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POSTER PRESENTATION

A Case of Giant Cell Arteritis Diagnosed Before 50 Years of Age

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Fever of unknown origin (FUO) refers to a prolonged febrile illness without an established etiology despite intensive evaluation and diagnostic testing. FUO may be caused by over 200 malignant/neoplastic, infectious, rheumatic/inflammatory, and miscellaneous disorders [1]. Systemic symptoms associated with giant cell arteritis (GCA) include fever, fatigue, and weight loss. Fevers occur in up to one-half of patients with GCA and are usually low-grade [2]. The most significant risk factor for developing GCA is aging. The disease rarely occurs before the age of 50 years, and its incidence rises steadily after that, peaking between the ages of 70 to 79 [3]. In this case report, we wanted to present a patient under the age of <50 years, diagnosed with giant cell arteritis while being investigated with a fever of unknown etiology.

CASE PRESENTATION

A 43-year-old female patient with no known internal disease was admitted to the rheumatology outpatient clinic with a fever that had increased in the afternoon for one year and lost 18 kilograms. At the same time, pain accompanied the fever, starting from the hip and spreading to the ankles. There was also headache that started from the neck and spread to the face, felt in the bilateral temporal region, and flashes of light in the eyes. Her rheumatological examination was unremarkable except for these complaints. Before the patient was admitted to our clinic, no infection or malignancy was detected in the external center investigation for the etiology of fever of unknown origin. On physical examination, fever was 37.2 °C, bilateral upper extremity blood pressure was 120/75 mmHg, bilateral temporal and carotid artery pulses were palpable, and other system examinations were normal. In the examinations performed when the patient applied to our clinic, PPD 17 mm, sedimentation 72 mm/hr, CRP 5.6 mg/dl, hemoglobin 9.5 g/dL, WBC 16.8 x10^3 µL, neutrophil 12.1 x10^3 µL, albumin 3.25 g/dL was detected. Temporal artery Doppler USG "left temporal artery diameter 1 mm, thickening of the temporal artery wall (halo sign), stenotic foci along the artery trace; right temporal artery diameter of 0.6 mm, stenotic foci along the artery tracing" were reported. Giant cell arteritis was diagnosed with the patient's current findings, and 48 mg of methylprednisolone (to be reduced by 4 mg per week), methotrexate six tablets per week, folbiol 1 per week, ASA 1x100 mg, and calcium-D vitamin were started. In the patient, who benefited from this treatment in a 3-month follow-up, a temporal artery biopsy was performed because they had a fever twice and the headache persisted even though it decreased. The biopsy result was "artery section with medial calcinosis."

Concurrent sedimentation and CRP with biopsy were respectively 45 mm/hr and 2.58 mg/dL. Vasculitis CT angiography was normal. In this period, methotrexate eight tablets/week and methylprednisolone 12mg were administered. PET CT was planned because she had a fever again in the follow-up. Due to diffusely increased FDG (SUVmax: 5.4) in the bone marrow in PET CT, she was referred to hematology. Bone marrow biopsy was performed. It was found normal. According to these results, tocilizumab was started once a week s.c. Methotrexate was adjusted five tablets/week, methylprednisolone 1x4 mg. The patient who responded to this treatment was followed up without complications and was fever-free.

DISCUSSION

Fever of unknown origin, defined as fever ≥38.3°C (101°F) for >3 weeks that remains undiagnosed after a hospital work-up. Fever of unknown origin work-ups may be done as an outpatient. FUO can be classified as infectious, malignant/neoplastic, rheumatic/inflammatory, and miscellaneous disorders. The most common causes of rheumatic diseases in the etiology of FUO are adult still's disease, juvenile rheumatoid arthritis (JRA), and giant cell arteritis (GCA). The fever-of-unknown-origin work-up should be a symptom (history) and sign (physical examination) driven. Based on history and physical clues, determine the appropriate category for the fever. Testing should be selective and based on diagnostic probabilities, not possibilities [1].

Giant cell arteritis (GCA, also known as Horton disease, cranial arteritis, and temporal arteritis) is categorized as a vasculitis of large- and medium-sized vessels because it can involve the aorta and great vessels. It causes stenosis and aneurysm of affected vessels. Fevers occur in up to one-half of patients with GCA and are usually low-grade. However, in approximately 15 percent of patients, fevers exceed 39ºC [2]. A characteristic laboratory abnormality in many patients with GCA is a high ESR, which can reach 100 mm/hour, and high CRP. Normochromic anemia is often present before therapy, moderately decreased serum albumin, also have a reactive thrombocytosis [4].

USG, MRI, CT, and PET CT can be used as imaging modalities for diagnosing GCA. Ultrasound sensitivity is 87%, and specificity is 96%. The temporal arteries are localized superficially, about 4 mm below the skin surface. Although they are small, with lumen and wall diameters of about 0.7 mm, they are easily accessible with ultrasound. Colour Doppler ultrasound of temporal arteries shows hypoechoic (dark) oedematous wall swelling in acute temporal arteritis. This tissue is not compressible. It disappears with glucocorticoid treatment after 2–3 weeks in most patients. In some patients, detecting a characteristic wall swelling becomes difficult after only three days of treatment [5].

KEY MESSAGES

In the etiology of fever of unknown origin, rheumatological diseases should always be included in the differential diagnosis.

Giant cell arteritis should be kept in mind in patients who present with fever, headache, and vision changes at a young age, even though it is a low probability.

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