

Sarcoid vasculitis: Case presentation

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Definition

Sarcoidosis is a multisystemic disease of unknown etiology that affects many organs and is characterized by non-caseating granulomas (1). Vascular involvement of sarcoidosis is an extremely rare finding. In the terminology of vasculitides published by the Chapel Hill Conference in 2012, vasculitis associated with systemic sarcoidosis was defined as sarcoid vasculitis (2).

Clinical Features

Sarcoid vasculitis is a very rare vasculitis, and vasculitides involving small, medium and large vessels can be seen. In the literature, sarcoidosis-related leukocytoclastic vasculitides, polyarteritis nodosa-like vasculitides involving medium vessels, and Takayasu-like large vessel vasculitides have been reported (3). It is well known that patients with pulmonary sarcoidosis have signs of vasculitis involving vessels of all diameters, from large vessels to venules in the lungs (4). However, extrapulmonary vasculitis is not an expected finding very often (1).

The clinic in sarcoid vasculitis varies according to the type of vessel involved. In patients with small vessel involvement, skin involvement in the form of palpable purpura is prominent, while symptoms similar to Takayasu's arteritis may be seen in patients with large vessel involvement (such as limb claudication, dizziness, blood pressure difference, and murmur on the vessel).

Diagnosis

Radiological and histopathological findings in sarcoid vasculitis patients vary according to the type of vessel involved. Imaging is an important tool in diagnosis when large vessel involvement is present, and CT-angio, MR-angio or PET-CT can be used for this purpose. In patients with small vessel involvement, histopathological examination is essential, and biopsy is required from the affected area in suitable patients.

Case Report

A 52-year-old female patient applied to our clinic with complaints of pain in both arms and fatigue for 10 years. The patient had diagnoses of diabetes mellitus, hypertension, and sarcoidosis and smoked 60 packs/per year. The patient had been stented 5 years ago in the right brachiocephalic artery and right common carotid artery, and balloon angioplasty was performed on the right subclavian artery. Coronary angiography was performed 3 times in 5 years, and a stent was placed in the left main coronary artery, circumflex artery, and left anterior descending artery. A year ago, the patient was evaluated in the chest diseases outpatient clinic with dyspnea, cough, and weight loss. Thorax computer tomography (CT) revealed lymph nodes that increased in size in the right paratracheal, subcarinal, and right hilar regions. Fine-needle aspiration biopsy taken by endobronchial ultrasonography from the lymph

node, the largest of which is 20 mm in the right lower paratracheal area, resulted in granulomatous lymphadenitis. The patient was diagnosed with sarcoidosis and was followed up with symptomatic treatment. Four months after the diagnosis of sarcoidosis, echocardiography performed due to newly developed hypertension revealed a 50% ejection fraction and mild hypokinesia in the posterior apicoseptum. Coronary angiography was performed again in the patient, and 60-70% stenosis was detected in the stent in LAD and CX, but recanalization could not be achieved. The patient who applied to our clinic due to recurrent arterial thrombosis had normocytic anemia (Hb: 9 gr/dl, MCV: 85 fl) in her laboratory. Sedimentation and CRP were in the normal range; there was no hypercalcemia. Her ACE level was high (119 U/L), she did not have hyperlipidemia. The patient's previous CRP values were between 13 and 57 mg/L. In CT angiography, a stent was observed in the right common carotid artery and 85-90% stenosis was observed in the stent, fibrocalcific plaques causing approximately 75-80% field stenosis in the right carotid bulb, fibrocalcific plaques causing 75-80% field stenosis in the left carotid bulb. The stent lumen was open in the right brachiocephalic artery, and there were plaques in the proximal part of the right subclavian artery, causing approximately 80-85% stenosis. No stenosis was detected in the pulmonary artery branches and abdominal aorta branches of the patient. In PET CT, there were multiple lymph nodes with the largest 2,5 cm in the right supraclavicular, inferior jugular, along the right paratracheal chain, subcarinal area, bilateral hilar region (SUV max 12.6), no sign of large vessel vasculitis was detected.

Differential diagnosis

1. Giant cell arteritis
2. Atherosclerotic disease
3. Sarcoid vasculitis
4. Poliarteritis nodosa

In our case, existing vascular involvements were associated with sarcoid vasculitis, since the patient with a known diagnosis of sarcoidosis had recurrent medium-large vessel thrombosis and acute phase elevation accompanying these vascular events, the patient did not have hyperlipidemia, and there was no family history of cardiovascular disease at an early age. The large vessel involvement of the patient was thought to be related to sarcoidosis and a diagnosis of sarcoid vasculitis was made. Methotrexate 15 mg/week and prednisolone 10 mg/day were started.

Key messages

- Sarcoid vasculitis is an extremely rare disease.
- In sarcoid vasculitis, vessel involvement of any diameter can be seen.
- Before making a diagnosis of sarcoid vasculitis, other possible causes of vasculitis must be excluded. If possible, histopathological examination gives extremely valuable information in terms of diagnosis.
- In the treatment, immunosuppressive treatments are used together with steroids.

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