Case Report

Right ventricular thrombus with pulmonary artery aneurysm in a young male: A rare presentation of Behçet’s disease

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ABSTRACT

We describe an adolescent patient presenting with hemoptysis. Detailed clinical work up of the patient showed right ventricular thrombus and bilateral pulmonary artery aneurysms along with the prescribed criteria for the diagnosis of Behçet’s disease. Younger age of the patient was another distinctive feature of this case. Six months of therapy with cyclophosphamide and prednisolone resulted in near complete clinicoradiological response.

KEY WORDS: Behçet’s disease, intracardiac thrombus, pulmonary artery aneurysms

INTRODUCTION

Behçet’s disease (BD) is a multisystem, chronic inflammatory disorder of unknown etiology. First described by Behçet Hulusi in the year 1937 in a patient with distinct picture of oral and genital ulcer with iridocyclitis.[1] Other organ involvement includes skin, joint, central nervous system, gastrointestinal tract, lung and cardiovascular system. This disease is frequent among the Mediterranean, Middle east and Far eastern population.[2]

Cardiopulmonary involvements include wide spectrum of abnormalities. Cardiac manifestations in Behçet’s disease occur in about 1-5% of cases.[3] It includes coronary artery disease, recurrent pericarditis, myocardiopathy, and endocardial abnormalities. Intracardiac thrombus formation is a rare and serious complication. It often occurs in association with pulmonary artery aneurysm (PAA). Young males seem to be most at risk and the right heart is the most frequent site of involvement.[4]

Pulmonary involvement includes abnormalities of the vessel lumen and its wall, lung parenchyma, pleura and mediastinal structures.[5] PAA is reported in 1% of adults with BD. Pulmonary arteries are the second most common site of arterial involvement preceded by the aorta.[6] Aneurysms are more common than thrombosis.[6] Thrombosis of the pulmonary arteries is usually in situ.[7]

Considering the rarity of this disease and its distinct presentation with intracardiac thrombus and pulmonary artery aneurysm, we report this patient.

CASE REPORT

A 17-year-old male presented with hemoptysis of 3 days duration. The patient also complained of low grade fever, exertional dyspnea, decreased appetite and weight loss (7 kg) since last 6 months. Two and a half months back, patient noted pain and redness of both eyes and skin rashes on both thighs which resolved with some form of local treatment in 15 days. Further, on active questioning, the patient revealed recurrent oral and genital ulcers during last 1 year. The patient denied any history of similar illness in family.

General physical examination showed respiratory rate 16 breaths/minute, blood pressure 112/76 mm Hg, and pulse rate 80 beats/minute. Pallor, cyanosis, clubbing and lymphadenopathy were absent.

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Examination of oral cavity [Figure 1] showed multiple oral ulcers. Scars of old ulcers were present on the scrotum.

Respiratory system examination revealed bilateral vesicular breath sounds. There were no added breath sounds including bruit. Ophthalmic examination was also normal.

Routine laboratory tests revealed hemoglobin 11.7 gm/dl, total leukocyte counts 9800/µl with neutrophils 75%, lymphocytes 20%, monocytes 3%, eosinophils 2% and ESR 100 mm in first hour. His fasting blood sugar, renal and hepatic functions were within normal limits. Serum immunological profile e.g. cANCA, pANCA, rheumatoid factor, antinuclear antibody and antibody to HIV were negative. Pathergy test was also negative.

Chest radiograph [Figure 2] showed a rounded poorly defined opacity of approximately 40 × 30 mm size in relation to descending branch of the right pulmonary artery. Rest of lung parenchyma and mediastinum were normal.

Contrast-enhanced computed tomography thorax [Figure 3] showed dilation of both pulmonary arteries containing thrombi. Walls of pulmonary arteries also appeared thickened. Lung window [Figure 4] showed focal area of pleural-based ground glass opacities/consolidation suggestive of pulmonary infarction in both basal lung fields.

CT angiogram thorax [Figure 5] was performed to delineate the details of pulmonary arterial anatomy. It showed dilatation with hypo dense soft tissue filling of the lumen of second order branch of the right descending pulmonary artery and third order branch of the left pulmonary artery. A soft tissue density focus in the right ventricle suggestive of thrombus was also present.
Echocardiography showed a $20 \times 15$ mm mass in right ventricular cavity with no evidence of any structural heart disease. Color Doppler of lower limbs showed normal venous flow.

In our patient, oral ulcer, genital lesions, vascular manifestations (PAA), skin, eye lesions and a negative pathergy test were present during disease duration. A systemic severity score of 7 out of 8 were present in our patient. Positive pathergy test is not necessary in all cases of Behçet’s disease.$^{[8,9]}$

The diagnosis of Behçet’s disease with intracardiac thrombus and pulmonary artery aneurysm was made on the clinical and radiological findings.

