Successful surgical treatment of pulmonary artery aneurysm in Behçet’s syndrome

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Abstract

We report herein an uncommon clinical case of pulmonary artery aneurysm in Behçet’s syndrome, for which only a few reports have been published in the literature to date. A 68-year-old Japanese male, who was referred for recurrent fever of unknown origin, anemia and pulmonary nodular opacities, was treated by right lower lobectomy followed by postoperative oral administration of colchicine and corticosteroid. Postoperative pathological examination confirmed a diagnosis of pulmonary artery aneurysm accompanied by vasculitis and thrombi. Clinical and radiographic feature are presented herein.

Keywords: Pulmonary arteries; Vascular disease; Aneurysm

1. Introduction

Behçet’s syndrome (BS) is a multi-system inflammatory disease of uncertain etiology, currently classified as a vasculitis [1, 2]. BS is characterized by recurrent oral aphthous ulcers, genital ulcers, hyperreactivity of the skin to penetrating trauma, other skin lesions, arthritis, panuveitis, and central nerve system lesions [2]. BS is not a chronic, persistent inflammatory disease, but rather a disease comprising recurrent attacks of acute inflammation. BS affects all sizes of arteries and veins, with large-artery involvement in 1.5–2.2% of all patients with BS [3, 4]. The pulmonary artery is the second-most affected vessel after the aorta and BS is the only vasculitis that causes pulmonary artery aneurysm [2, 4]. Pulmonary artery vasculitis affects mainly young men, presenting with dyspnea, cough, chest pain and hemoptysis. Prognosis in such patients is poor, due to the risk of rupture with fatal hemoptysis [3]. Although surgical intervention with adjunct immunotherapy is considered the best therapy for systemic aneurysms, discussion is ongoing about the role of surgical therapy in the treatment of pulmonary artery aneurysm [5]. We report herein successful surgical treatment of a pulmonary artery aneurysm for which conservative surgical treatment was attempted before the definitive diagnosis of BS.

2. Case report

A 68-year-old man was referred to our institution with recurrent fever of unknown origin and nodular opacity in the lower lobe of the right lung, which was first noted on chest radiography in May 2007. The patient had no history of pulmonary tuberculosis or hemoptysis. Laboratory studies revealed chronic hypochromic anemia, hemoglobin 10.2 g/dl and hematocrit 32.0% and elevation of C-reactive protein level to 9.2 mg/dl on admission to our institution. Repeated flexible bronchoscopy showed extraluminal compression of the right lower bronchus and histological examination revealed no malignancy. Chest computed tomography (CT) enhanced by contrast media revealed dilatation of the pulmonary artery of the right lower lobe, accompanied by thrombi and intracardiac thrombosis in the right ventricle (Fig. 1a,b). Perfusion scintigraphy using technetium demonstrated hypovascularization of the right lower lobe. Exploratory thoracotomy was performed to obtain definitive diagnosis and treat the rapid dilatation of the pulmonary artery. The pulmonary artery aneurysm was localized in the lung parenchyma of right lower lobe. Right lower lobectomy and pulmonary artery plasty were performed for right lower lobe pulmonary artery aneurysm, with an uneventful recovery except for prolonged elevation of C-reactive protein and intermittent fever. The margin between the damaged wall of the pulmonary artery and the intact wall for the arterial suture was approximately 3 mm. Macroscopic appearance of the resected specimens revealed unruptured aneurysm of the lower pulmonary artery, measuring 18×12 mm (Fig. 2a). Microscopic histopathological examination showed intimal thickening and destruction of elastic fibers in the media of the pulmonary artery. The aneurysm wall was infiltrated by polymorphonuclear and mononuclear cells and had penetrated into the adjacent bronchus with adherent thrombus (Fig. 2b). Postoperatively, detailed history-taking elicited the presence

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Fig. 1. Preoperative enhanced CT-scan demonstrated aneurysmal dilatation of right lower pulmonary artery, with thrombus within lumen of aneurysm (a) and thrombus in the right ventricle (black arrow) (b). No aneurysmal dilatation of pulmonary artery was detected in other pulmonary arteries or arterioles.

Fig. 2. (a) Macroscopic appearance of the resected specimens revealed that an unruptured aneurysm of lower pulmonary artery, measuring 18×12 mm. The structure of the pulmonary artery wall was destroyed and replaced by inflamed fibrous tissue. The pulmonary artery aneurysm was filled with thrombus. Right pulmonary artery (black arrow); right lower bronchus (white arrow). (b) Microscopic histopathological examination showed intimal thickening and destruction of the elastic fibers in the media of the pulmonary artery. The aneurysm wall was infiltrated by polymorphonuclear and mononuclear cells and penetrated into the adjacent bronchus with an adherent thrombus (hematoxylin and eosin; original magnification, ×200). Right pulmonary artery (black arrow); right lower bronchus (white arrow).

of recurrent painful oral and genital aphthae and episodes of epididymitis, leading to a diagnosis of BS. In addition, the patient displayed positive results for HLA-B51 allele [2]. Treatment was initiated with colchicine (1 mg/day) and prednisone (40 mg/day) with tapering of prednisone. After initiation of colchicine and prednisone, C-reactive protein fell from 22.1 mg/dl to 0.11 mg/dl and intermittent fever was diminished within a week and the intracardiac thrombus vanished. As of the time of writing, eight months postoperatively, the patient remains well with no evidence of recurrent pulmonary artery aneurysm.

