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Thoracic Determination Behçet's Disease

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Abstract

Behçet's disease is a multisystem vasculitis of unknown etiology. Thoracic lesions are dominated by venous and arterial diseases. We identified 21 cases with a hospital prevalence of Behçet's disease between January 1997 and June 2012. The prevalence was of 0.2%. Hemoptysis was the most common symptom and superior vena cava syndrome was the most common sign. Chest Computerized Tomography (CT) scan had confirmed thrombosis of the superior vena cava in 15 patients, and the pulmonary artery aneurysms in 6 patients, three of them were complicated by thrombosis. Patients followed for thrombosis of the superior vena cava received anticoagulant therapy, corticosteroid and immunosuppressive. The combination of corticosteroids, colchicine and immunosuppressive drugs was prescribed in patients with pulmonary aneurysms before surgical treatment. The authors concluded through this study for the rarity and severity of thoracic manifestations of Behçet's disease. Therapeutic means are limited and the prognosis is poor. Thoracic lesions of Behçet's disease are not rare in Mediterranean countries, diagnosis is easy with CT scan, but there's a late in treatment. In these cases, prognosis is conditioned by thoracic lesions.

Keywords: Behçet's disease; Chest; Aneurysms; Thrombosis

Introduction

Behçet's disease (BD) is a multisystem vasculitis that evolves in spurts. It was once part of the great vessels vasculitis, currently it is one of vasculitis vessels of varying size and type [1]. BD is more common in young adults born around the Mediterranean and the Middle East. Thoracic involvement is rare. Chest with include pulmonary infarction, alveolar haemorrhage, thrombosis of the superior vena cava (SVC) and pulmonary aneurysms. Thrombosis of the superior vena cava (SVC) and pulmonary aneurysms are the most common thoracic manifestations and can develop life-threatening. We report 21 cases of thoracic determination in the BD patients hospitalized in ours. Through this study, the authors emphasize the seriousness of these events.

Materials and Methods

This is a retrospective analytical study conducted between January 1997 and June 2012. We identified 21 cases of BD with chest wall involvement. Data were collected through a record operating preset. We are based on the new diagnostic criteria for BD [2] (Table 1). Epi info statistical software Version 6.04 has data analysis. The difference is significant when the P<0,05.

Results

Epidemiological profile

All patients were of Moroccan origin. The average age was 34 years with extremes of 17 and 58 years. Male sex was predominant in 86 % of cases. Females accounted for 14 % of cases.

oral ulceration	**1 point**
genital ulceration	2 points
Skin lesions (pseudofolliculitis, erythema nodosum)	1 point
eye injury	2 points
Vascular involvement (thrombophlebitis, deep vein thrombosis, arterial thrombosis, aneurysm)	1 point
Positive pathergy test	1 point
Diagnosed if ≥ 3 positive points	

Table 1: Criteria for Behçet's disease

Pathergy test: This hypersensitivity reaction is manifested as a pustule or papule that appears after 24-48 intradermal injection of saline. Negative pathergy test: no papule or pustule

Background

Six of our patients were followed and for BD are 29% of cases, the department of internal medicine for skin and eye damage.

Clinical symptoms

Respiratory symptoms were dominated by hemoptysis in 10 cases, with a significant difference, followed by chest pain, dyspnea and dry cough in 3 cases each. Dry cough was found in 2 cases.

Among the physical signs (Table 2), genital ulcers were found in 14 cases with a significant difference (Figure 1), and the upper cave syndrome was observed in 11 cases.

oral ulceration	**21 cases (100%)**

genital ulceration	14 cases (67%)*
Lesions pseudofolliculitis	10 cases (48%)
Erythema nodosum	6 cases (29%)
Syndrome cellar higher	11 cases (52%)
Effusion syndrome	7 cases (33%)
Positive pathergy test	16 cases (76%)*

Table 2: physical signs

*statistically significant value (P< 0,05)

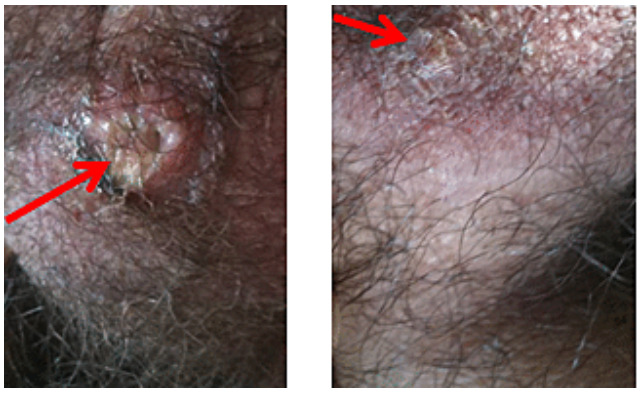

Figure 1: Genital ulcers

Radiological findings

Chest radiography and CT scan were performed in all patients (Figures 2, 3 and 4). CT scan confirmed the diagnosis of thrombosis in 15 cases (71%) and pulmonary aneurysm in 6 cases (29%), the difference is significant. Among these six cases were complicated with two endoluminal artery thrombosis.