The patient was given cyclophosphamide 1 gm along with mesna 600 mg intravenous monthly pulse therapy and tablet prednisolone 30 mg/daily tapered over 1 month to 10 mg daily. After 6 months of therapy, oral and scrotal lesions showed improvement. No fresh skin or eye lesion appeared. Repeat CT pulmonary angiogram showed near normal pulmonary arteries with partial resolution of right ventricular thrombus [Figure 6].

**DISCUSSION**

Present day diagnosis of BD is considered on the basis of newer international criteria for Behçet’s disease (ICBD)$^{[10]}$. This new criteria has additionally incorporated vascular manifestations (VMs) e.g., superficial phlebitis, deep vein thrombosis, large vein thrombosis, arterial thrombosis and aneurysm to the earlier five criteria of international study group.$^{[11]}$

Intracardiac thrombi in BD may result from endomyocardial fibrosis, which may be a sequele of vasculitis involving endocardium, myocardium or both.$^{[11]}$ As intracardiac thrombus is tightly attached to the endocardium, embolism from the cardiac cavity seems to be relatively uncommon.$^{[12]}$

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.$^{[14,13]}$

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 from a tumor and myxoma.$^{[12,14]}$

In a series of 137 patients with BD only one patient was found to have right ventricular thrombus.$^{[15]}$ Similarly, only 1 patient was observed to have intracardiac thrombus out of 56 (1.78%) patients by Uçan et al.$^{[16]}$

Recently, two BD patients with intracardiac thrombi and pulmonary artery aneurysms have been reported.$^{[17,18]}$ Luo et al.$^{[19]}$ analyzed the clinical characteristics of BD with intracardiac thrombus, diagnosed over a period of 1 year.

Pulmonary artery aneurysm (PAA) although rare is more commonly seen than intracardiac thrombi in patient of BD. Various other conditions which can also cause pulmonary artery aneurysm include trauma (often iatrogenic),$^{[20]}$ infections,$^{[21]}$ pulmonary hypertension,$^{[22]}$ congenital heart disease$^{[23]}$ and neoplasm.$^{[24]}$ A detailed history and work up of patients is often sufficient to arrive an etiological diagnosis in a given patient. Helical CT is considered superior to MRI imaging in diagnosis of PAA.$^{[6]}$

In a series of 534 patients with BD, only eight suffered from PAA and six of those died despite immunosuppressive treatment or surgery, underlining
the mortality associated with PAA.\(^\text{11}\) Hamuryudan et al.\(^\text{23}\) reported that 12 of 24 patients (50%) of PAA died after an average of 10 months after the onset of hemoptysis. However, Emad et al.\(^\text{26}\) observed a higher frequency of PAA in 9 out of 16 patients with BD using multislice CT. Such higher frequency of PAA can be attributed to the excellent delineation of vessels lumen and wall on multislice CT scan.

Cyclophosphamide and corticosteroids are the main stay of medical treatment of this life-threatening disease carrying poor prognosis. However, early diagnosis and aggressive therapy result in remission with significant reduction in size of the pulmonary artery aneurysm and partial resolution of right ventricular thrombus. Surgical resection may be considered in patient with massive hemoptysis as a life-saving strategy. The major complication following surgery includes recurrence, false aneurysm and A-V fistula at the site of anastomosis. There are no controlled trial of anticoagulant and thrombolytic agents in BD.\(^\text{15}\) Recently, Piga et al.\(^\text{27}\) successfully used thrombolytic therapy for recurrent right ventricular thrombosis in a patient with BD. Due to the presence of hemoptysis in our case, we did not consider thrombolytic therapy.

