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Behçet's disease: Spectrum of MDCT chest and pulmonary angiography findings in patients with chest complaints



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ABSTRACT

Objective: The aim of the work was directed to evaluate the value of multi-detector computed tomography pulmonary angiography study in evaluation of known patients with Behçet's disease.

Materials and methods: This study was done retrospectively and included eighteen known patients with Behçet's disease and referred for MDCT pulmonary angiography.

Results: Pulmonary artery aneurysm was the most common finding as it was found in 16 patients, followed by pulmonary embolism which was found in 14 patients, 12 patients with pulmonary hypertension, right ventricular strain in 6 patients, intracardiac thrombus in 4 patients, dilated bronchial arteries in 8 patients, venous occlusion in 4 patients, mosaic attenuation of the lung in 12 patients, pulmonary infarcts in 4 patients, and pleural effusion in 4 patients.

Conclusion: MDCT pulmonary angiography is an important diagnostic imaging tool for diagnosis of vascular complications in patients with Behçet's disease.

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1. Introduction

Behçet's disease (BD) is a chronic, relapsing, and debilitating systemic vasculitis of unknown etiology with diverse spectrum of clinical manifestations including mucocutaneous lesions, ocular, vascular, articular, neuro-

logic, gastrointestinal, urogenital, and pulmonary involvement [1–3].

Onset of BD usually occurs in the third decade of life with an equal sex predilection in most patient series; however, its course in males is more severe than in females [4].

BD results in vasculitis and the major manifestations are venous and arterial thrombosis and arterial aneurysms, but venous involvement is typically more evident. Arterial occlusions and aneurysms sometimes coexist, as well as arterial lesions and venous thrombosis that may affect the same patients. After a first event, other vascular lesions tend to occur [1,5–7].

Cardiovascular involvement, an important and serious manifestation occurs in 7–46% of patients. BS is a distinctive perivasculitis that may involve both veins and arteries

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Table 1

Clinical data of the studied cases.

Clinical findings	Number of patients
Oral ulcer	9
Genital ulcer	9
Ocular lesions	6
Skin lesions	8
CNS	1
GIT	5
Joint affection	7
Pathergy test	4

Table 2

MDCT angiographic findings of the studied cases.

CT findings	Number of patients
Pulmonary hypertension	12
Pulmonary artery aneurysm	16
Pulmonary embolism	14
Right ventricular strain	6
Intracardiac thrombus	4
Pleural effusion	4
Pulmonary infarcts	4
Mosaic attenuation	12
Venous occlusion	4
Dilated bronchial arteries	8

of all sizes, ranging from great blood vessels to capillaries [8].

The pulmonary artery is the second most commonly affected artery after the aorta; it may show single or multiple aneurysmal dilatations. The arterial aneurysmal dilatation is more common than occlusion and this is more dangerous due to complications of rupture [2,9].

Vascular manifestations are the main predictors of mortality and morbidity in Behcet's disease. Arterial complications may lead to difficult surgical problems. Any invasive methods to arterial system may cause pseudoaneurysms. Repairs by using autogenous veins or synthetic grafts may lead to anastomotic false aneurysms [10].

