A case report of pulmonary artery aneurysms in Behcet's disease

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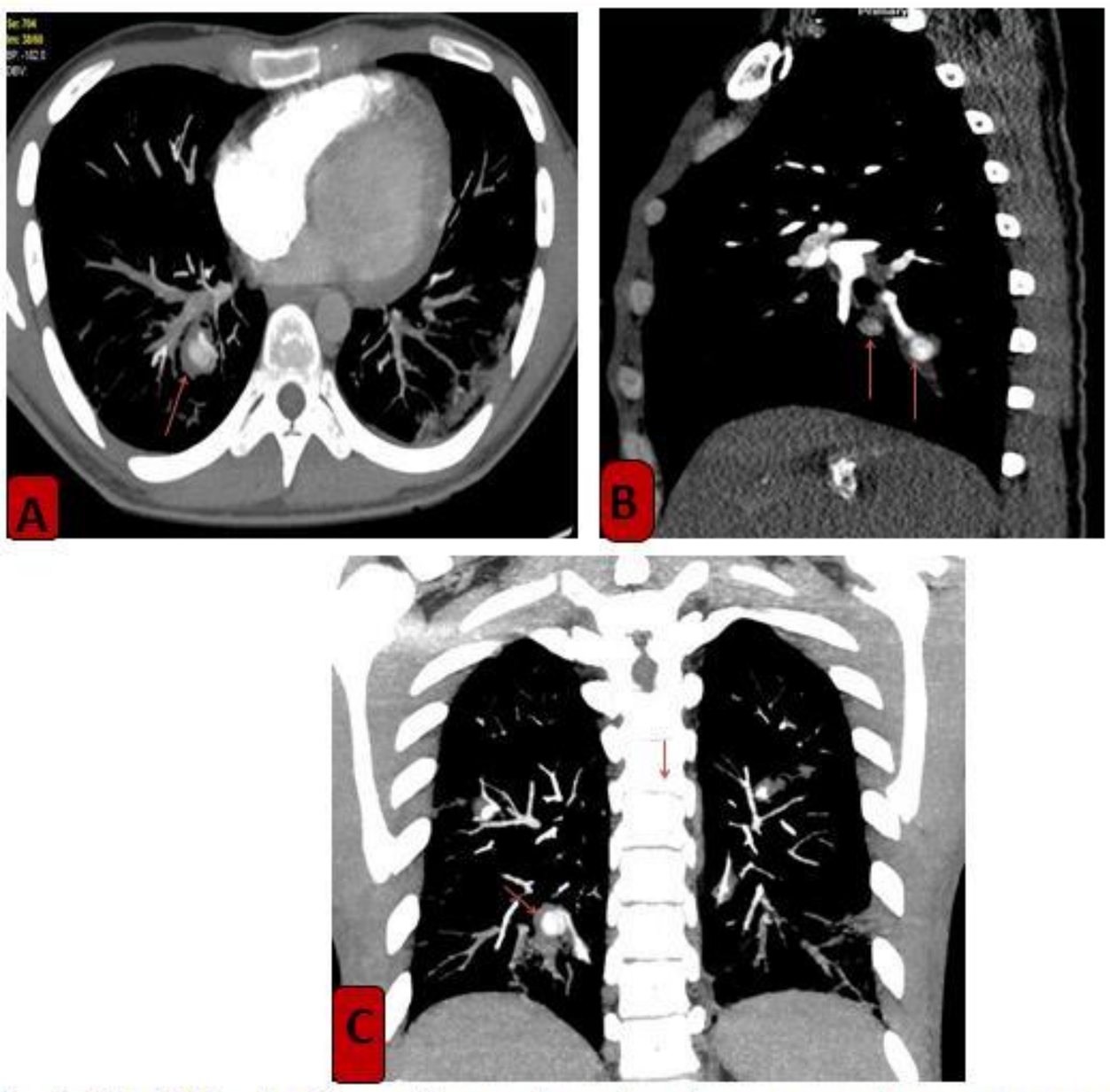
Introduction

Behçet disease is a rare multisystemic and chronic inflammatory disorder with an unknown cause. 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.Vasculitis can affect arteries and veins of any size; however, venous system involvement is more frequent than arterial system involvement. Rapid advances in MDCT scanners allow a more detailed analysis of arterial and venous structures. Arterial system involvement is rare and accounts for only 10– 15% of vascular system involvement. Inflammation of the vasa vasorum destructs elastic fibers and results in vessel lumen dilatation and aneurysm formation. The pulmonary artery is rarely affected with pulmonary aneurysms formation that has a poor prognosis of the disease.