Treatment and course

Treatment of thrombosis SVC: Patients with thrombosis of the SVC received treatment with anticoagulants, corticosteroids and immunosuppressants.

Treatment of pulmonary aneurysms: The combination of corticosteroid, colchicine was immunosuppressive and prescribed for patients with pulmonary aneurysms before surgery. Two patients were operated.

Evolution: The evolution was stable in 11 patients. Seven patients were lost to view. One patient had recurrence. Two patients died immediately after the diagnosis of pulmonary aneurysms. The cause of death was massive hemoptysis.

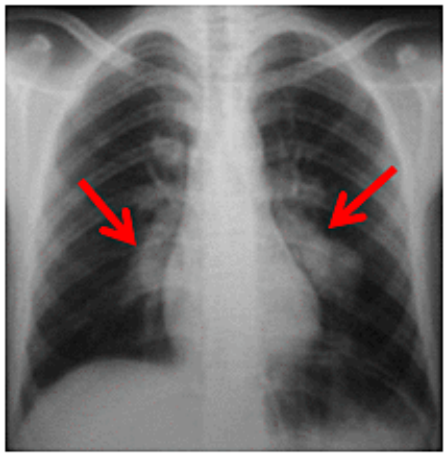

Figure 2: Bilateral hilar opacity

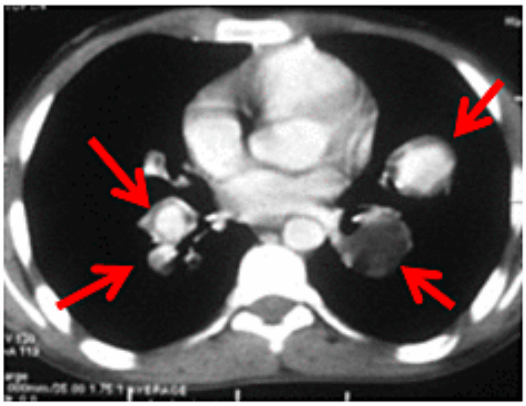

Figure 3: Pulmonary aneurysms

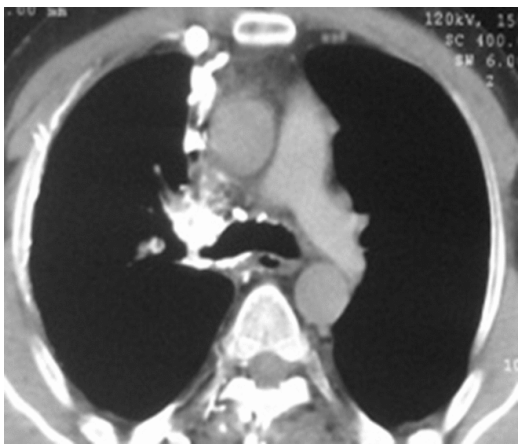

Figure 4: Thrombosis of the superior vena cava

Discussion

The retrospective nature of the study is a limitation of it. However this does not compromise the quality of work, especially as the chest wall involvement of BD is rare.

The BD was first described by Hulusi Behçet times in 1937 involving aphtoses oral, genital ulcers and uveitis [3] (Table 3). It most often affects young patients ranging in age between 20 and 30 years with a male predominance. The young age of our patients and male predominance is consistent with literature data. Currently male dominance is controversial [4]. In Morocco the prevalence of BD was 10 per 10 000 population [5], it varies from one geographic area to another. Hemoptysis and SVC syndrome were predominant in our series. These clinical symptom and sign are not specific to the BD as hemoptysis and SVC syndrome can be seen in the respiratory, infectious or non-tumor pathologies.