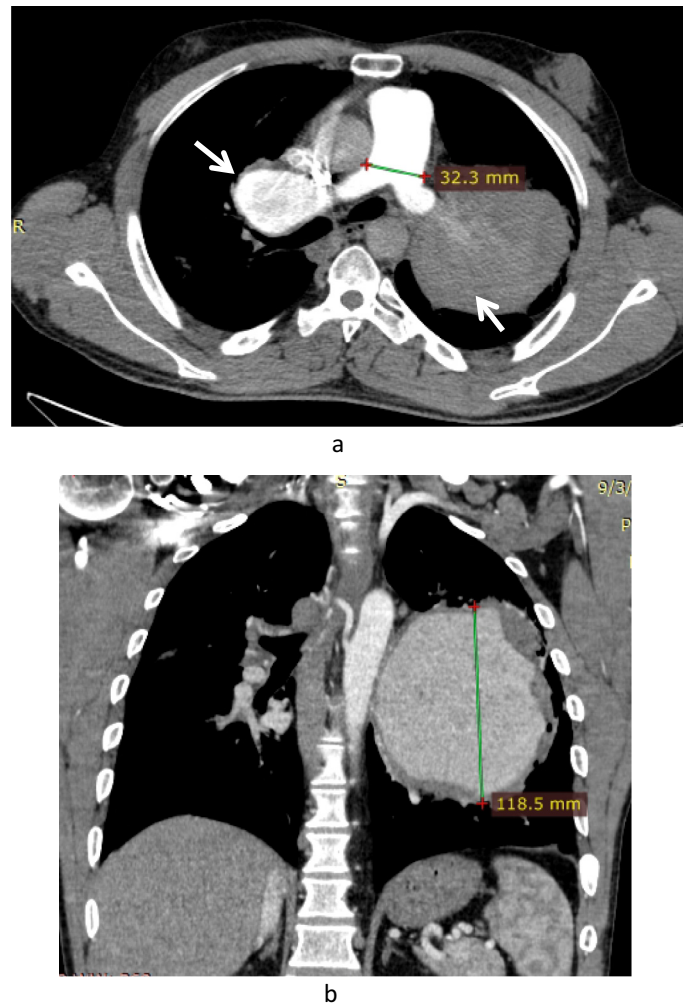


Fig. 1. (a) Axial CT pulmonary angiography of 55 years male patient showing bilateral pulmonary aneurysms (arrows) larger on left side as well as pulmonary hypertension with the diameter of pulmonary trunk (32 mm). (b) Coronal CT pulmonary angiography of the same patient showing left pulmonary aneurysm measuring 11.8 cm with partial thrombosis evident in delayed scans.

Chest X-ray is usually used for patients with respiratory symptoms and in BD it will diagnose thoracic aorta or pulmonary artery dilatation but not specific for the findings of lung parenchyma as commonly to appear non-specific opacities [1].

After development of MDCT and pulmonary angiography, now they are playing an important role in diagnosis of chest findings related to BD including pulmonary parenchymal, vascular as well as cardiac complications [1].

2. Aim of the study

The aim of this study was to identify the possible chest imaging findings in patients with Behcet's disease and complaining of chest symptoms by using MDCT chest and pulmonary angiography to assess the lung, pleura, mediastinum as well as the thoracic vasculature.

3. Patients

3.1. Inclusion criteria

During the period from February/2014 till February/2016 there were 200 known patients with Behcet's disease who were referred from the outpatient clinics, or admitted in Alexandria University Hospitals; 110 out of 200 patients had chest complains, out of these 110 patients, in only 85 patients MDCT chest was recommended and out of these 85 patients only 40 cases did CT pulmonary angiography to explain their chest symptoms according to imaging findings.

This retrospective study included 18 patients out of 40 who did CT pulmonary angiography and showed positive findings.

All patients fulfilled the International Study Group Criteria for diagnosis of Behcet's Disease (International Study Group Criteria, 1990) [11].