### REFERENCES

1. Criteria for diagnosis of Behçet’s disease. International study group for Behçet’s disease. Lancet 1990;335:1078-80.
2. Düzgün N, Küşçükan O, Atasoy KÇ, Topay İşkay C, Gerede DM, Erden A, et al. Behçet’s disease and intracardiac thrombosis: A report of three cases. Case Rep Rheumatol 2013;2013:637015.
3. Wechsler B, Du LT, Kieffer E. Cardiovascular manifestations of Behçet’s disease. Ann Med Interne (Paris) 1999;150:542-54.
4. Mogulkoc N, Burgess MJ, Bishop PW. Intracardiac thrombus in Behçet’s disease: A systematic review. Chest 2000;118:479-87.
5. O’Duffy JD, Carney JA, Deodhar S. Behçet’s disease. Report of 10 cases, 3 with new manifests. Ann Intern Med 1971;75:561-70.
6. Erkan F, Gül A, Tasaş E. Pulmonary manifestations of Behçet’s disease. Thorax 2001;56:572-8.
7. Yilmaz S, Cimen KA. Pulmonary artery aneurysms in Behçet’s disease. Rheumatol Int 2010;30:1401-3.
8. Krause I, Molad Y, Mitrami M, Weinberger A. Pathergy reaction in Behçet’s disease: Lack of correlation with mucocutaneous manifestations and systemic disease expression. Clin Exp Rheumatol 2000;18:71-4.
9. Davatchi F, Shahram F, Chams-Davatchi C, Shams H, Nadji A, Akhlaghi M, et al. Behçet’s disease: From East to West. Clin Rheumatol 2010;29:823-33.
10. International Team for the Revision of the International Criteria for Behçet’s Disease. “Evaluation of the International Criteria for Behçet’s disease (ICBD)”. Clin Exp Rheumatol 2006;24 Suppl 42:S13.
11. Houman M, Ksontini I, Ben Ghribel I, Lamloum M, Braham A, Mnif E, et al. Association of right heart thrombosis, endomyocardial fibrosis, and pulmonary artery aneurysms in Behçet’s disease. Eur J Intern Med 2002;13:455.
12. Erkan F, Esen K, Tunaci A. Pulmonary complications of Behçet’s disease. Clin Chest Med 2002;23:493-503.
13. Vanhalewijk G, el-Ramahi KM, Hazmi M, Sieck JO, Zaman L, Fawzy M. Right atrial, right ventricular and left ventricular thrombi in (incomplete) Behçet’s disease. Eur Heart J 1990;11:957-9.
14. El Houari T, Ouokrell I, Ghzaiel L, Fellat I, Azeroosal M, Serraj K, et al. Management of Behçet disease with multiple complications. Hellenic J Cardiol 2009;50:420-2.
15. Kiraz S, Ercanli I, Ozturk MA, Haznedaroğlu IC, Celik I, Calginener I. Pathological haemostasis and “prothrombotic state” in Behçet’s disease. Thromb Res 2002;105:125-33.
16. Uçan ES, Kiter G, Abadoğlu Ö, Karlikaya C, Akoğlu S, Bayinding Ü. Thoracic manifestations of Behçet’s disease: Reports of the Turkish Authors. Turkish Respiratory Journal 2001;2:39-44.
17. Kaya A, Ertan C, Gürkan OL, Fitoz S, Atasoy C, Kılıçkap M, et al. Behçet’s disease with right ventricle thrombus and bilateral pulmonary artery aneurysms- A case report. Angiology 2004;55:573-5.
18. Düzgün N, Anıl C, Ozer F, Acıcan T. The disappearance of pulmonary artery aneurysms and intracardiac thrombus with immunosuppressive treatment in a patient with Behçet’s disease. Clin Exp Rheumatol 2002;20 Suppl 26:S56-7.
19. Luo L, Ge Y, Liu ZY, Liu YT, Li TS. A report of eight cases of Behçet’s disease with intracardiac thrombus and literatures review. Zhonghua Nei Ke Za Zhi 2011;50:914-7.
20. Rai VK, Malireddy K, Dearmond D, Myers J, Dent DL. Traumatic pseudoaneurysm of the pulmonary artery. J Trauma 2010;69:730.
21. Patankar T, Prasad S, Deshmukh H, Mukherji SK. Fatal hemoptysis caused by ruptured giant Rasmussen’s aneurysm. AJR Am J Roentgenol 2000;174:262-3.
22. Nienaber CA, Spielmann RB, Montz R, Bleifeld W, Mathey DG. Development of pulmonary aneurysm in primary pulmonary hypertension: A case report. Angiology 1986;37:319-24.
23. Tami LF, McElderry MW. Pulmonary artery aneurysm due to severe congenital pulmonary stenosis. Case report and literature review. Angiology 1994;45:383-90.
24. Geddes DM, Kerr IH. Pulmonary arterial aneurysms in association with a right ventricular myxoma. Br J Radiol 1976;49:374-6.
25. Hamuryudan V, Er T, Seyahi E, Akman C, Tüzün H, Fresko I, et al. Pulmonary artery aneurysms in Behçet’s syndrome. Am J Med 2004;117:867-70.
26. Emad Y, Abdel-Razek N, Gheita T, el-Wakd M, el-Gohary T, Samadoni A. Multislice CT pulmonary findings in Behçet’s disease (report of 16 cases). Clin Rheumatol 2007;26:879-84.
27. Piga M, Puchades F, Mayo I, D’Cruz D. Successful thrombolytic therapy for recurrent right ventricular thrombosis in Behçet’s disease. Clin Exp Rheumatol 2010;28 Suppl 60:S56-8.

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