Radiological abnormalities whether aneurysmal or thrombotic respectively artery and SVC are not as specific to the BD. Thoracic involvement of the BD is dominated by venous and arterial disease reached as is the case in our series and this is also noted by several authors [6-8]. The frequency of these attacks would vary from 1 to 8% [6-8]. Call symptoms include cough, dyspnea, chest pain and hemoptysis. Hemoptysis can be explained by three mechanisms: a fistula into the bronchial lumen aneurysm, thrombosis and infarction with alveolar hemorrhage [9]. Hemoptysis was the dominant symptom in our series, probably due to the anxiety that patients come in consultation call. In our study, the incidence of thrombosis of the SVC (71%) is higher compared to other countries reported in the mediterranean basin [10] studies.

mandatory criteria	
Recurrent oral ulceration	Minimum 3 episodes in 12 months
other criteria	
Recurrent genital ulcerations	Minimum 3 episodes in 12 months
Eye lesions	uveitis, retinal vasculitis
skin lesions	erythema nodosum, pseudo-folliculitis
Positive pathergy test	read at 24-48 hours

Table 3: Diagnostic criteria for Behçet's disease (International Study Group for Behçet's Disease)

Need for the combination of mandatory criterion at least two other events [3].

The achievement of aneurysmal pulmonary arteries is rare. We collected 6 cases (29%) of pulmonary aneurysms. It is less common than thrombosis of the SVC. They are often multiple and bilateral this is the case in our series. It is most often overlooked, underestimated or under-diagnosed. However it may be indicative of the BD. Hemoptysis is the symptom of pulmonary aneurysms. On one of our patients, the diagnosis was revealed after hemoptysis, despite a year before he presented neurological manifestations of BD to type thrombosis after upper lateral sinus aseptic meningitis. Our prevalence pulmonary aneurysm is less than that of venous thrombosis. And this is also consistent with data from studies [11], but this prevalence is higher than the results of studies of other authors [12]. These are a series in their prevalence is 50 % higher than ours.

Chest CT scan is a reference examination chest with the BD. It allows you to view endovascular arterial and venous thrombosis [13]. Similarly, it strengthens the diagnosis in case of doubt on the vascular nature of mediastinal widening, hilar opacities. The diagnosis of thrombosis SVC based on computed tomography [14].

Chest CT scan allows visualizing the pulmonary aneurysms, counting them, and clarifying their headquarters and their contents. In our series 2 cases of pulmonary arterial aneurysms sit thrombosis. The main headquarters of lesions are the main arteries and lobar [15]. Aneurysms are viewed as saccular or fusiform dilatation is enhancing concomitantly with the pulmonary artery. Their boundaries are blurred when cracking [16].

Angiography is no longer given, due to thrombosis secondary to the injection of contrast and arterial aneurysms puncture sites. The magnetic resonance imaging is necessary in the exploration of mediastinal vessels, however it is less sensitive for the detection of vasculitis of small peripheral arteries and detect lung parenchymal abnormalities [16].

Medical treatment is not specific and often disappointing. It is based empirically on corticosteroids and immunosuppressants (Table 4). The treatment is based on clinical manifestations. The high surgical mortality risk treatment and spares war recurrence.

Insufficient level of evidence for the treatment of major vascular involvement	No controlled trial demonstrated no benefit of anticoagulants, antiplatelet, fibrinolytic in venous or arterial thrombosis
Deep vein thrombosis (DVT): immunosuppressive - Azathioprine - Cyclophosphamide - Corticosteroids - Cyclosporine A	 Risk of fatal hemorrhage when associated aneurysm Response to immunosuppressive
arterial aneurysms - Cyclophosphamide	No preventive effect of recurrences

- And corticosteroids	

Table 4: Dependent therapeutic recommendations lesion site

The functional prognosis is sometimes brought into play in cases of infringement eye and vital after cataclysmic hemoptysis by rupture of aneurysms. Delay in diagnosis fosters this poor prognosis because the therapeutic management of acknowledgment delays. In a series of studies, where the prognosis is far better with 62 % survival at 5 years, although 20% of patients have died during the first year. The authors explain the improved prognosis for a short period of two months on average diagnosis, and treatment consisted of corticosteroids and immunosuppressants for 24 months, both resorted to cyanoacrylate embolization for bleeding who undertake the prognosis [17].

Conclusion

Through this study, thoracic manifestations of Behçet's disease are serious. Thoracic lesions of Behcet's disease are not rare in the Mediterranean countries. The diagnosis is easy with chest CT scan. It lacks therapeutic consensus support. In these cases, the prognosis is conditioned by thoracic lesion.

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