Thoracic manifestations of Behcet's disease by MSCT and CT angiography

Thesis
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Abstract

Behçet's disease (BD) is a multisystem inflammatory disorder classified among the vasculitides, which can affect all types and sizes of blood vessels characterized by major symptoms of oral aphthous ulcers, uveitis, skin lesions and genital ulcers. Involvement of intestines, vessels and central nervous system (CNS) sometimes leads to a poor prognosis.

Although cases of Behçet's disease were reported from all around the world, it is more prevalent in Far East (Japan, Korea); Middle East (Iran, Iraq, Israel, Saudi Arabia, Kuwait, Syria) and countries around Mediterranean see (Turkey, Italy, Egypt, Greece, Morocco, Algeria, Tunis). Therefore, Behçet's disease occurs most commonly in the countries along the ancient “silk road” Clinical Diagnosis of Behçet Disease:According to the diagnostic criteria of the International Study Group for Behçet Disease , the diagnosis is based on the presence of recurrent oral ulcerations, along with two of the following criteria: (a) recurrent genital ulcerations, (b) eye lesions, including uveitis and retinal vasculitis, (c) skin lesions (folliculitis, erythema nodosum), and (d) positive skin pathergy test (pustule formation 24–48 hours following skin prick) .

Behçet disease involving the chest can manifest as a wide spectrum of abnormalities. Aneurysms of the pulmonary arteries with or without thrombosis are a typical manifestation of Behçet disease. Involvement of the SVC and aorta may occur, and pulmonary findings include pulmonary hemorrhage and atelectasis, fibrosis, and air trapping. The mediastinum and pleura may also be involved. Knowledge of these various manifestations can be useful in diagnosing Behçet disease, documenting the cause of symptoms in patients who present with hemoptysis, and initiating appropriate therapy.

Spiral CT is useful in demonstrating the entire spectrum of thoracic manifestations of Behçet disease. Spiral CT is noninvasive and provides excellent delineation of the vessel lumen and wall and perivascular tissues as well as detailed information concerning the lung parenchyma, pleura, and mediastinal structures. CT angiography can be performed with only a small quantity of contrast material and may be used as an alternative to venography and angiography.

Conventional chest radiography is commonly used for initial assessment of pulmonary signs and symptoms of Behçet disease, for follow-up, and for assessment of response to therapy. The pulmonary parenchymal changes are nonspecific and appear as focal and diffuse areas of increased opacity . Chest radiographs are also useful for detection of hilar enlargement, which may be due to pulmonary artery aneurysms , and of mediastinal widening, which may be an ominous sign of an aneurysm developing in the thoracic aorta.

In our study, we are aiming to evaluate those patients diagnosed as Behcet disease whether symptomatic or asymptomatic for early detection of chest manifestations.

Keyword

CNS-BD-CT- MSCT- *PAA*

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LIST OF ABBREVIATIONS

3D	*Three dimensional*
BD	*Behcet's disease*
BS	*Behcet's syndrome*
CT	*Computed Tomography*
CTA	*CT angiography*
CTPA	*CT pulmonary angiography*
CTV	*CT venography*
DVT	*Deep venous thrombosis*
FOV	*Field of view*
IVC	*Inferior vena cava .*
MDCT	*Multi-detector computed tomography*
MSCT	*Multi-slice computed tomography*
MIP	*Maximal intensity projection*
MPR	*Multi-planner reconstruction*
MRA	*Magnetic resonance angiography*
NBD	*Neuro-Behcet's disease*
PAA	*Pulmonary artery aneurysm*
SVC	*Superior vena cava.*
VR	*Volume rendering*

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INTRODUCTION

INTRODUCTION

Behçet disease is a rare multisystemic and chronic inflammatory disorder with an unknown cause. Genetic predisposition (human leukocyte antigen [HLA], HLA-B51) and certain microorganisms are believed to play roles in its cause. It is most frequently seen in the third decade of life and is more frequent in men. The course of the disease is more severe in men than women and in those younger than 25 years at disease onset. Behçet disease is most frequently seen in Mediterranean, Middle East, and Far East countries. The highest prevalence rate, to our knowledge, was reported from Turkey as 80–370 per 100,000 **(Ceylan et al, 2010).**

Recurrent oral and genital ulcers and uveitis are the clinical triad of the disease. According to the diagnostic criteria of the International Study Group for Behçet's Disease **(1990)**, the diagnosis is made on the detection of oral ulcers and at least two of the following criteria: recurrent genital ulcers, ocular lesions including uveitis and retinal vasculitis, skin lesions (folliculitis, erythema nodosum), and positive skin pathergy test (i.e., pustule formation 24–48 hours after skin prick) **(Ceylan et al, 2010).**

. Behçet’s disease involving the chest can manifest as a wide spectrum of abnormalities. Aneurysms of the pulmonary arteries, with or without thrombosis, are a typical manifestation of Behçet’s disease **(Hiller et al, 2004)** .

Pulmonary involvement is relatively infrequent, having been reported in 1%–10% of patients **(Erkan, et al, 2001).**

computed tomography (CT) is the preferred imaging modality for depicting the thoracic manifestations of Behçet disease **(Hiller et al, 2004)** .

The evident advantages of CT for the diagnosis of pulmonary vascular problems including pulmonary aneurysms and pulmonary embolism have become further enhanced by the introduction of multislice CT technology. It is now feasible to acquire a 1-mm scan of the entire thorax within one breath-hold **(Schoepf et al, 2001)**.

Behçet disease may also affect joints, the gastrointestinal system, CNS, cardiovascular system, and the lungs. Vascular system involvement is seen in approximately 25–30% of patients and is the most common cause of mortality. The main pathologic process in Behçet disease is vasculitis and perivascular infiltration affecting vessels of various sizes **(Ceylan et al, 2010).**

In this literature, we review , discuss and illustrate the spectrum of CT findings in Behçet disease of the thorax, including vascular, pulmonary parenchymal, pleural, and mediastinal abnormalities.

Aim of work

The purpose of this study is to evaluate the radiological findings related to thoracic vascular system involvement and pulmonary, pleural, and mediastinal involvement of the patients diagnosed as Behcet's disease (BD) according to the diagnostic criteria of the International Study Group for Behçet's Disease . CT chest with contrast and CT pulmonary angiography are important diagnostic imaging techniques in the evaluation of patients with Behçet disease.

Review of literature

Anatomy