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CASE REPORT – OPEN ACCESS

Coexistence of Behçet Disease and Sarcoidosis in a Young Man with Femoral Artery Aneurysm: An Uncommon Case

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ABSTRACT

Background: Behçet disease may be confused with sarcoidosis because they are both multisystem inflammatory disorders presenting with uveitis, polyarthritis, cardiovascular disease, erythema nodosum, and other cutaneous lesions. The coexistence of Behçet disease and sarcoidosis is extremely rare but not impossible. Vascular involvement is a significant pathological finding in Behçet disease, and arterial aneurysm formation is more common than occlusion. However, large-vessel vasculitis associated with sarcoidosis is extremely rare.

Case Report: A 23-year-old man was admitted to the hospital because of swelling and pain in the right groin. The patient, who had predominant Behçet disease, was also diagnosed with sarcoidosis during the examination. He had inguinal distention associated with femoral artery aneurysm and underwent successful surgery without complication.

Conclusion: Assessment of the coexistence of Behçet disease and sarcoidosis is critical because of their potentially fatal effects by triggering systemic inflammation. Furthermore, arterial diseases, especially those developing at young ages, can be a sign of immunoinflammatory disease.

Keywords: Behçet disease, sarcoidosis, vasculitis, femoral artery aneurysm, vascular surgery

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INTRODUCTION

Femoral artery aneurysms are advanced-age diseases; however, they can develop in young patients with noninfectious autoimmune or inflammatory arteritis and connective tissue disorders. Behçet disease (BD) and sarcoidosis are well-known multisystemic inflammatory diseases that can affect the cardiovascular system. Although they overlap in terms of clinical features, the presence of both BD and sarcoidosis in a patient is scarce (1). Vasculitis is the main pathological finding in BD and can affect vessels of all sizes. However, information on vascular involvement in sarcoidosis is unclear. Small- and medium-sized vessels are frequently involved in pulmonary sarcoidosis, but extrapulmonary vascular involvement is uncommon (2).

We report the case of a young patient who was admitted to the hospital with groin pain and diagnosed with BD, sarcoidosis, and femoral artery aneurysm.

CASE REPORT

A 23-year-old Turkish man was admitted to the emergency room with right groin pain. He had no previous medical history and did not smoke and use illicit drugs. On physical examination, a pulsative mass in the right groin and venous collaterals on the right side of the abdomen were noted and the thrombus was detected in the distal part of the vena cava inferior on computed tomography (Fig. 1a, b). A review of his clinical symptoms showed that the patient had a history of recurrent oral aphthae (five to six times a year). The patient has never visited any hospital for treatment of oral aphthae and venous collaterals because he thought these were varicose veins. A dermatologist and ophthalmologist also performed clinical evaluation of the patient. Ocular manifestations (such as uveitis, episcleritis, scleritis, conjunctival ulcers, and keratitis) and skin manifestations (such as genital aphthous ulcers, erythema nodosum, pseudofolliculitis, and acneiform nodules) were not detected. Still, the skin pathergy reaction for BD was found positive. The results of the initial laboratory investigations showed that the following were within the normal range: complete blood cell count, electrolyte and calcium, renal function indices, liver enzymes, acute phase reactants (erythrocyte sedimentation rate and C-reactive protein), fibrinogen, and D-dimer. In the tests performed for cancer screening, prostate-specific antigen, alpha-fetoprotein, and carcinoembryonic antigen levels were within normal limits, and fecal occult blood screening was negative. Thrombophilia screening showed normal protein C and S, homocysteine, and antithrombin III levels. The patient did not have mutations in methylenetetrahydrofolate reductase, factor V

