REV BRAS REUMATOL. 2014;xxx(xx):xxx–xxx

REVISTA BRASILEIRA DE REUMATOLOGIA

www.reumatologia.com.br

Case report

Clinical course of Behcet's disease in a patient with delayed diagnosis and radiological follow-up of the thrombi with computed tomography angiography: a five-year follow-up under immunosuppressive treatment

Muhammet Cinar[a,*], *Sedat Yilmaz*[a], *Sinan Akay*[b], *Ugur Bozlar*[b], *Ayhan Dinc*[a]

[a] *Division of Rheumatology, Gulhane Military Medical Academy School of Medicine, Ankara, Turkey*
[b] *Department of Radiology, Gulhane Military Medical Academy School of Medicine, Ankara, Turkey*

ARTICLE INFO

Article history:
Received 23 March 2013
Accepted 12 August 2013
Available online xxx

Keywords:
Behcet's disease
Thrombus
Intracardiac
Superior vena cava
Pulmonary artery

ABSTRACT

Behçet's disease (BD) is a form of chronic systemic vasculitis and almost any organ can be involved. Cardiac and vascular involvement in BD is often associated with poor prognosis. We present a case of a 27-year-old woman diagnosed with BD 9 months after the first symptom appeared. Intracardiac, superior vena cava, and bilateral pulmonary artery thrombi were diagnosed with the aid of echocardiography, cardiac magnetic resonance imaging, and computed tomography angiography (CTA). The patient was treated with immunosuppressive therapy. Then, serial CTA investigations were performed to examine changes in thrombi. Most symptoms except those in the superior vena cava syndrome resolved; CTA showed that thrombi had shrunk but were not completely resolved 5 years later.

Evolução clínica da doença de Behçet em paciente com atraso do diagnóstico e seguimento radiológico dos trombos com angiotomografia computadorizada: seguimento por 5 anos durante tratamento imunossupressor

Palavras-chave:
Doença de Behçet
Trombo
Intracardíaco
Veia cava superior
Artéria pulmonar

RESUMO

A doença de Behçet (DB) é uma forma de vasculite sistêmica crônica e quase qualquer órgão pode ser acometido. O acometimento cardíaco e vascular da DB costuma associar-se a um mau prognóstico. Apresentamos o caso de uma mulher de 27 anos com diagnóstico de DB 9 meses depois que o primeiro sintoma surgiu. Com o auxílio da ecocardiografia, da ressonância magnética cardíaca e da angiotomografia computadorizada (ATC), foram diagnosticados trombos intracardíacos, na veia cava superior e bilateralmente na artéria

* *Corresponding author.*
E-mail: muhcinar@hotmail.com (M. Cinar).
http://dx.doi.org/10.1016/j.rbre.2013.08.004

Please cite this article in press as: Cinar M, et al. Clinical course of Behcet's disease in a patient with delayed diagnosis and radiological follow-up of the thrombi with computed tomography angiography: a five-year follow-up under immunosuppressive treatment. Rev Bras Reumatol. 2014. http://dx.doi.org/10.1016/j.rbre.2013.08.004

pulmonar. A paciente foi tratada com terapia imunossupressora. Depois, foram realizadas investigações por ATC para examinar alterações nos trombos. A maioria dos sintomas, exceto os da síndrome da veia cava superior, foi resolvida; a ATC mostrou que os trombos haviam ficado menores, mas não estavam completamente resolvidos após 5 anos.

Introduction

Behçet's disease (BD) is a chronic inflammatory disease of unknown etiology.[1] The diagnosis is made on the basis of the combination of clinical findings; therefore delay in the diagnosis is not rare. Sometimes, cardiovascular and pulmonary involvement is seen before making diagnosis of BD. Such manifestations can be life-threatening and failure to diagnose BD in such a patient may be very serious.[2–4] In this paper, we describe a BD patient diagnosed late with intracardiac, superior vena cava, and bilateral pulmonary artery thrombi. We used computed tomography angiography (CTA) to study the time course of thrombus development from the time of initial diagnosis throughout treatment. In this aspect, this is the first report to use CTA to explore the long-term course of intracardiac, superior vena cava, and bilateral pulmonary artery thrombi.

