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Review

Imaging manifestations of Behcet's disease: Key considerations and major features

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ARTICLE INFO

Abbreviations:
BD
Behcet's disease
PTE
pulmonary thromboembolism
SVC
superior vena cava
BCS
Budd-Chiari syndrome
CNS
central nervous system
MeSH
Medical Subject Heading
DVT
deep vein thrombosis
CT
computed tomography
MR
magnetic resonance
CXR
chest x-ray
MRI
MR imaging
ECG
electrocardiography
MIP
maximum intensity projections
TTE
transthoracic echocardiography
TEE
transesophageal echocardiography
HASTE
half-fourier acquisition single-shot turbo spin-echo
IVC
inferior vena cava
ADC
apparent diffusion coefficient

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ABSTRACT

Behcet's disease is an autoimmune disease most commonly seen in the Middle East. Although primarily known with painful oral and genital ulcers, it can lead to vasculitis. Therefore, several associated complications such as thrombotic syndromes, aneurysmal arterial disease may arise. In many cases, it might be difficult to make the diagnosis purely based on clinical grounds; however, imaging plays an important role for both diagnosis and assessment of the disease's complications. We provide a comprehensive review of the most notable imaging findings of Behcet's disease.

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Autoimmune disease
Vasculitis
Thrombosis

1. Introduction

Behcet's disease (BD) is an autoimmune disease most commonly seen during the second and third decades of life in people living in countries around the historical Silk Road. The prevalence is reported to be up to 370 patients per 100,000 persons in Turkey and 40 per 100,000 persons in Iran [1]. However, the epidemiology goes beyond, considering the dynamic shift in the populations, including immigration. In fact, BD has been reported everywhere around the world, with prevalence estimates around 4.2 per 100,000 people in Germany, 7.2 per 100,000 persons in France, and 8.6 per 100,000 persons in the United States [1]. BD is primarily known with painful oral and genital aphthous ulcers and uveitis [2] and is a unique vasculitis that is not confined to a specific vessel size [3]. BD could manifest with serious complications, such as blindness and visual impairment, thrombotic complications such as pulmonary thromboembolism (PTE), arterial aneurysms, and central nervous system (CNS) involvement, among others; causing severe functional disabilities and death [1]. The diagnosis is usually made clinically by the International Study Group criteria or the International Criteria for Behcet's Disease; however, in many cases it is challenging to make a definite diagnosis with the current criteria. In other cases, the disease might not even be suspected prior to review of specific imaging features, especially in non-endemic areas [4–7]. Moreover, imaging findings can have a key role in prognostication, and response to treatment for patients with BD [8]. Imaging plays an important role in assessing the disease involving different organ systems. As such, collaborative communication between radiologists and clinicians and careful review of the imaging findings is key in comprehensive assessment of patients with suspected or proven BD. We provide a contemporary summary of the most notable extraocular manifestations of BD, focusing on the key imaging findings that radiologists and clinicians should be aware of.

1.1. Data source

We searched PubMed for studies related to the imaging manifestations of BD, published in English from January 1, 1960 to April 24, 2017, using a combination of keywords and Medical Subject Heading (MeSH) terms (Table 1). We identified 240 citations and screened all of them; as well as the reference list of related articles. Where possible; we tried to use the data from large cohort studies and systematic reviews or large case series. Inevitably; however; at times use of data from different regions with variable baseline prevalence and/or dissimilar screening or diagnostic techniques; leading into variations in the reported relative frequency of organ system involvements or specific imaging manifestations. Further; at times; for rare manifestations; and more recent imaging modalities case reports and smaller case series are used to describe the imaging manifestations (Table 2).

2. Cardiovascular involvement

Cardiovascular involvement in BD is reported in about 9% of patients [9], although there might be variations in other reports. Cardiovascular complications may have life-threatening consequences, sometimes constitute the first presentation of the disease and associated with worse prognosis [10,11]. Vascular involvement includes deep vein thrombosis (DVT; the most common complication), PTE, vascular aneurysm formation, and thrombotic occlusion (venous and/or arterial) [12]. Cardiac manifestations include intracardiac thrombosis, sinus of Valsalva and coronary aneurysm, pericarditis, myocarditis, endocarditis, endomyocardial fibrosis, valvular dysfunction and conduction system disorders [11,13].

2.1. Aortic vasculitis

Aortic vasculitis could be seen in the course of BD, and is characterized by the presence of leukocytes within the aortic wall, with reactive damage to mural structures [14]. Clinical presentations are nonspecific and may include fever, chest or abdominal pain, and vascular insufficiency [15]. Computed tomography (CT) and magnetic resonance imaging (MRI), in early stages, can show irregular wall thickening, with fat stranding and, mural contrast enhancement, while stenosis, occlusion, and aneurysm formation are seen in later stages (Fig. 1) [14–16]. Delayed contrast enhancement has been described as a feature of active disease [14].

2.2. Venous thromboembolism and superior vena cava syndrome

DVT is a common vascular complication in the course of BD. In large studies, DVT has been reported in 4–6.6% of patients with BD [6,17]. Asymptomatic lower extremity DVT is the most common presentation, frequently occurring in the popliteal veins with male predominance [18], although symptomatic presentation, has been reported as well [19].

Ultrasonography is an accurate noninvasive method for the initial assessment of lower extremity DVT, especially proximal DVT. Assessment of the distal veins, and the upper extremities may be warranted in the right clinical scenarios [20]. Interestingly, embolism is rare in patients with BD who develop DVT, due to the adherent nature of the thrombi in the inflamed veins. PTE in the course of BD may stem from in-situ pulmonary thrombosis and distinguishing such events from embolic phenomena is challenging [12,16]. In the case of PTE, chest x-ray (CXR) is similar to PTEs in other conditions. CT pulmonary angiography is used for making the diagnosis, and echocardiography can be used to identify thrombus in-transit, or to determine the right ventricular function [21]. On CT, pleural-based wedge-shaped opacities might be considered as foci of pulmonary infarction [22]. CT can also show mosaic attenuation in lung parenchyma or dark regions with smaller vessels compared with nearby normal parenchyma and vasculature, and occasionally vessel wall calcification in the case of chronic thromboembolic pulmonary hypertension [8]. Ventilation-perfusion scan may show segmental perfusion defect with normal ventilation [23]. Considering the young age of patients with BD, magnetic resonance imaging (MRI), including MR angiography may be of value in the diagnosis of complications such as PTE, to minimize the exposure to ionizing radiation, especially among young population [21,24].

In BD, Superior Vena Cava (SVC) syndrome has been reported due to vasculitis, thrombosis, or extrinsic compression as a result of mediastinal fibrosis [25,26]. Based on the acuity and extent of SVC obstruction, the presentation varies from flushing and dyspnea to cyanosis, respiratory distress, and altered mental status [27]. Chylothorax and chylopericardium may also complicate the course, possibly due to

Table 1
Search Strategy.

(Behcet*[TI] OR "Behcet Syndrome"[MAJR] OR "Behçet"[TIAB]) AND (imag*[TI] OR radiolog*[TI] OR radiograph*[TI] OR tomograph*[TI] OR ultrasonogra*[TI] OR sonogra*[TI] OR magnetic*[TI] OR radionuc*[Ti] OR Echo*[TI] OR angiography[TI] OR Venography[TI])