Case Presentation

Male patient 27 years old suffering from Behcet disease 9 years ago, presented with hemoptysis and sent for CT chest angiography. CT pulmonary angiography revealed multiple well-defined saccular aneurysmal dilatation are seen arising from the posterior segmental branch of the pulmonary artery of the right lower lung lobe with partial thrombosis surrounded by perifocal alveolar opacity, distally the related segmental branch shows filling defects that is impressive of vasculitis with partial thrombosis in segmental pulmonary aneurysm. Other multiple small saccular aneurysmal dilatation are seen arising from related segmental branch of pulmonary arteries of the superior segment of the right lower lobe, anterior segment of left upper lobe , postro-medial segment of left lower lobe , posteromedial segment of the right lower lobe. The patient started to take anticoagulant in addition to immunosuppressive agents and sent to the thoracic surgeon and interventional radiology for consultation and take action for treatment of the pulmonary aneurysms.



A. Axial CT chest angiography showing saccular aneurysm (arrow) of the posterior inferior branch of the right pulmonary artery with peripheral thrombus. Subpleural patchy areas of consolidations are seen in the lateral segment of the left lower lung lobe.
B. Sagittal reformate of CT chest angiography shows multiple bilateral variable sized saccular aneurysms are seen arising from the pulmonary artery branches (arrows).
C, Coronal reformate of CT chest angiography shows multiple bilateral variable sized saccular aneurysms are seen arising from bilateral pulmonary artery branches (arrows) with peripheral thrombosis and patchy areas of distal consolidation.

Discussion

In Behcet's disease; one cause of exacerbation of the disease is pulmonary artery aneurysm and, rarely, pulmonary embolism [1]. PAA may be multiple and have high rates of recurrence [2]. PAA have a poor prognosis, especially when the diameter is more than 3 cm. It is estimated that 30% of BD patients with PAA die within 2 years. Hemoptysis (sometimes massive) is often the first clinical sign [1]. In this case, emergency treatment with open surgery or embolism is the only choice. Otherwise, invasive methods are recommended in isolated PAA. Also it is important that enhanced immunosuppressive therapy is preceded, as it seems to prevent postoperative complications such as pseudoaneurysms and bronchopulmonary arterial fistulas [3]. In our case; computed Tomography angiography was done to detect the cause of haemoptysis. CT angiography revealed pulmonary arteries aneurysms with clots inside. In this case, the use of anticoagulants is controversial, since there is an increased risk of bleeding and even fatal haemoptysis. Many studies show that immunosuppressive therapy combined with corticosteroids results in remission and even disappearance of PAA [4]. Administration of anticoagulants is considered by many researchers with skepticism because of the different mechanism of thrombosis in the BD [3]. It is an in situ thrombosis as the result of vasculitis [2]. Inflammation affects all layers of the vessels and leads to diffuse endothelial cell injury, which is the key factor that predisposes to thrombosis. In non-extensive PE, the use of antiplatelet agents was regarded sufficient in the past [4]. Thus, if there is any indication of pulmonary vasculitis, increased bleeding risk and suspicion of hidden aneurysms, anticoagulants are contraindicated [5].

Conclusion

In view of the above, we conclude that BD as a vasculitis predisposes to pulmonary aneurysms with high possibility of thrombotic events that lead to high mortality rate, the treatment of which should be personalized. The new guidelines on PE correctly include autoimmune diseases among the risk factors for thrombosis. For these reasons, clinicians should include them in their differential diagnosis.

References

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