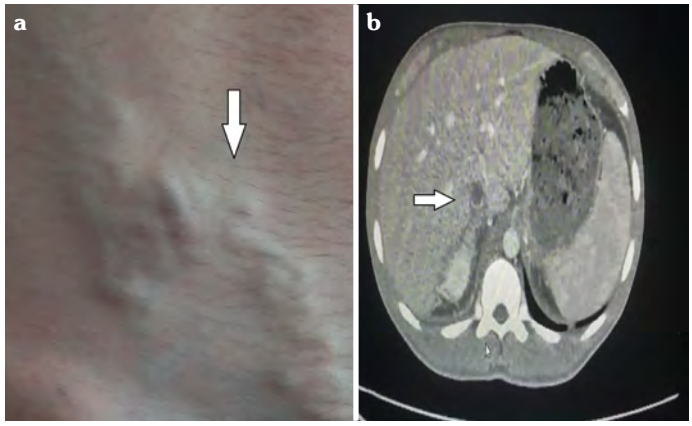

Figure 1. (a) Venous collaterals on the right side of the abdomen. (b) Image of thrombus in the vena cava inferior

and prothrombin, lupus anticoagulant, and anticardiolipin antibody. An HLA B27 test showed negative results, whereas HLA B51 was found to be positive. Antinuclear antibody (ANA) determined using an indirect immunofluorescence method was mildly positive with 1/80 titration, and the serum angiotensin-converting enzyme (ACE) level was 65 U/L (normal range, 8–52 U/L). Laboratory values obtained during follow-up in the hospital are shown in Table 1.

The patient underwent peripheral computed tomography angiography for the pulsatile mass in the right groin, which showed a femoral artery aneurysm and intraoperative view of the aneurysm shown in Figure 2b (Fig. 2a and b). In addition, thorax computed tomography (TCT) was performed to detect the presence of aneurysms in other anatomical regions. TCT and chest radiographic images led us to suspect sarcoidosis and sarcoidosis diagnosis confirmed by transbronchial lung biopsy (Fig. 3a–c). TCT scans showed mediastinal lymphadenopathy and disseminated parenchymal infiltration of typical for sarcoidosis. In late-phase tomography, the thrombus was detected in the distal part of the vena cava inferior (Fig. 1b). Moreover, it did not show pulmonary embolism, and transthoracic echocardiography showed normal left and right ventricle functions and pulmonary artery pressure. The presence of active infection was not considered in the patient because there was no fever and infection parameters were within normal limits.

The femoral artery graft interposition was performed following the excision of the artery aneurysm using a 10-mm biosynthetic vascular graft (Omniflow II, LeMaitre Vascular, USA) (Fig. 2b). The patient was treated with low-molecular-weight heparin (heparin sodium 7500 IU daily) for 10 days. After the 10th day, acetylsalicylic acid (100 mg/day) and warfarin (with INR values of 2.5) was administered instead of heparin. Moreover, the rheumatology department suggested 1 mg/kg/day of oral prednisolone with gradual tapering and colchicine (0.5 mg twice daily). He was referred to the pulmonary disease department when his general condition improved. Flexible bronchoscopy was performed, and transbronchial lung biopsy revealed non-caseating granulomas consistent with sarcoidosis (Fig. 3c). The patient underwent follow-up with the rheumatologist, pulmonologist, and cardiovascular surgeon.

