Case Report

Right hilar mass with hemoptysis: An unusual presentation of uncommon disorder

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ABSTRACT

Common differential diagnosis of lung and hilar opacity includes infectious pathology or a mitotic lesion. Behcet's disease (BD) is a rarely diagnosed disease in Indian subcontinent. BD is a multisystem inflammatory disorder that presents with recurrent orogenital ulceration, uveitis, and erythema nodosum. We present here the case of a patient who presented with recurrent hemoptysis with radiological picture of hilar mass, during the evaluation of which the diagnosis of BD was established.

KEY WORDS: Behcet's disease, hemoptysis, lung, vasculitis

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INTRODUCTION

When a patient presents with symptoms of hemoptysis and has a hilar opacity on chest radiograph, the likely diagnosis is bronchogenic carcinoma or less commonly tuberculosis in Indian context. We report a rare case of such a presentation where cause of hemoptysis was Behcet's disease.

CASE REPORT

A 38-year-old man, nonsmoker presented with the history of multiple episodes of hemoptysis of 18 months duration. With each episode of hemoptysis he coughed out 50–100 ml of blood. He had no symptoms of fever, breathlessness, chest pain/discomfort, hoarseness of voice, difficulty in swallowing or weight loss. He had been previously treated with antitubercular treatment on one occasion in the past 18 months with an empirical diagnosis of pulmonary Koch's. Since he did not have any clinical or symptomatic benefit with the antitubercular treatment, his anti-Koch's drugs were stopped after a period of 4 months. He had history of recurrent oral ulceration treated as aphthous ulcers multiple times. He also had history of recurrent genital ulceration.

On examination, he was afebrile, had no lymphadenopathy and had no signs of pulmonary disease. He was found to have multiple oral ulcers on the lower lip and a healed ulcer on the tongue. He had a scrotal ulcer measuring 2 × 2 cm [Figure 1].

On investigation, he was found to be severely anemic

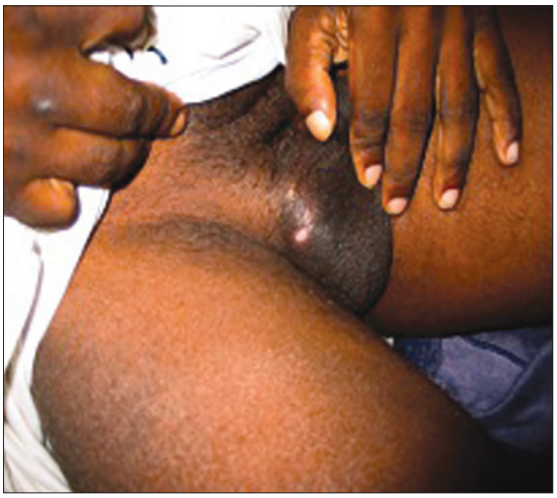

Figure 1: Genital ulceration

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Table 1: International study group criteria for the diagnosis of Behcet's disease (1990) (in the absence of other clinical explanations, patients must fulfill the following criteria)

1	Recurrent oral ulceration (aphthous or herpetiform) observed by the physician or patient recurring at least three times in one 12 months period + two of following
2	Recurrent genital ulceration
3	Eye lesions: anterior uveitis posterior uveitis (cells in the vitreous observed by slit lamp examination); or retinal vasculitis observed by an ophthamomologist
4	Positive skin pathergy test read by a physician at 48 h (2 mm erythematous papule or pustule at prick site 48 h after the application of a sterile hypodermic 20–22 gauge needle which obliquely penetrated avascular antecubital skin to a depth of 5 mm)

(hematocrit – 18) with total WBC count of 8500/cumm. His platelet counts were normal, so were his bleeding and clotting parameters. Peripheral blood smear was suggestive of microcytic hypochromic anemia. Sputum examination by gram stain and Ziehl–Neelsen stains were negative. His serum ANA, ANCA, RA factor, VDRL, and TPHA were reported negative. His chest X-ray PA view showed right hilar mass [Figure 2]. Ultrasound abdomen was normal. Biopsy of the oral ulcer was done which showed spongiosis and features of acute inflammation. A fiber-optic bronchoscopy was done to evaluate for the source of hemoptysis. Bronchoscopy revealed extrinsic compression of right intermediate bronchus however no focus of bleed was found. Contrast CT scan of the chest was done which showed marked thickening of the right pulmonary artery along with aneurysmal dilatation with thrombus *in situ* extending along the descending branch of the left main pulmonary artery proximally [Figure 3]. Pathergy skin test was negative. Ophthalmic examination excluded eye involvement. Since our patient had oral and genital ulceration with features of systemic vasculitis, a diagnosis of Behcet's disease was made. He was started on pulse dexamethasone and cyclophosphamide. He has been clinically stable with no further episodes of hemoptysis after 6 months of treatment. He was subsequently lost for follow up.

