

Isolated Aortic Aneurysm with Aortic Valve Regurgitation in Takayasu Disease

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Received date: September 07, 2017, Accepted date: September 26, 2017, Published date: September 30, 2017

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Abstract

Takayasu arteritis is a chronic inflammatory disease of the aorta, its main branches, and the pulmonary arteries. It is characterized by arterial stenoses and ischemic manifestations.

Aneurysmal dilatation of the ascending thoracic aorta associated with aortic regurgitation is rare in Takayasu disease.

We report the cases of a 43-year-old woman with a Takayasu disease since 18 years, who presented a dyspnea and chest pain. Computed tomography and trans-thoracic echocardiography confirmed an ascending aortic aneurysm associated to a severe aortic insufficiency.

The patient will undergo surgical replacement of the ascending aorta and the aortic valve.

Keywords :

Takayasu's arteritis; Aortitis; Aneurysm; Ascending aorta; Aortic regurgitation

Introduction

Takayasu arteritis is a chronic inflammatory disease predominantly seen in young Asian women, that affects the aorta and its major branches, and less frequently affects the pulmonary arteries [1,2].

It leads to histologic fibrosis of the intima and adventitia and occlusion of the large arteries.

Aneurysm formation and arterial dissection are uncommon complications [1]. We report a severe aortic insufficiency with aneurysmal dilation of the ascending aorta in a 43-year-old woman with Takayasu disease.

Case report

A 43-year-old woman, is treated for Takayasu disease since 18 years. Vascular investigation showed a thickening of the two common carotid arteries and the left subclavian artery, but without anomalies of the aorta and the renal arteries. The patient underwent medical therapy with corticosteroids and immunosuppressive agents. She developed a dyspnea and chest pain. Her physical examination showed a diastolic murmur in the chest wall. Blood pressure was 115/70 mm Hg. The blood tests revealed an inflammatory and microcytic anemia, a chronic inflammatory syndrome with elevated C-reactive protein, and erythrocyte sedimentation rate.

A thoracoabdominal computed tomography-scan showed an ascending aortitis (Figures 1 and 2) (5 mm) aortic wall thickening, with an aneurysmal dilation of the ascending aorta (the diameter of the sinus of Valsalva was 41 mm, and the diameter of the ascending aorta was 43 mm).



Figure 1: Aneurysmal dilation of the ascending aorta.

No abnormality was reported in the duplex scanning of the supra-aortic trunks. Transthoracic echocardiography confirmed a severe aortic valve insufficiency with a tricuspid valve, and without left ventricular dysfunction.



Figure 2: CT scan showing an aneurysm of the aorta.

So, the patient will undergo a surgical replacement of the ascending aorta and of the aortic valve.

Discussion

Takayasu disease or aortitis is a chronic, inflammatory arteriopathic condition of undetermined etiology, that leads to occlusion and ectatic changes in the aorta and its main branches. Clinical presentation is heterogeneous. Early systemic symptoms of Takayasu arteritis may include fever, night sweats, malaise, weight loss, arthralgia, myalgia, and mild anemia [3]. Predominant arterial complications of Takayasu aortitis are stenotic or occlusive lesions. So, it can be manifested by diminished or absent pulses associated with limb claudication, and sometimes hypertension due to renal artery stenosis [3,4]. Isolated aortic aneurysms are rare in Takayasu disease [1]. Although the original reports of Takayasu aortitis are occlusive symptoms, more recent reports have emphasized the increased frequency of aneurysms [5]. Sheikhzadeh et al. [6] described isolated aneurysms in 2% of their patients with Takayasu disease. Matsumura et al. [7] described higher frequency in patients older than 40 years, and mostly within the ascending aorta.

A limited number of reports have been published that describe Takayasu aortitis with aneurysm or dissection of the ascending aorta and aortic valve regurgitation, without obstructive lesions. Tavora F et al. [8] reported a rare case of 57-year-old woman who died by a

ruptured dissection of the ascending aorta into the pericardium. At autopsy, histologic examination of the aortic wall confirmed aortitis of Takayasu. Diagnosis is based on clinical presentation, relevant laboratory findings such as elevated ESR and CRP that may reflect inflammation, and imaging findings of wall thickening and stenosis of medium and large vessels [4]. Imaging of the arterial tree is based on ultrasound, computed tomography, magnetic resonance imaging, or catheter-based angiography showing stenoses of medium and large vessels, and thickened vessel walls. 18-fluorodeoxyglucose (18F-FDG) positron emission tomography may also be helpful for initial diagnosis and monitoring of disease activity [9]. Transthoracic echocardiography detect an associated mitral or aortic valve disease. Aortic regurgitation and aortic or arterial aneurysms left untreated result in greater morbidity and mortality [10].

Glucocorticoid therapy is the primary treatment modality for Takayasu arteritis. Adjunctive, steroid-sparing immunosuppressants such as methotrexate, and azathioprine, are frequently paired with glucocorticoids [11]. Other agents that have been tried include, cyclophosphamide and mycophenolate mofetil [12]. The aim of surgical intervention for aortitis of the ascending aorta should be to correct the anatomic disease problem, to reduce the risk of pseudoaneurysm formation, to do valve replacement for moderate or severe aortic valve disease, and to prevent perivalvular aortic valve leaks. Nevertheless, there are many difficult problems in surgical treatment of this lesion, because of its inflammatory nature, so steroid therapy before and after surgery is therefore vital.

Conclusion

Our paper represents a rare case of Takayasu arteritis with isolated aneurysm of the ascending aorta and aortic valve insufficiency symptomatic with dyspnea.

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