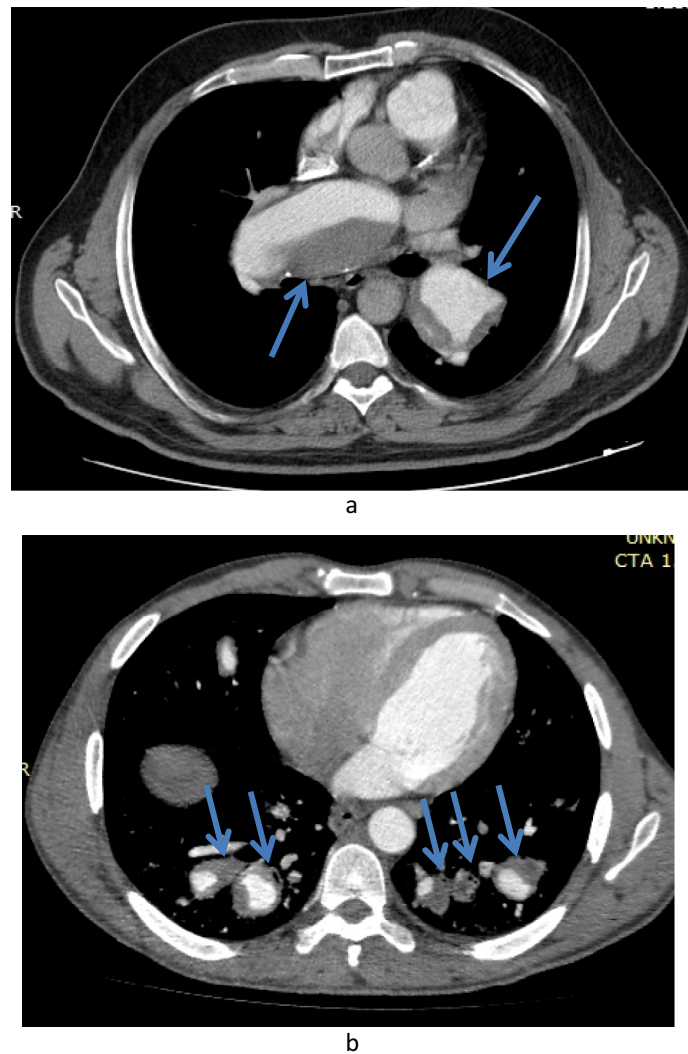


Fig. 2. Axial CT pulmonary angiography (a and b) in 35 years male patient showing multiple variable sized bilateral pulmonary aneurysms at different levels with partial thrombosis (arrows).

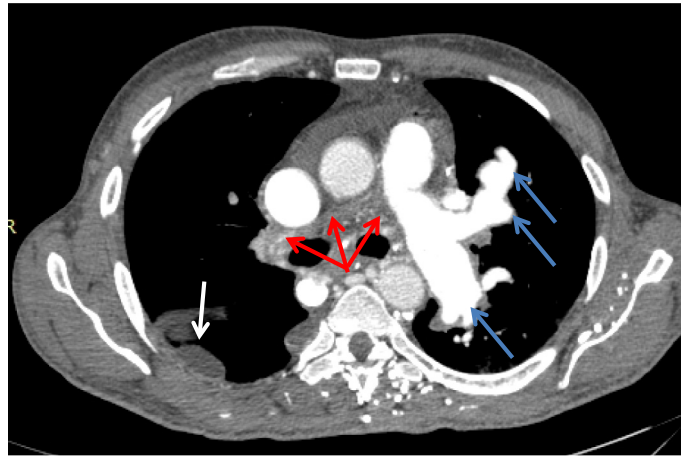
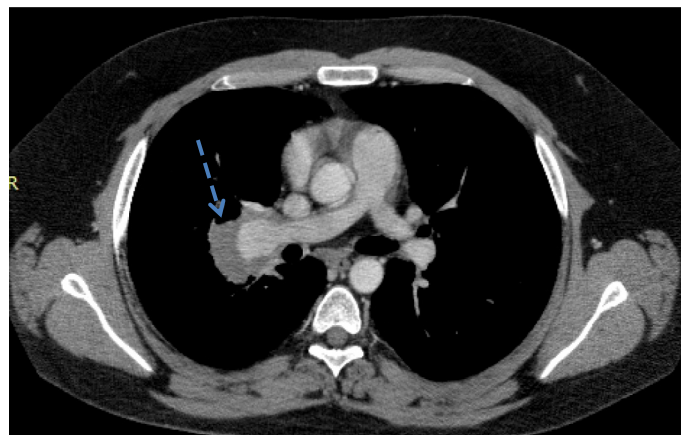
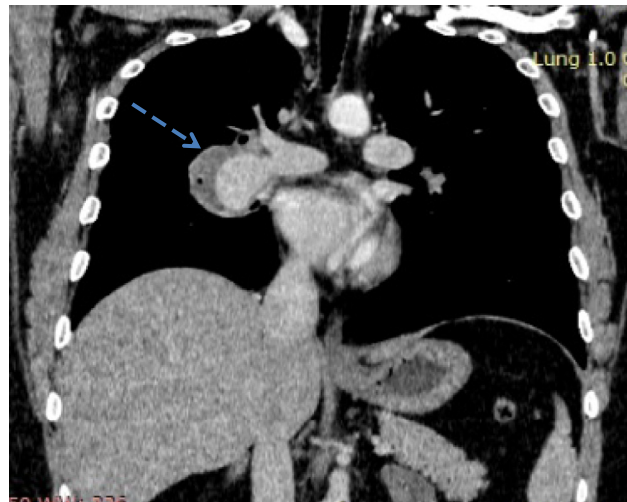