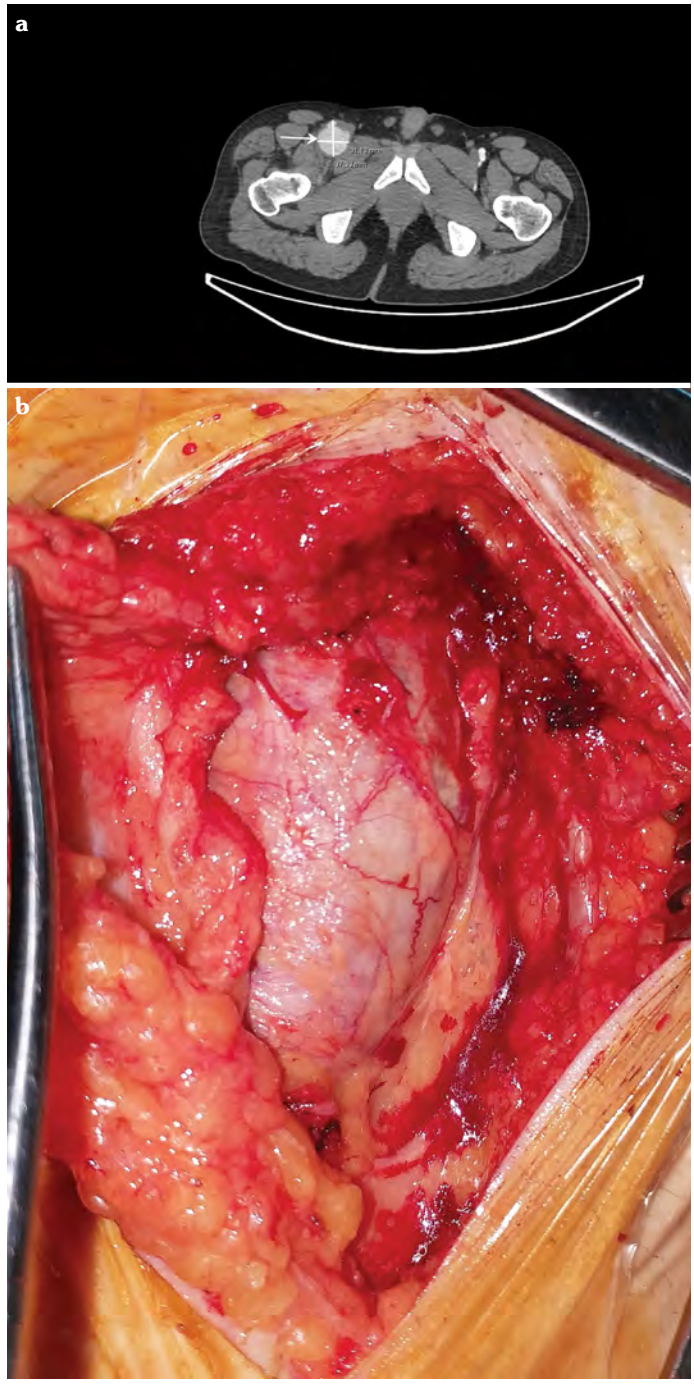

Figure 2. (a) Image of femoral aneurysm on peripheral computed tomography angiography indicated by white arrow. (b) Image of intraoperative femoral artery aneurysm

DISCUSSION

BD and sarcoidosis are chronic multisystemic inflammatory disorders that usually present with uveitis, polyarthritis, cardiovascular disease, erythema nodosum, and cutaneous lesions in chest imaging methods. The general prevalence of BD is 350 and 6000 per million of the population, whereas the prevalence of sarcoidosis is 2.2 to 160 cases per 100,000 people. However, reports on incidence and prevalence vary in relation to factors such as geography, gender, age, and ethnicity. Although

Table 1. Demographic/clinical data, and laboratory findings

	Patient 1	Reference value
Demographic characteristics		
Sex	M	
Age (years)	23	
BMI (kg/ml)	24	
Smoking	No	
EKG	Sinus rhythm	
Laboratory findings		
White blood cell (10^3/uL)	7540	3.7–10.1
Hemoglobin (g/dL)	15.1	13–18.1
Platelet (10^3/uL)	283	3900–10800
D-dimer (mg/L)	0.1	0–0.55
Fibrinogen (mg/dL)	306	245–400
Troponin-I (pg/ml)	0.3	<34.2
BUN (mg/dL)	14.8	8.4–25.7
Creatinine (mg/dL)	0.8	0.7–1.2
AST (U/L)	34	10–40
ALT (U/L)	21	0–55
Albumin (g/dL)	4.38	3.5–5.0
INR	1.02	0.8–1.2
PT (s)	12.1	10.5–14.9
aPTT (s)	32.9	21–35
CRP (mg/L)	3.95	0–5
ESR (mm/h)	18	0–20
ACE (U/L)	65	8–52
Factor V (%)	85	70–120
Protein C (%)	103	70–130
Protein S (%)	116	65–140
Antithrombin III activity	101	80–120
Rheumatoid factor (IU/mL)	<9.6	0–16
Antinuclear antibody	+1 (1/80 titration)	
Anticardiolipin antibody (GPL-U)		
IgG	1.9	0–10
IgM	0.4	0–7
MTHFR	Normal	
Factor V leiden	Normal	
HLA B27	Negative	
HLA B51	Positive	

