RADIOLOGIC FEATURES OF PULMONARY ARTERIAL ANEVRYSMS DUE TO BEHÇET'S DISEASE WITH IMMUNOSUPPRESSIVE THERAPY : A CASE REPORT AND REVIEW OF THE LITERATURE

CARACTÉRISTIQUES RADIOLOGIQUES DES ANEVRYSMES ARTÉRIELS PULMONAIRES SECONDAIRE A LA MALADIE DE BEHÇET SOUS TRAITEMENT IMMUNOSUPPRESSEUR

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Abstract

Behçet's disease (BD) is a systemic vasculitis characterized by recurrent oral and genital ulcers, uveitis, arthritis, and involvement of the gastrointestinal tract, central nervous system, and blood vessels. In the thorax, it may cause vascular complications, affecting the aorta, brachiocephalic arteries, bronchial arteries, pulmonary arteries, pulmonary veins. After the aorta, the pulmonary arteries are the most common site of arterial involvement among the pulmonary manifestations in patients with BD. We report a case of a 24-year-old man with an established diagnosis of Behçet's disease since ten years presented with cough and hemoptysis. Pulmonary arterial aneurysms (PAA) was diagnosed. This report aims to demonstrate the imaging findings of PAA seen in BD, and the effectiveness of immunosuppressive treatment which led to partial resolution and thrombosis of PAAs.

Key -words: Pulmonary artery aneurysm ;Behçet's disease ; CT Scan ; Imaging features.

Résumé

La maladie de Behçet est une vascularite systémique caractérisée par des ulcères buccaux et génitaux récurrents, une uvéite, une arthrite, une atteinte du tractus gastro-intestinal, du système nerveux central et des vaisseaux sanguins. Au niveau du thorax, elle peut entraîner des complications vasculaires, affectant l'aorte, les artères brachiocéphaliques, les artères bronchiques, les artères et les veines pulmonaires. Après l'aorte, les artères pulmonaires sont le site le plus fréquent d'atteinte artérielle avec des manifestations thoraciques chez les patients atteints de BD. Nous rapportons le cas d'un homme de 24 ans ayant un diagnostic établi de la maladie de Behçet depuis dix ans qui consulte pour toux et hémoptysie. Le diagnostic posé était des anévrismes des artères pulmonaires (AAP). Le but de cette observation est de présenter les signes radiologiques des AAP observés dans la maladie de Behçet, et l'efficacité du traitement immunosuppresseur qui a conduit à la diminution en taille et à la thrombose partielle des AAP.

Mots - clés : Anévrismes des artères pulmonaires ; Maladie de Behçet ; Scanner ; Aspects radiologiques.

ملخص

مرض بهجت هو التهاب الأوعية الدموية الجهازي الذي يتميز بقرح متكررة في الفم والأعضاء التناسلية والتهاب القزحية والتهاب المفاصل وتلف الجهاز الهضمي والجهاز العصبي المركزي والأوعية الدموية. في القفص الصدري ، يمكن أن يسبب مضاعفات الأوعية الدموية التي تؤثر على الشريان الأورطي والشرايين العضدية الرأسية والشرايين القصبية والشرايين والأوردة الرئوية. بعد الشريان الأورطي ، تعد الشرايين الرئوية أكثر الأماكن شيوعًا لتأثير الشرايين مع المظاهر الصدرية في مرضى بهجت. قمنا بدراسة حالة رجل يبلغ من العمر 24 عامًا تم تشخيص إصابته بمرض بهجت لمدة عشر سنوات و قد تمت استشارتنا بعد تعرضه لحالة سعال ونفث الدم. كان التشخيص هو تمطط الأوعية الدموية في الشريان الرئوي (PAA). الهدف من هذه الملاحظة هو عرض العلامات الإشعاعية لـ PAA التي لوحظت في مرض بهجت ، وفعالية العلاج المثبط للمناعة الذي أدى إلى انخفاض الحجم والتخثر الجزئي الناجم عن المرض.

الكلمات المفاتيح: تمطط الأوعية الدموية في الشريان الرئوي ; مرض بهجت ; الأشعة المقطعية ; المظاهر الإشعاعية.