Fig. 3. Axial CT pulmonary angiography in 55 years male patient showing multiple unilateral (left) pulmonary aneurysms (blue arrows) and mild right pleural effusion (white arrow), and also noted total occlusion of the right pulmonary artery (red arrows). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)



a



b

Fig. 4. (a and b) Axial and coronal CT showing single partially thrombosed right lower lobar pulmonary artery aneurysm (dashed arrows).

Exclusion criteria:

- (1) Known patients with BD referred for non-contrast CT chest or only routine CT chest without pulmonary angiography.
- (2) Patients with incomplete medical records.

Patient consent was waived by the Research Ethics Board, assuring respect of the confidentiality of the medical record.

4. Methods

All patients were subjected to the following:

- (1) Full history taking.
- (2) Clinical assessment with emphasized chest examination.
- (3) Routine laboratory investigations included ESR, CRP, CBC, liver and renal function tests.
- (4) Ophthalmological examination.
- (5) MDCT chest with pulmonary angiography using Toshiba Aquilion 64 slices.
 - (a) A post contrast study of 1-mm collimation multislice CT acquisition with a 500-ms gantry rotation time and a pitch of 6 (12-mm table feed/s), which covers a range from the aortic arch to the base of the heart in 8–10 s, was performed. Non-ionic water-soluble contrast medium was

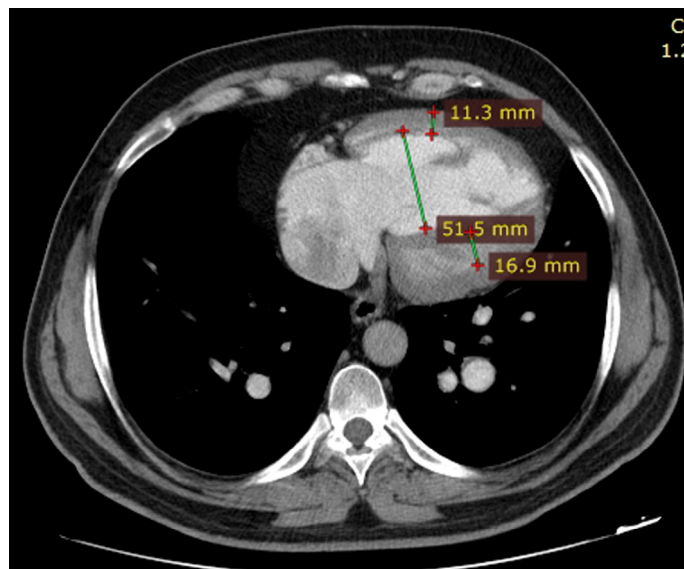


Fig. 5. Axial CT pulmonary angiography of a 35 years male patient showing right ventricular strain in the form of increased thickness of right ventricular wall reaching 11.3 mm and dilated right ventricle compared to the left ventricle.

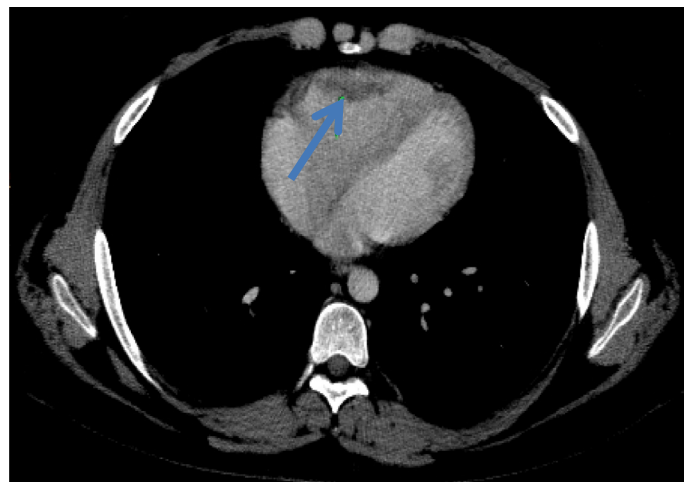


Fig. 6. Axial CT pulmonary angiography of a 38 years male patient showing right ventricular thrombus in the form of filling defect adherent to the anterior wall of the right ventricle (arrow).

injected using an automated injector according to the pulmonary circulation time adjusted after performing the time–density curve.