BMI: Body mass index; LDH: Lactate dehydrogenase; AST: Aspartate transaminase; ALT: Alanine aminotransferase; INR: International normalized ratio; PT: Prothrombin time; aPTT: Activated partial thromboplastin time; CRP: C-reactive protein; ESR: Erythrocyte sedimentation rate; ACE: Angiotensin-converting enzyme; MTHFR: Methylenetetrahydrofolate reductase

the incidence of BD and sarcoidosis coexistence is not known precisely, patients presenting with both BD and sarcoidosis have been described in the literature.

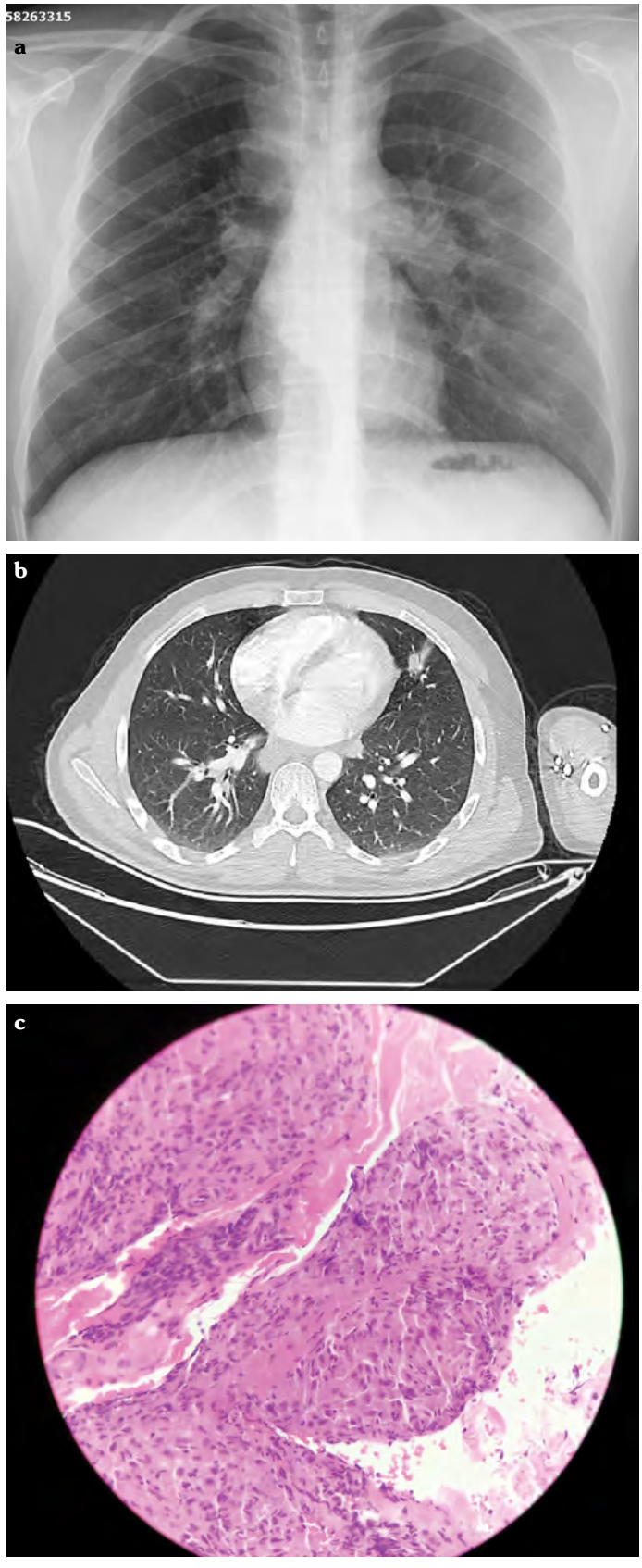

Figure 3. (a) Chest X-ray image of the patient. (b)TCT scans showed mediastinal lymphadenopathy and parenchymal infiltration. (c) Transbronchial noncaseating granuloma on histopathological image

Femoral artery aneurysms are advanced-age diseases and are have the same risk factors as other aneurysms (3). The present case was a young man with no risk factors such a smoking, hypertension, or diabetes mellitus. The anamnesis revealed no history of trauma, invasive procedures, or drug addiction. First, we believed that the patient might have BD because the patient has a history of oral aphthae occurring five to six times a year. Therefore, BD should be considered in young patients with an artery aneurysm without risk factors (4). Vascular involvement is joint in BD since this disease is a chronic, autoimmune systemic vasculitis. Deaths related to the aneurysm and pseudo-aneurysm rupture have a significant rate in BD (5). Because of its anatomical location and similar features to lymph node and inguinal hernia, diagnosis of femoral aneurysm may be delayed until complications develop. Our patient presented with inguinal pain and enlargement in the groin. The physical examination performed by a careful emergency room specialist showed a pulsatile mass.

Another interesting issue in our patient was venous collateral in the abdominal region. Interestingly, the patient thought these collaterals were varicose veins and did not present to any hospital. A pathology involving the inferior vena cava was supportive of the venous system effect of BD. We confirmed the diagnosis of BD with positive HLA B51 and pathergy tests. These findings were consistent with those carried out by Berriche et al. (6).