INTRODUCTION

Behçet's disease (BD) was firstly described by Hulusi Behçet in 1937[1]. It is a systemic vasculitis characterized by recurrent oral and genital ulcers and uveitis, arthritis, and involvement of the gastrointestinal tract, central nervous system, and blood vessels[1]. In the thorax, it may cause vascular complications, affecting the aorta, brachiocephalic arteries, bronchial arteries, pulmonary arteries, pulmonary veins. Pulmonary artery aneurysm (PAA) is reported in 1.5 % of adults with BD and affects mainly young men. It carries a bad prognosis. However, it may become smaller or disappear with medical treatment. Mural thrombotic changes may be observed during the regression of pulmonary artery aneurysms [2,3].

We report a case of a 24-year-old man with an established diagnosis of Behçet's disease since 2010 presented with cough and hemoptysis. Pulmonary arterial aneurysms(PAA) was diagnosed. This report aims to demonstrate the imaging findings of PAA seen in BD and the effectiveness of immunosuppressive treatment which led to partial resolution and thrombosis of PAAs.

CASE REPORT

A 24-year-old man with an established diagnosis of Behçet disease since 2010.

In July 2018, he was admitted to the internal medicine department complaining of cough, hemoptysis, and shortness of breath. An angio-CT scan was performed.

It showed bilateral pulmonary embolism in both lobar and segmental pulmonary arteries.

The patient was treated with Low-molecular-weight heparin (LMWH) than Vitamin K antagonists (VKA) associated with methylprednisolone followed by oral azathioprine.

In November 2018, a Chest CT scan showed arterial pulmonary aneurysms and the persistence of pulmonary embolism.

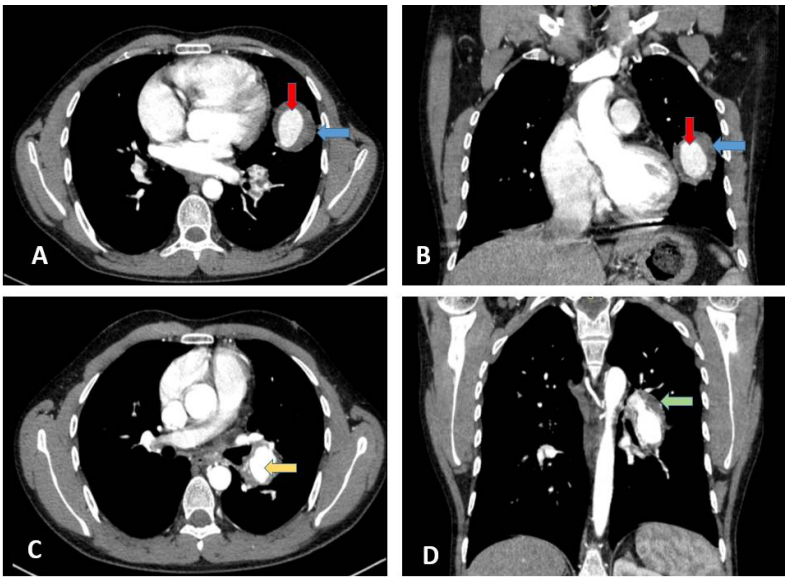
Because of the radiological aggravation of the lesions, we decided to change the treatment. The patient was given subsequent corticosteroids and cyclophosphamide.

In June 2019 (Seven months after cyclophosphamide therapy), the patient presented with a cough and acute hemoptysis. Chest computed tomography (CT) was performed showing large arterial pulmonary aneurysms interesting the lingular artery and inferior lingular segment, an intracardiac thrombus in the right ventricle and the absence of pulmonary embolism (Figure 1). The decision was to continue the immunosuppressive therapy.

In August 2020 (after 21 months of cyclophosphamide therapy), the patient was readmitted for checkup and reevaluation.

CT scan was performed and showed the increase of thrombosis and decrease of the aneurysmal lumen interesting the lingular pulmonary artery. We have seen a total regression of the inferior lingular segmental artery which was replaced by a cavity with an air-fluid level communicating with the inferior lingular segmental bronchus (Figure 2).

The patient was a candidate for surgical treatment.



