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Case Report

Pulmonary Artery Aneurysms (PA) Presented in Behcet Disease: Case Report

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Abstract

Introduction: Behcet disease is poly-systemic complication secondary to occluded vasculitis. It generally presents with mouth sometimes with reproductive organ ulcers however and other system involvements may be the initial sign. It may also involve the thorax and manifesting by the presence of pulmonary artery (PA) aneurysms.

Case Report: A 29-year old man was referred to our thoracic ward for his many episode of hemoptysis. However, the taking history included with other complaints like mouth, reproductive organ ulcers and one episode of blindness. On investigation, HLA B5 was positive and left pulmonary artery aneurysm and vasculitis was present. Behcet disease however presentation with PA aneurysm may warrant an urgent lobectomy or Pneumonectomy.

Keywords: *Behcet Disease; Aneurysm; Vasculitis; Lobectomy; PA*

Introduction

Behcet, disease is a chronic poly-systemic inflammatory disease whose etiology primes not known and with big incidence in men gender aged between third to fourth decade of life [1,2]. This disease is commonly happened in Mediterranean countries and Asia. The biggest ratio of patients in Turkey, was characterized by Hulusi Behcet through the triad: (recurrent mouth, reproductive organ ulcerations and eye lesion uveitis).

Incidence occurred in many organs have been described with vasculitis being the prime pathological diagnosis [1].

Behcet disease diagnosis is based on some criteria advised by the International Study Group for Behcet Disease (ISGBD) [3]. The prognosis is determined by the engagement of the CNS, GIT and CVS [2]. The vascular system compromise seen in twenty five to thirty percent of cases in Behcet disease although this event is the highest cause of mortality linked to the disease [4,5].

The thorax engagement shows as many findings may be manifestation by radiological studies. The studies on such presentations lead in the disease diagnosis.

Case Presentation

A male, twenty nine year-old patient resident of Chakri, Rawalpindi, Pakistan married, security guard by profession, now presented with fever and hemoptysis was referred from PAEC hospital to Combined Military Hospital, Rawalpindi because of findings seen pulmo-

nary artery aneurysms, for a definitive procedures. The patient reported the onset of high grade fever, night blindness, and some episodes of hemoptysis 2 years back. More, he reported recurrent mouth with reproductive organ ulceration from his adolescence.

At PAEC hospital he was managed for diplopia, reduced vision and uveitis after that he was treated for testicular ulcers and mouth ulcer. Keeping in view the history and clinic-pathological evidence in opinion of Behcet disease and lymphoproliferative disorder. Extensive workup was done at that time which was HLA B5 diagnosed finally and placed patient on steroid, immunosuppression and proton pump inhibiter After diagnosed Behcet disease patient had two episode of hemoptysis in which he advise CT scan thorax which reported pulmonary artery vasculitis and on CTPA suggest pulmonary primary idiopathic large vessel vasculitis with aneurysmal dilatation (Figure), the biggest at lower lobar pulmonary artery showing mural thrombosis and concern for rupture (Figure).

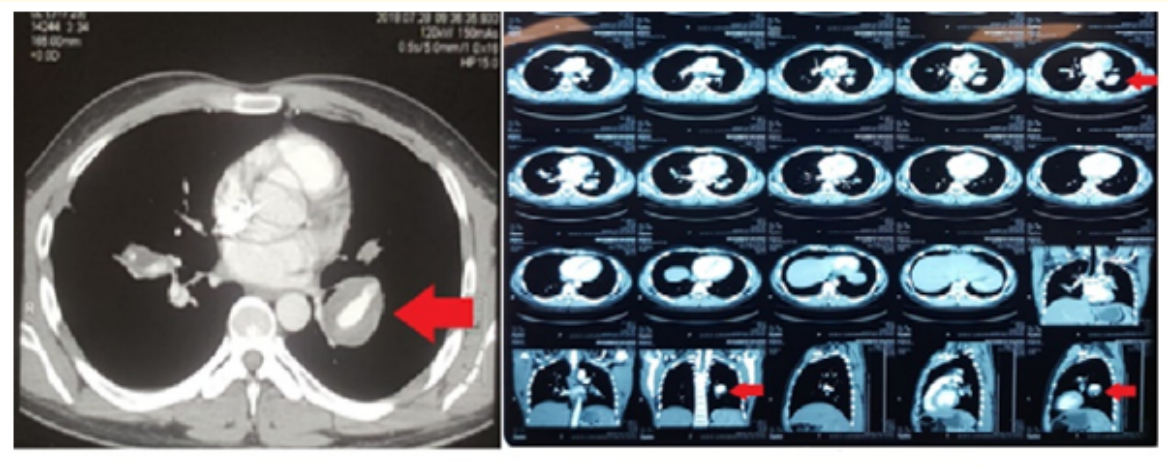

Figure : *Red arrowhead showing pulmonary vessel vasculitis with aneurysmal dilatation.*

Meanwhile at department of thoracic surgery, CMH, Rawalpindi, Pakistan where MDT held with radiologist and they finally suggest left lower lobe lobectomy to avoid rupture catastrophes. At 07/12/2018 under one lung anesthesia poster-lateral thoracotomy performed in which left lower lobe lobectomy performed and on 2nd days all thorax drained removed and on 09/12/2018 patient discharged.

Discussion

As per the classification developed by the ISGBD, the history of recurrent mouth ulcers and any 2 of the mention sings like reproductive organ ulcers, typical eye lesions such as uveitis or retinal vasculitis, dermal lesions or a pathergy test positive define the diagnosis of Behcet disease [3].

Thorax engagement is seen in eight percent of patients and may show through a huge range of differentiation in the mediastinum, going to complications such as fibrosing mediastinitis; in the pulmonary parenchyma where infarction, atelectasis, hemorrhage, diffuse airspace nodules, pneumonia and fibrosis may be occur; in the pleural space as well the blood vessel system [4,6].

Pulmonary vasculitis occurs in Behcet disease affect large and medium caliber vessels "by the Chapel Hill classification for vasculitis", most generally affecting the venous system in eighty five percent, in the shape of thrombophlebitis [4,6]. The venous system complications in Behcet disease are Superior vena caca syndrome, and Budd Chiari syndrome [4]. Arterial system changes are less commonly happened.

The destruction of elastic fibers produced by the inflammatory mechanism of the vasa vasorum consequences in vascular lumen dilatation and development of aneurysms in sixty five percent [4]. Behcet disease is the primary cause of PA aneurysms which may occurs in the segments, or in sub-segmental branches, and that can be seen at CT scan, showing a poor prognosis [4,5]. Aneurysms may regress with specific medical therapy usage of steroid and immunosuppressant therapy, but disease progression and recurrence are more often seen in such patients [4,7].

Ct scan and CT angiography has big role in the diagnosis of Behcet disease. The knowledge on the changes occurs to Behcet disease is useful in the diagnosis, help in the finding of the cause of symptoms, chiefly hemoptysis and leading in the choice of the accurate treatment.

Conclusion

Behcet disease may be diagnose with ease, when it presents with mouth and reproductive organ ulcers or panuveitis. Presentation with PA aneurysm may need an urgent lobectomy or Pneumonectomy.

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