- (b) The images were revised using dedicated workstation (vitrea) and reconstructed in both soft tissue and high resolution windows. Also the source images were reconstructed in sagittal, coronal and oblique planes.
- (c) Image interpretation.
- o Pulmonary vasculature, aneurysm or thrombosis.

- o Venous structures: SVC, IVC and innominate veins.
- o Parenchymal, pleural and mediastinal findings.

5. Results

The study included 18 male patients. Their ages ranged from 28 years to 55 years with mean age of 42 years. Clinical data of the studied cases: (Table 1) MDCT angiographic findings of the studied cases: (Table 2).

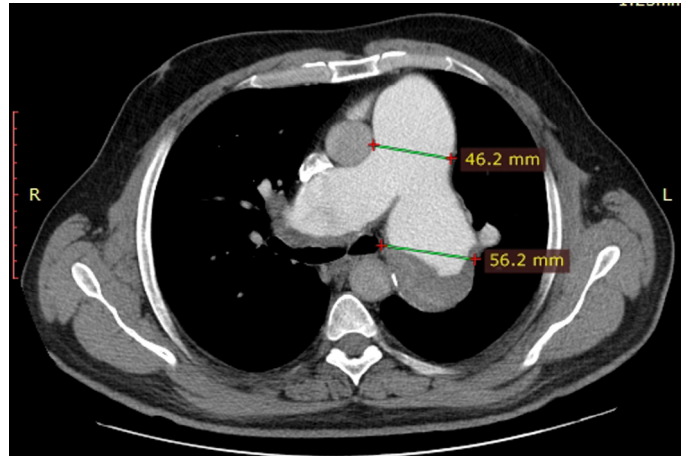


Fig. 7. Axial CT pulmonary angiography of a 35 years male patient with pulmonary hypertension showing increased diameter of the pulmonary trunk above 30 mm and main pulmonary artery above 23 mm.

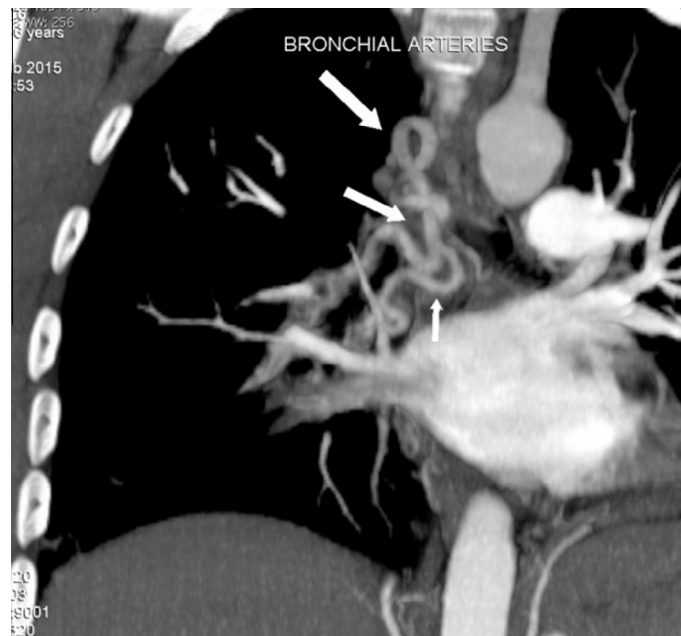


Fig. 8. Male patient 42 years showing multiple dilated bronchial arteries in coronal CT pulmonary angiography (arrows).

Clinical presentation was variable among the studied cases but most of them (16 cases) presented with dyspnea, 12 cases with hemoptysis, 9 cases with chest pain.