The coexistence of BD and sarcoidosis is extremely rare but not impossible (1). Furthermore, it is clinically challenging to differentiate sarcoidosis from BD. Sarcoidosis may be asymptomatic or can present several symptoms (7). According to Baughman et al. (8), the diagnosis of sarcoidosis is based on observation, knowledge, and the clinician's experience. Our patient had no symptoms, such as dry cough, shortness of breath, chest pain, and rarely low-grade fever suggestive of sarcoidosis, and if we had not investigated whether there was an aneurysm due to BD elsewhere, we would not have found suspicious findings in TCT. Therefore, we evaluated some laboratory findings in our patient. ACE and ANA levels were in the upper limit of normal. The absence of symptoms related to sarcoidosis and laboratory values showed that BD had a predominant clinical picture in our patient.

BD and sarcoidosis are multisystem inflammatory disorders and can lead to cardiovascular system complications. Although the vascular effects of BD are clearly defined, sarcoidosis also has vascular effects. Systemic vascular involvement in sarcoidosis includes granulomatous angiitis and microangiopathy (9). A study conducted in 2011 reported that patients with sarcoidosis have impaired endothelial function and increased arterial stiffness (10). In light of this information, it is not surprising that arterial pathology (e.g., aneurysm and occlusion) and venous thrombosis develop in patients with sarcoidosis. In our patient, the presence of arterial and venous involvement and positive pathergy test and HLA B1 suggested that femoral aneurysm developed owing to BD.

This case showed that chronic inflammatory diseases could progress with atypical and unpredictable clinical signs.

CONCLUSION

BD and sarcoidosis are chronic, complex, and immunoinflammatory disorders, where the cardiovascular effects of BD have overshadowed those of sarcoidosis. Therefore, further studies are needed. In addition, chronic inflammatory diseases should be considered in patients presenting with atypical symptoms. Therefore, clinicians should be aware of vascular involvement in immunoinflammatory diseases.

Informed Consent: Written informed consent was obtained from patients who participated in this study.

Peer-review: Externally peer-reviewed.

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Conflict of Interest: The authors have no conflict of interest to declare.

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