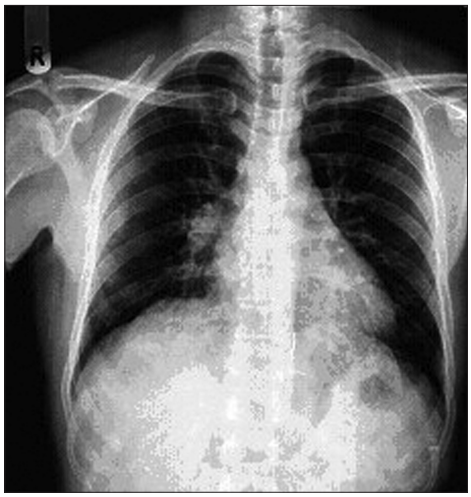

Figure 2: Right hilar mass

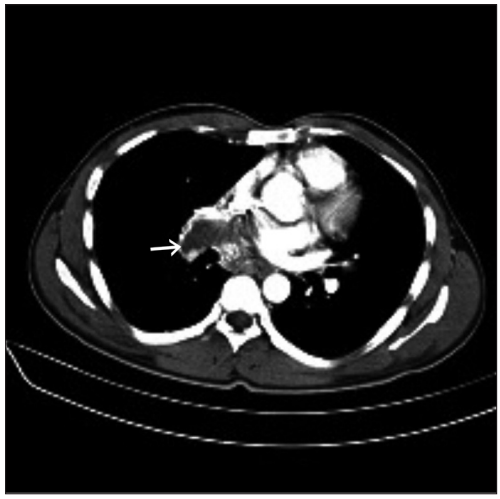

Figure 3: CT Chest showing right pulmonary artery aneurysm with thrombus *in situ*

DISCUSSION

Behcet's syndrome is a rare multisystem disorder of unknown etiology presenting with recurrent oral and/or genital ulcerations, chronic relapsing uveitis which may cause blindness and/or neurological impairments.[1-3] The prevalence ranges from two to thirty per 100 000 populations in Asian countries.[4] There are very few reported cases from India.[5] It generally begins in the third decade of life, but could present at any age. It tends to occur more often in men than in women.[2,3] However, in the Indian patients no such gender predisposition has been observed.[6] BD presents with varied systemic features and pulmonary involvement is one of the features of the disease, which could present as hemorrhage, vasculitis, thrombosis, aneurysm, arterio-venous shunting, etc. In world literature only about 1% of patients have pulmonary artery aneurysmal involvement.[7] Reports of Behcet's disease in India commonly have noted predominantly ''mucocutaneous'' and ''arthritic'' involvement.[6] The pathergy test is rarely positive.[6]

Our patient had oral and genital ulcers, but had negative pathergy test and had no eye or skin involvement. This is not unusual in a patient with pulmonary involvement due to Behcet's disease. In a case series reported by Akpolat *et al*.[8] patients with Behcet's disease, who also had pulmonary manifestation did not fulfill all four criteria required for the diagnosis (ISG criteria are quoted in Table 1).

The pulmonary problems associated with BD are multiple, and these can be classified into three groups:[8-10]

1. Pulmonary artery aneurysm (PAA).
2. Pulmonary parenchymal changes including pulmonary artery occlusion, pleural effusion.
3. Pulmonary obstructive airway disease.

Among all these manifestations of BD, the pulmonary artery aneurysm is the most common as well as most dangerous type. The commonest sites of involvement are the right and left pulmonary artery. It usually affects young male patients.[3,8] The cause of hemoptysis is either rupture of aneurysm or erosion into a bronchus. In our patient, the CT scans revealed the presence of right pulmonary artery aneurysm. There was no evidence of active bleeding at the time of the scan. Another probable cause of hemoptysis is development of the *in situ* thrombus from active vasculitis. Thrombotic aneurysms are the most frequent type of aneurysm in Behcets patients as stated in the literature[11] In our patient, the CT images' appearance showed thrombus *in situ* with features of vasculitis.

Empirical anti-inflammatory and/or immunosuppressive drugs tailored to the severity of the disease remain the mainstay of treatment. A combination of cyclophosphamide and methyl-prednisolone is used most frequently for patients with pulmonary artery aneurysms.[8] Our patient was treated with pulse dexamethasone and cyclophosphamide with which his hemoptysis subsided. He was not started on any anticoagulants.

PAA has a very poor prognosis and is one of the leading causes of death in Behcet's disease.[12] Thirty percent of patients with this condition die within 2 years. Mean survival after the onset of hemoptysis in a patient with Behcet's Disease with PAA was about 10 months in a case series by Ben Taarit *et al.*[12] Our patient continued follow-up with us for 6 months following which he had to move away on account of his job and has not reported for follow up thereafter.

CONCLUSION

In conclusion, in the Indian/Asian context, when a well preserved, young male patient presents with hilar opacity and hemoptysis, apart from the history-taking and evaluation for infective and malignant etiology, it is important to consider the rarer possibility of Behcet's disease if they have other features of the disease such as oro-genital ulcerations.

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