Aneurysmal dilatation of the pulmonary artery was the commonest finding in the studied cases as it was found in 16 out of 18 patients. Multiple bilateral pulmonary artery aneurysms were detected in 10 patients, and the largest aneurysm reached 11.8 cm and showed partial thrombosis (Figs. 1 and 2). Two patients showed multiple unilateral small aneurysms on the left side (Fig. 3), four patients showed single aneurysm involving the right lower lobar branch and both of them with partial thrombosis (Fig. 4).

6 out of 18 patients showed right ventricular strain (Fig. 5) and only 4 patients showed right ventricular thrombus (Fig. 6).

Pulmonary hypertension was detected in 12 patients in the form of dilated main pulmonary artery with their diameters ranged from 30 to 46 mm (Fig. 7) while the remaining patients showed normal diameter of the pulmonary artery.

8 patients out of 18 showed dilated bronchial arteries representing extra-pulmonary systemic collaterals in the context of chronic thrombo-embolism (Fig. 8).

14 patients showed pulmonary embolism (PE), including 6 cases with acute embolism involving the sub-segmental branches of pulmonary arteries bilaterally in the form of filling defects while the other 8 patients showed chronic embolism in the form of attenuated irregular sub-segmental branches of both pulmonary arteries,

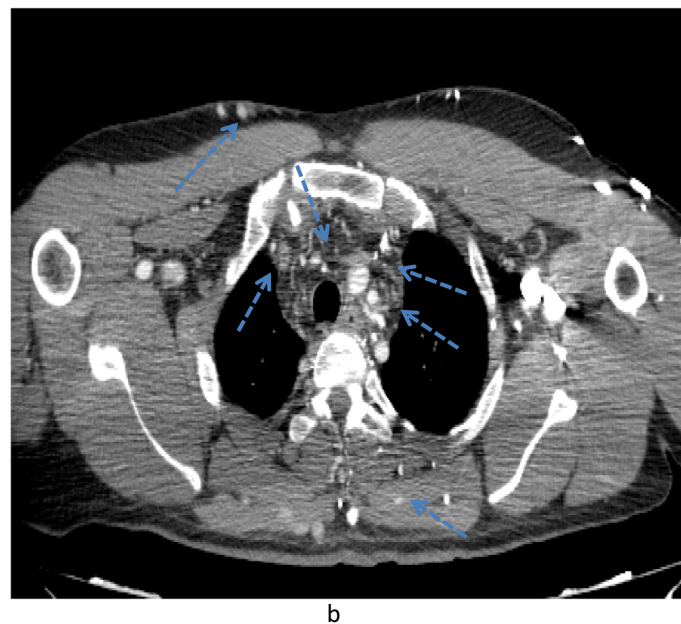
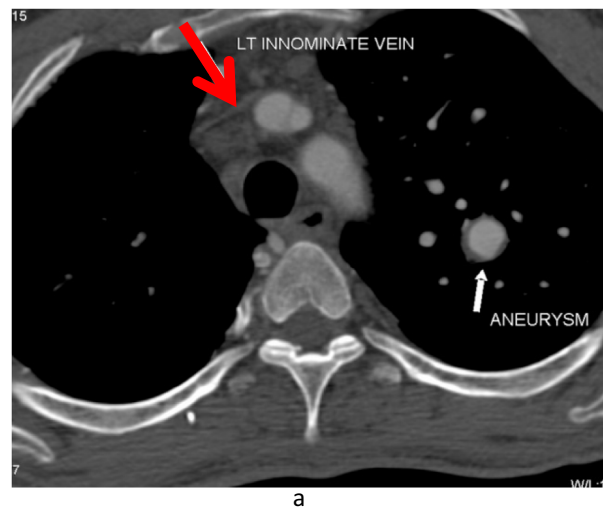


Fig. 9. Axial CT pulmonary angiography of 48 years male patient showing occlusion of innominate vein (red arrow) (a) and SVC occlusion. The patient also shows left upper lobar pulmonary artery aneurysm (white arrow) (b) with extensive chest wall and mediastinal collateral veins (dashed arrows). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