3. Discussion

BS is one of the known acquired syndromes leading to the formation of pulmonary artery aneurysm. The frequency within families is 2–5%, except in Middle Eastern countries, where it is 10–15% [2]. Epidemiologic findings suggest that both genetic and environmental factors contribute to the development of BS. The pulmonary arteries affected in BS reportedly range from the lobular and segmental branches down to the arterioles [3, 5]. The prevalence of the HLA-B51 allele is high among patients with BS who live in areas along the Silk Road (up to 81% of Asian patients have the allele) but not among white patients who live in Western countries (13–70%) [2]. The incidence of HLA-B51 is significantly higher among patients with BS (55–70%) than those without BS (10–19%) [2, 4, 6]. HLA typing may be helpful in the differential diagnosis for BS [2]. This allele affects the severity of disease, since it is more common among patients with posterior uveitis or progressive central nervous system disease than among those with milder disease. However, no previous reports demonstrated that the prevalence of HLA-B51 allele affected the poor prognosis of BS patients with pulmonary artery aneurysm. The prognosis of
BS patients with pulmonary artery aneurysm is serious and hemoptysis is the most common symptom in these patients, with more than half of the patients dying of pulmonary hemorrhage within three years [4, 5]. Hamuryudan et al. reported that the overall survival rate of 62% was five years in BS patients with pulmonary artery aneurysm [4]. Kural-Seyashi et al. reported that death due to pulmonary artery aneurysm had occurred 12 years after the initial diagnosis of BS [7]. Because arterial involvement is a late complication of BS, long-term treatment and follow-up may be required [4]. The underlying pathology is inflammation of the vasa vasorum of the tunica media, causing destruction of the elastic fibers of the media and dilatation of the vessel lumen, which in turn results in aneurysm formation [3, 5]. No randomized controlled studies have evaluated treatment options for pulmonary vasculitis with BS. Colchicine has been recommended as a basic treatment for BS, but a recent controlled trial has again confirmed that this pharmacotherapy is only effective relieving artritis, erythema nodosum and urogenital ulceration [8]. Various treatment modalities in pulmonary aneurysm include immunosuppressive drugs alone or in combination with steroids, surgery, and endovascular embolization [5, 9]. In case of massive hemoptysis, surgery or endovascular embolization is necessary. To avoid pulmonary amputation, aneurysmectomy or occlusion of the arterial defect by direct suture was applied. However, these techniques might not be feasible due to the associated high mortality rates. Massive hemoptysis has been reported as a certain result of suture line rupture and the penetration to the adjacent bronchi [9]. We performed lobectomy and pulmonary artery plasty for resection of the pulmonary artery aneurysm, allowing complete resection of the damaged pulmonary artery wall. This procedure was safe and simple and could be the definitive treatment for a pulmonary artery aneurysm. However, it has been reported that the incidence of recurrent anastomotic aneurysm was 25% after the surgical treatment for pulmonary artery aneurysm [10]. BS is a multi-systemic vasculitis, and colchicum with steroids or systemic immunosuppressive therapy with steroids may be required in the postoperative period to avoid the recurrence of pulmonary artery aneurysm.

References


eComment: Treatment of pulmonary artery aneurysm in Behçet’s disease – few things can make big differences

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We read with the interest report on pulmonary artery aneurysm in Behçet’s disease (BD), presented by Takahama et al. [1]. Pulmonary artery aneurysms are life-threatening complications of BD, frequently reported over the past decades [2, 3]. The advent of computed tomography made diagnosis of these complications readily available. Though cardiovascular involvement in BD is not common, major arterial and venous events are responsible for morbidity and mortality in a significant proportion of these patients [2]. Rupture of pulmonary artery aneurysm leads to death in about 50% of the affected patients [3].

Pathophysiology of BD and its complications are uncertain. The disease has been classified as spondyloarthritides, vasculitides and auto-inflammatory disorders [4]. Neutrophils have been shown to be target cells of inflammation in BD, similar to some others, well-responding to colchicine auto-inflammatory disorders (e.g. familial Mediterranean fever).

The introductory notes and description of the current report [1] prompted us to stress out the following. The authors claimed that BD is not a chronic inflammatory disorder, which contradicts current views on this and other auto-inflammatory disorders, making continuous anti-inflammatory treatment unnecessary. On the contrary, the stable course of inflammation is more likely to cause vasculopathy with destruction of the vasa vasorum and elastic structures of vessels in BD. Hence, monitoring of inflammatory biomarkers and their suppression with steroids and cyclophosphamide would be necessary in the presented case. The choice of colchicine, which is mostly effective for arthritis in BD, is somewhat unexpected.

A search for other locations of vascular destruction (i.e. aneurysms, pseudo-aneurysms, stenosis and thrombosis), and a description of the status of the aorta and the vena cava, in particular, would be useful [2].

Regular radiographic assessment of the vascular system is especially important after the surgical treatment, which itself can mechanically damage other sites of the pulmonary artery as well as other vessels.

The detection of intracardiac thrombus had to necessitate the assessment of tricuspid valve function and pulmonary artery pressure. The latter could allow estimating the efficiency of the procedure. Moreover, the affected site of the lung was removed, but at that stage, the thrombus was still inside the right ventricle, posing a threat of embolisation. It would be important to know what was behind the decision to leave the thrombus.

Finally, BD runs the course of exacerbations and remissions, and complications usually occur when inflammation is out of suppression. With the exception of emergent cases, surgical interventions may be postponed until remission is reached by conservative treatment [2]. As for emergent interventions in BD, they mandatorily should be performed under high dose of immunosuppressive therapy [2].

References