Case report

Our patient is a woman who was 27 years at the time of diagnosis, and her first complaint was fever, which commenced in January 2005. Prior to that time, she had suffered from aphthous lesions, but did not seek medical attention. In April 2005, investigations revealed elevated levels of acute phase reactants including the erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) level; these were 38 mm/h and 149 mg/L, respectively. She was admitted to hospital at that time. On physical examination, multiple oral aphthous ulcers were detected. Although BD was considered in differential diagnosis, she was not diagnosed with the disease because other signs of BD were absent. In fact, she did have a genital ulcer but, unfortunately, it was not mentioned to the physician, and the genitalia were not examined. Exhaustive tests seeking the etiology of the fever were conducted; all of infectious, autoimmune, and malign etiologies were considered. Echocardiography revealed a cardiac mass 24 mm × 13 mm in dimensions on the lateral wall of the right ventricular cavity. This was confirmed by cardiac magnetic resonance imaging (MRI); a soft tissue mass 25 mm × 40 mm × 40 mm was evident in the right ventricular cavity. The main features of this soft tissue mass were its partially moving character during systole and diastole, iso-hypointense according to the myocardial tissue and without contrast enhancement. A cardiac thrombus was initially suspected. A biopsy was performed, but it was non-diagnostic. Subsequently, the patient experienced her first episodes of pleuritic chest pain and hemoptysis. Lung ventilation/perfusion scintigraphy was performed; perfusion loss in multiple segments of both lungs was evident. Possible foci of thrombosis and causes of thrombophilia were sought. She was heterozygous for the prothrombin G-A20210 mutation, but neither the V Leiden nor the MTHFR gene was mutated. The activated partial thromboplastin time, and the levels of lupus anticoagulant, protein C, protein S, anti-thrombin III, anticardiolipin antibodies, anti-beta 2-glycoprotein 1 antibodies, and homocysteine were all normal.

One month later, the right ventricular mass was biopsied once more and found to contain only normal heart muscle fibers and adipose tissue. Hemoptysis re-occurred after biopsy and persisted for about 1 week. Pulmonary CTA was then performed, and a hypodense filling defect was evident in the right ventricle and right pulmonary artery (Fig. 1A1 and B1). Therefore, anticoagulant therapy (low-molecular weight heparin followed by warfarin) was commenced.

In September 2005, while still on anticoagulant therapy, the patient was hospitalized with fever, cough, neck and facial swelling, dyspnea, and palpitations. On physical examination, she had fever (38.5 °C), bilateral jugular venous distention, face and neck edema, osteofolliculitis, and erythema nodosum on the right pretibial region. In addition, two genital ulcer scars and oral aphthae were observed. Human leukocyte antigen B51 test was positive and pathergy test was negative, so the diagnosis of BD was made. Pulmonary CTA was performed again. The intracardiac thrombus (ICT) noted earlier remained, but now, dilatations of 2 cm of the ascending and 2.5 cm of the descending branches of the right pulmonary artery were evident, together with a dilatation of 3 cm of the descending branch of the left pulmonary artery. All dilatations were associated with the presence of mural thrombi. Also, neither the right brachiocephalic vein nor the superior vena cava could be visualized because of thrombosis. The clinical findings that developed over the 9-month period prior to subsequent treatment are shown in chronological order (Table 1).

First, warfarin therapy was discontinued because it was possible that both a pulmonary arterial aneurysm and arteritis were present. Methylprednisolone (1 g/day for 3 days) was administered, followed by 1 mg/kg/day of oral prednisolone. A cyclophosphamide (CYC) pulse of 1 g was started and continued monthly thereafter. Prednisolone was tapered 4 weeks later. Symptoms were relieved, and both the CRP level and the ESR fell to normal ranges. Hemoptysis gradually decreased and then disappeared.

In November 2005, the patient was re-evaluated by pulmonary CTA. Thrombi persisted in the intracardiac region, the superior vena cava (Fig. 1D2) and both pulmonary arteries. Multiple collateral intercostal veins, which drain the azygos vein, were serving to drain the upper extremities. A filling defect was observed in the descending branch of the left pulmonary artery (Fig. 1C2). This created a dilatation in the vessel wall, which was associated with minimal intraluminal

Please cite this article in press as: Cinar M, et al. Clinical course of Behcet's disease in a patient with delayed diagnosis and radiological follow-up of the thrombi with computed tomography angiography: a five-year follow-up under immunosuppressive treatment. Rev Bras Reumatol. 2014. http://dx.doi.org/10.1016/j.rbre.2013.08.004