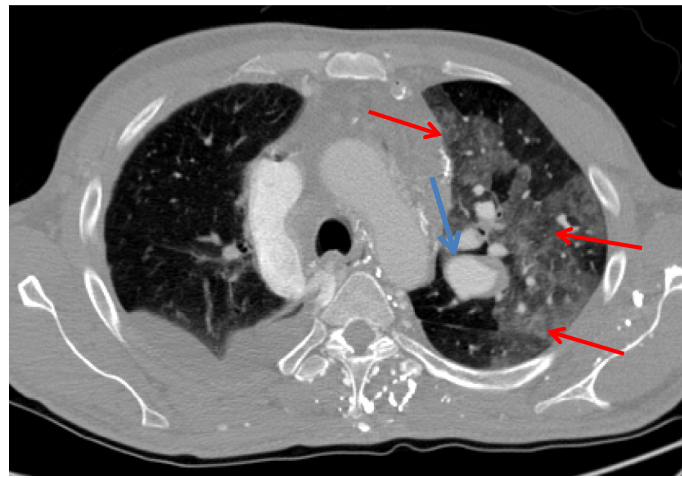


Fig. 10. Axial CT lung window of a male patient aged 48 years presented by dyspnea with evidence of left lung Mosaic attenuation (red arrows) secondary to chronic PE. Also noted left upper lobar pulmonary artery aneurysm (blue arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

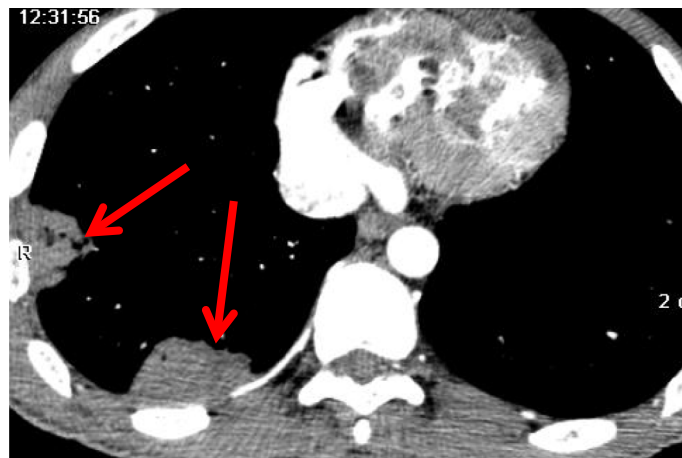


Fig. 11. Male patient aged 28 years presented by frank hemoptysis followed by dyspnea, CT showing right sided pleural based, wedge shaped, consolidative parenchymatous lung lesions seen at the right lower lobe mostly representing small pulmonary infarcts (red arrows). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

secondary pulmonary hypertension and dilated bronchial arteries (Figs. 7 and 8).

Two patients showed occlusion of superior vena cava and two patients showed occlusion of both superior vena cava and innominate vein (Fig. 9).

Right pulmonary artery occlusion was detected only in one patient (Fig. 3).

5.1. MDCT chest findings in the studied cases

12 patients out of 18 showed mosaic attenuation of both lungs (Fig. 10) and 4 out of 18 cases had pulmonary infarcts (Figs. 11 and 12).

In the studied cases only four patients had pleural effusion, and it was mild in amount on the right side (Fig. 3).

6. Discussion

Behçet's disease is chronic multisystem idiopathic inflammatory disease with vasculitis being the main pathology in the course of disease. It is common to occur in Mediterranean countries and in Asia in middle age group [12–14]. In our study the mean age of the studied cases was 42 years.

Multidetector CT as well as CT angiography has an important role in diagnosis of BD related complications including pulmonary parenchymal and vascular structures in addition to mediastinum and pleura [14]. So we included in our study BD patients who had MDCT pulmonary angiography and excluded other patients with only non-contrast or non-angiographic CT study of the