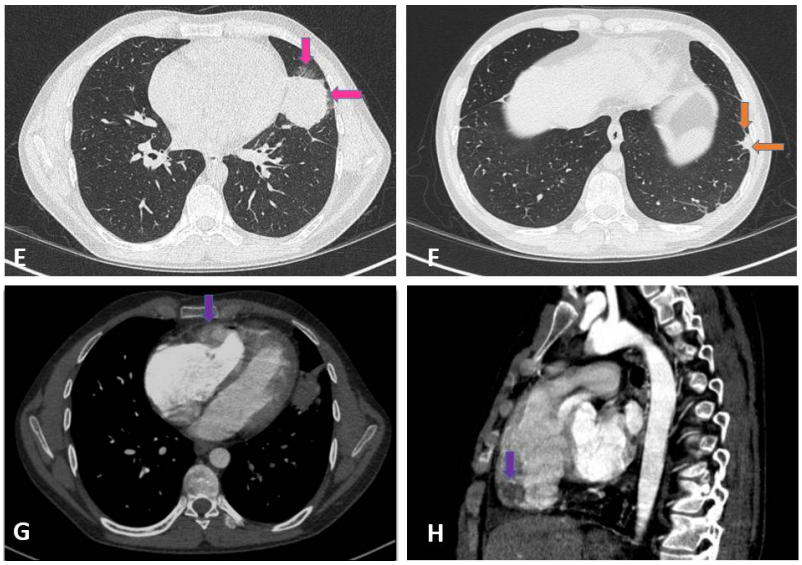


FIGURE 1 : Contrast-enhanced chest CT scan
(7 months after cyclophosphamide treatment)

(A, B) Axial and coronal mediastinal window images showing large pulmonary artery aneurysm of the inferior lingular segmental artery (red arrow) lined by thrombi (blue arrow) and densely opacified on a volumetric high-resolution CT. (C,D) Axial and Coronal mediastinal window images showing the lingular artery aneurysm (yellow arrow) with irregular mural thrombi(green arrow), densely opacified on a volumetric high-resolution CT. (E) Axial lung window image shows the lingular pulmonary artery aneurysm with circumferential ground glass opacification because of surrounding hemorrhage (pink arrow): It was attributed to intra-parenchymal hemorrhage. (F) axial lung window image demonstrates a focal triangle peripheral area of high attenuation that represents peripheral pulmonary infarction (orange arrow). (G, H) Axial and sagittal mediastinal window images demonstrate a low attenuating mass-like lesion (purple arrow) in the right ventricle without enhancement proven an intracardiac thrombus.

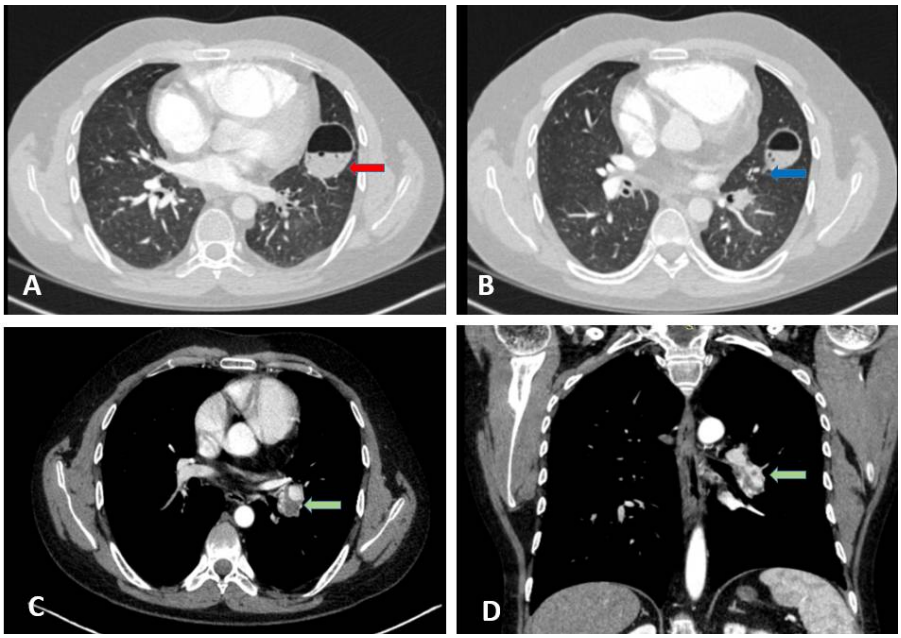


FIGURE 2: Contrast-enhanced chest CT scan
(21 months after cyclophosphamide therapy)

(A, B) Axial lung window images showing total regression of the inferior lingular segmental artery. Parenchymal change as perianeurysmal consolidation. The aneurysm was replaced by a cavity with air-fluid level (red arrow) communicating with the inferior lingular segmental bronchus (blue arrow). (C, D) Axial and Coronal mediastinal window images showing increased thrombosis and decreased aneurysmal lumen (green arrow) interesting the lingular pulmonary artery.

