimotor peripheral neuropathy. Temporal artery biopsy revealed giant cells in the degenerated internal elastic lamina (Figure 1). A diagnosis of ANCA-associated vasculitis with temporal artery involvement was made and a 3-g steroid pulse was given, followed by 1 mg/kg/day oral prednisolone and 2 mg/kg/day azathioprine. The patient informed consent was obtained.

Case 2

A 55-year-old woman was admitted with fatigue and widespread pain. She had numbness of her feet, headache, night sweats, fever, and weight loss. She also had jaw claudication. The physical examination was normal. There was no tenderness or induration in the temporal region. Laboratory testing showed CRP 250 mg/L and ESR 117 mm/h. The 24-hour urine protein was negative and there was no hematuria. ANA-IFA was negative, while c-ANCA was positive. Doppler ultrasonography and MRA of temporal arteries were normal. Thorax and abdomen CT of thorax and abdomen were normal. Temporal artery biopsy revealed intense inflammation in the media, and adventitia consisting of lymphocytic infiltration with segmental involvement (Figure 2). Diagnosis of ANCA-associated vasculitis with temporal artery involvement was made and a 3 g steroid pulse was given, followed by 1 mg/kg/day oral prednisolone and 2 mg/kg/day azathioprine. One month later, she presented with leg weakness. Her right foot dorsiflexion motor strength was 2/5. Hypesthesia of the right foot was also found. EMG revealed asymmetric sensorimotor neuropathy, consistent with mononeuritis multiplex and azathioprine was discontinued, and monthly intravenous cyclophosphamide was started. The patient informed consent was obtained.

Discussion

Vasculitides are subdivided according to their clinical characteristics and the size of the affected vessels. ANCA-associated vasculitis affects small to medium vessels, while giant cell arteritis affects medium to large vessels. Upper respiratory tract, lung, and renal involvement are more frequent in ANCA-associated vasculitis, while the carotid or aortic involvement is seen in giant cell arteritis. The treatment of the two vasculitides differs, and ANCA-associated vasculitis have a poorer prognosis.[2,4]

The first patient had widespread pain, fatigue, weakness, and numbness of the hands and feet. He had asthma and eosinophilia was detected. The p-ANCA, eosinophilia, and asthma were consistent with ANCA-associated vasculitis, as was the sensorimotor peripheral neuropathy. The jaw claudication and headache were thought to indicate the temporal artery involvement. Histologically, temporal artery inflammation was detected. ANCA-associated vasculitis with temporal artery involvement was diagnosed.

The second patient had headaches and jaw claudication, which were consistent with giant cell arteritis. However, the mononeuritis multiplex is not typical of giant cell arteritis.

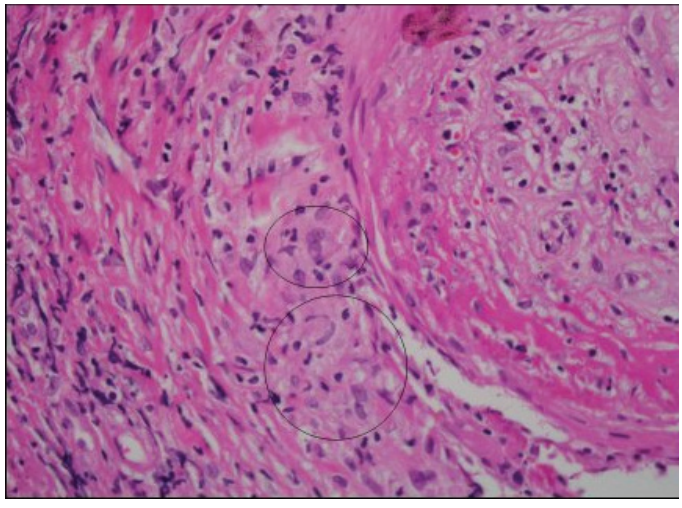

Figure 1. Histopathologic findings of the temporal arteries show giant cells at the degenerated internal elastic lamina with H&E

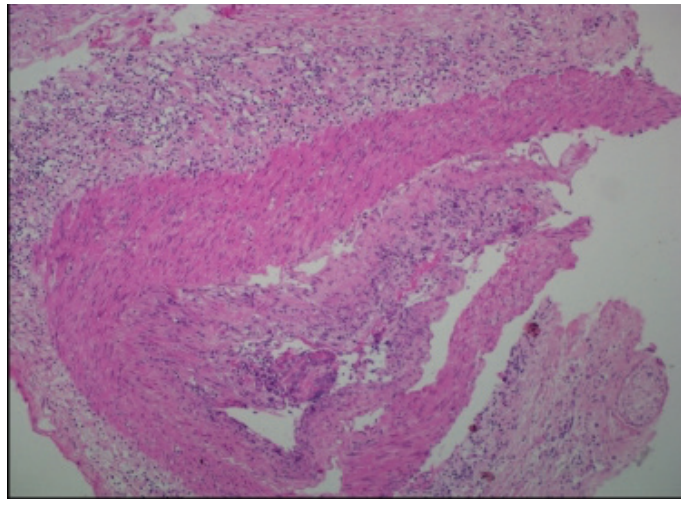

Figure 2. Histopathologic findings of the temporal artery show intimal thickening, lymphocytic infiltration and thrombosis with H&E

The c-ANCA positivity suggested a diagnosis of ANCA-associated vasculitis despite the absence of upper respiratory tract, lung, or renal involvement. Histologically, temporal artery inflammation was detected, but pathological findings were not typical of giant cell arteritis. A diagnosis of ANCA-associated vasculitis with giant cell artery involvement was made and the subsequent development of mononeuritis multiplex supported our diagnosis.

Endo et al.[7] reported a patient presenting with pain and tenderness in the temporal region in whom giant cell arteritis was diagnosed by biopsy. The clinical complaints were relieved with steroid treatment, but 9 months later the patient was admitted with bronchial asthma and peripheral eosinophilia. Although ANCA was negative, they suggested a diagnosis of eosinophilic granulomatosis with polyangiitis with temporal artery involvement. Tanaka et al.[6] reported a patient with microscopic polyangiitis with temporal artery involvement. Unlike our patient, they found necrotizing crescentic glomerulonephritis. In a cohort of 120 patients with giant cell arteritis, Hamidou et al.[8] detected different vasculitides in seven patients, including three patients with microscopic polyangiitis, two with polyarteritis nodosa, and one with granulomatosis with polyangiitis. Nishino reported that 5 of 347 granulomatosis with polyangiitis patients had temporal artery involvement, all of whom had headache and jaw claudication with loss of vision.[5]

ANCA-associated vasculitis and giant cell arteritis have different clinical presentations, treatments and prognoses. Therefore, it is important to make the correct diagnosis in a patient with temporal artery involvement. Our cases suggest that ANCA testing is beneficial in patients presenting with symptoms of giant cell arteritis but have unusual findings. ANCA positivity seems to be associated with involvement of the peripheral nervous system.

Ethics

Informed Consent: Informed consents were obtained from the patients.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Concept: Ö.V., B.G., Design: Ö.V., B.G., A.T., Data Collection or Processing: Ö.V., H.K., A.T., Analysis or Interpretation: B.Ö., A.D., Literature Search: Ö.V., Writing: Ö.V.

Conflict of Interest: No conflict of interest was declared by the authors.

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