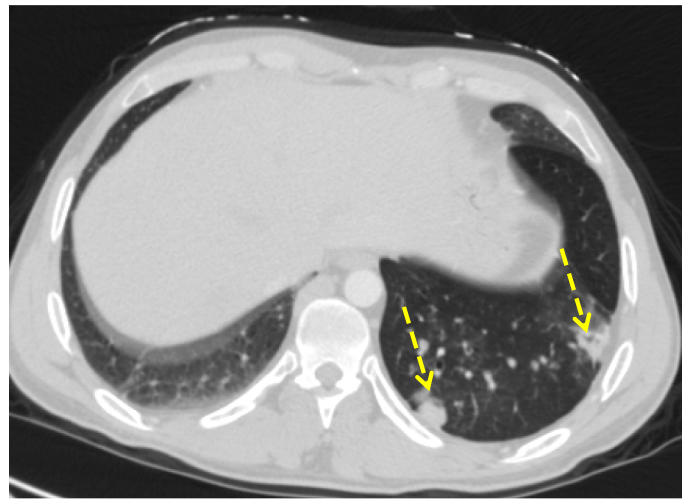


Fig. 12. Axial CT lung window showing infarcts (dashed arrows) in another 55 years patient presented with dyspnea and hemoptysis.

chest for better evaluation of pulmonary vasculature and diagnosis of the presence of vascular complications.

Hiller et al. in 2004 [15] proved that the most common pulmonary vascular finding in Behçet's disease is pulmonary artery aneurysm with or without thrombosis, and this is in agreement with our cases as we found pulmonary artery aneurysms in 16 out of 18 cases (89%).

Pulmonary vasculitis observed in BD is affecting great and medium sized vessels (according to the Chapel Hill classification criteria for vasculitis), with venous affection in the form of thrombophlebitis more than arterial affection; however, arterial complication with aneurysm formation is more dangerous for fear of rupture and it is the main cause of mortality among BD patients [1,2,14]. In contrary to our study we found venous occlusion only in four patients (22.2%), pulmonary artery occlusion in one patient (5.5%) and pulmonary artery aneurysm in 16 patients (89%) either single or multiple aneurysms with the largest of them showing partial thrombosis. This may be explained by small number of selected patients according to inclusion criteria in our study.

Emad et al. [16] concluded that BD may affect the pulmonary arteries at different levels either the main pulmonary arteries, lobar, segmental or the subsegmental branches. It has been also reported that the pulmonary arteries affected in Behçet's disease range from the lobar and segmental branches down to the arterioles. This was confirmed in the current study as we detected lobar and segmental arteries aneurysms; multiple bilateral aneurysms were found in 10 cases, and multiple unilateral aneurysms in 2 case while single aneurysm in 4 cases.

Neves et al. in 2013 [9] mentioned that intracardiac thrombosis is a rare complication of BD which is common in males and common to occur at right ventricle. The mechanism of intracardiac thrombosis is still unknown

but may be due to in situ process, secondary to venous thrombosis or endomyocardial fibrosis. This is in agreement with our study as we found intracardiac thrombosis only in two male patients which involve the right ventricle.

Pulmonary vasculitis in Behçet's disease can occur in different forms: stenosis, occlusion or thrombosis of pulmonary vessels. Pleural effusion can occur in Behçet's disease secondary to pulmonary vascular complications [17,18]. In the current study we found four patients (22%) with pleural effusion and it was mild in amount.

Naim et al. [1] described that pulmonary parenchymal and pleural affection can occur in 1–10% of patients with Behçet's disease. In the current 4 patients (22%) were complicated with mild pleural effusion, 4 patients (22%) with pulmonary infarcts and 12 patients (66.6%) with mosaic attenuation of the lung.

Pulmonary parenchymal damage in Behçet's patients can occur and may be replaced by fibrosis, and consequently air way narrowing with air trapping and the end result will be mosaic attenuation [19]. This was confirmed in our study as 12 cases (66.6%) in the current study showed mosaic attenuation of the lung.

Due to small number of patients with positive findings in CT pulmonary angiography who were included in this study, it was difficult to correlate statistically the CT findings with the clinical data.

7. Conclusion

Behçet's disease is an idiopathic multisystem disease with vasculitis (including the pulmonary vessels) as a major complication and it is the main cause of morbidity and mortality. MDCT pulmonary angiography is an important diagnostic imaging tool for diagnosis of vascular complications in patients with Behçet's disease and we should

keep high suspicion in these patients for early detection of vascular complications in order to reduce the mortality rate.

Conflict of interest

The authors declared that there is no conflict of interest.

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