DISCUSSION

BD is a systemic vasculitis characterized by recurrent oral and genital ulcers, uveitis, arthritis, and involvement of the gastrointestinal tract, central nervous system, and blood vessels.

Vascular involvement in the form of venous or arterial occlusion and aneurysm formation is seen in 25% of patients, and it is the most common cause of mortality in Behçet's disease [4]. Such a form of Behçet's disease is called ''vascular-Behçet's disease[5]. Arterial involvement is less common than venous involvement and accounts for 12% of vascular manifestations[5]. Arterial lesions may develop in the aorta or the pulmonary artery and its major branches, occurring as an aneurysm in 65% of patients and as an occlusion in 35%[5].

Pulmonary artery aneurysms (PAA) are mainly seen in young males and very rarely in females. PAAs are located most frequently in the right lower lobe pulmonary artery, followed by the right and left main pulmonary arteries[6].

Focal enlargement of the pulmonary artery can be the first radiologic sign of a pulmonary aneurysm.

Hemoptysis of varying degrees is the most common and predominant symptom, our patient presented with acute hemoptysis in multiples occasions, decreased gradually after immunosuppressive therapy. Possible causes for hemoptysis include rupture of an aneurysm with erosion into a bronchus[7] and thrombosis of pulmonary vessels[8]. Pulmonary artery aneurysms may cause major bleeding and is often associated with a poor prognosis with less than 50% 2-year survival, requiring early prompt diagnosis and treatment.

Intracardiac thrombi in BD may result from endomyocardial fibrosis[9], which may be sequelae of vasculitis involving in endocardium, myocardium, or both[8]. As intracardiac thrombus is tightly attached to the endocardium, embolism from the cardiac cavity seems to be relatively uncommon[10]. Young males seem to be most at risk than female and the right heart is the most frequent site of involvement for intracardiac thrombus formation[11].

The diagnosis of intracardiac thrombi in BD may be made using cardiac magnetic resonance imaging, computed tomography, and transthoracic echocardiography, which may show a mass in the heart chambers; sometimes, indistinguishable from infective vegetations or a tumor and myxoma[12].

Therapeutic options for the management of PAAs in Behçet's disease include medical treatment[13],[14], surgical repair[6], and trans catheter embolization[7].

The European League Against Rheumatism currently recommends the use of immunosuppressive agents such as corticosteroids, cyclophosphamide, and azathioprine for treating major vessel diseases.

Anticoagulant therapy is not recommended for the treatment of DVT(deep vein thrombosis) and/or pulmonary artery thrombosis due to the increased risk of fatal hemoptysis [15], but ,there are no controlled studies of anticoagulants or antiplatelet aggregation therapy and there is a lack of consensus on their use[15].

In this case, Combined anticoagulant and immunosuppressive therapy played an important role in the treatment of pulmonary embolism.

Immunosuppressive therapy is found to be most beneficial when administered at the early stages of the disease before irreversible damage of the arterial wall develops[6]. However, the course of pulmonary arterial aneurysms treated with immunosuppressive therapy can vary from an aneurysm to another. The ultimate response of treated PAA is progressive regression and the duration of immunosuppressive therapy may vary.

Tunaci et al. reported complete disappearance or regression of PAA during a 3–42 (mean 21)-month follow-up period of 13 Behçet's patients receiving immunosuppressant treatment [16]. Thirty-one initially non thrombosed aneurysms (67%) first became thrombosed during treatment: later, the thrombus regressed and the aneurysm decreased in size. Peri-aneurysmal consolidation and air-space nodules detected in seven patients disappeared in the early stages of treatment. Mosaic attenuation areas were seen in eight patients and disappeared in seven (88%) after treatment [16].

Medical treatment should be the first-line treatment if PAAs are multiple and have a tendency to recur unless there is massive hemoptysis[17]. Surgical treatment may be necessary in cases of massive hemoptysis, but mostly consists of major anatomical resection rather than preserving lung tissue. Moreover, it has the disadvantages of aneurysm recurrence requiring repeated thoracotomy and other complications such as perivascular leaks, graft thrombosis, and anastomotic leaks due to the pathologic nature of the disease[17].

CONCLUSION

PAA is the most frequent form of thoracic involvement in BD, with hemoptysis being the most common clinical presentation. PAAs associated with BD tend to be multiple.

Chest CT scan plays an important role for the diagnosis and evaluation of PAA after medical or surgical treatment.

The anti- inflammatory and immunosuppressive drugs are essential for the treatment of PAAs in patients with BD.

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