Case report

Isolated supradiaphragmatic descending thoracic aorta stenosis in a Takayasu’s disease: surgical cure

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Abstract

A 21-year-old male patient presented with a typical middle aortic syndrome. Echography disclosed a severe narrowing of the lower thoracic aorta with parietal thickening. The isolated character of the lesion was confirmed by magnetic resonance imaging and aortography. The surgical cure was realized by a Dacron bypass between the upper thoracic descending aorta and the juxta-diaphragmatic thoracic aorta. Aortic biopsy confirmed Takayasu’s disease. Postoperative course was uneventful with normalized blood pressure. The therapeutic options, surgery versus percutaneous dilatation and stent, are discussed. © 2001 Elsevier Science B.V. All rights reserved.

Keywords: Takayasu arteritis; Middle aortic syndrome; Arterial reconstruction

1. Introduction

Takayasu’s disease involving the lower thoracic aorta is rare, especially as an isolated lesion [1]. A new case is presented.

2. Case report

A 21-year-old male patient was referred for evaluation after a routine checkup disclosed a systemic arterial hypertension. He complained of vague, not exercise-related, thoracic pains and of some excessive fatigability of the lower limbs. Four years previously, he had been treated for 3 months for supposed pulmonary tuberculosis without bacteriological confirmation.

At examination the patient was in good general condition. Blood pressure was 180/100 mmHg in both upper limbs. There was no precordial murmur. At the level of the left costo-vertebral sinus, a loud systolo-diastolic murmur was heard. Both femoral pulses were present, but weak. No abnormal collateral circulation was palpated.

Routine laboratory work including C-reactive protein and erythrocyte sedimentation rate was normal. EKG showed left ventricular hypertrophy. Chest X-ray was normal.

Transthoracic echography and transesophageal echography (TEE) confirmed concentric left ventricular hypertrophy without aortic valve or ascending aorta anomalies, and showed a significant stenosis of the juxta-diaphragmatic descending thoracic aorta. At this level a mean pressure gradient was evaluated at 50 mmHg. The aortic wall was grossly thickened, but not calcified.

A complete exploration of the aorta and its branches by magnetic resonance imaging (Fig. 1) and aortography (Fig. 2) determined the characteristics of the lesion. Aortic arch, supracervical branches, abdominal aorta and its main branches were normal. The descending aorta wall was thickened from its middle portion; in the distal third, a fusiform 3.5-cm long stenosis was present; immediate juxta-diaphragmatic aorta appeared normal. Two important intercostal arteries originated at or near the level of the stenosis. The Adamkiewicz’s artery could not be identified. The pressure gradient was confirmed invasively.

At operation, the descending thoracic aorta was controlled through a left 8th intercostal thoracotomy. A thrill was palpated at the T8–T9 level; at this site the aorta external caliber was normal, but its wall was thickened with a fibrotic adventitia. Juxta-isthmic aorta and the last few centimeters of the terminal descending thoracic aorta appeared normal.

Under lateral cross-clamping, a Dacron bypass was performed between the post-isthmic and the terminal descending thoracic aorta. At the site of the implantations, aortic wall was normal except at the lower end of the upper...
arteriotomy where a biopsy was performed and confirmed Takayasu’s disease.

The postoperative course was uneventful. Control TEE showed no residual gradient and control arteriography a patent bypass. At a 6-month check-up, blood pressure was normal.

3. Discussion

The stenotic lesions lying within the aortic segments II–IV are defined either as ‘atypical’ coarctation or as ‘middle aortic syndrome’. Atypical coarctation applies more adequately to hypoplastic types classified as congenital defects. These hypoplasias are rare: 0.5–1.5% in the series of patients with coarctation [2]. Middle aortic syndrome was introduced by Sen et al. [3] for inflammatory subisthmic aortic narrowings. Inflammatory diseases such as Takayasu’s arteritis, aortoarteritis and granulomatous arteritis are indeed the most common causes of subisthmic stenoses [1].

The common clinical picture [3] associates an age below 40 years, upper extremity arterial hypertension, weak femoral pulses and a systolic bruit in the low interscapular or abdominal area. Various ischemic symptoms, including abdominal angina, can be present, but severe arterial hypertension is the main justification for an aggressive therapeutic attitude [4].

Our observation demonstrates that transthoracic and transesophageal echography can adequately localize and evaluate supradiaphragmatic aortic stenoses. Magnetic resonance angiography provides anatomical information and can assess the functional obstruction; however, conventional aortography is still necessary as detailed information about the collateral circulation is required [1]. The supradiaphragmatic narrowing can involve the Adamkiewicz’s artery which should be visualized.

Since Sen’s initial report, middle aortic syndrome series generally report diffuse stenotic or aneurysmal lesions of the thoraco-abdominal aorta and its visceral branches [1]. Suspected Takayasu’s disease deserves a complete vascular
exploration including aorta, its main branches, and the pulmonary artery. In the inflammatory phase, corticosteroids and antiplatelet drugs are classically recommended and surgery is usually avoided because of a high incidence of complications [5,6].

Percutaneous transluminal angioplasty (PTA) and wall stent have been used in Takayasu’s arteritis [7,8]. The mechanism of successful angioplasty is quite unclear as Takayasu’s lesions, panarteritis, diffuse inflammation and periarterial fibrosis differ totally from atherosclerotic pathology. Thrombosis and aortic dissection are the principal risks of PTA; incomplete results are frequent as vessel walls do not easily yield to mechanical distension. Long segment lesions can necessitate multiple stents. According to Sharma et al. [9] “angioplasty of thoracic aortic stenosis can be hazardous in the presence of diffuse underlying disease” and it is usually the case in Takayasu’s disease. Finally, any type of direct intervention to the narrowing may compromise the blood supply to the spinal cord if the Adamkiewicz’s artery originates from this segment [1].

If a simple descending to descending aorta bypass was acceptable in our case, surgery often requires an extensive thoracoabdominal graft [5,6,10]. The operative risk is very low in elective operations [6]. Over the long term, surgery can be considered as generally curative. Yet Tada et al. [5] report an 11% incidence of anastomotic aneurysms in 93 patients followed up to 22 years. To avoid this complication, a few controversial recommendations [5,6,10] have been done: operate in the ‘burnt out’ phase of the disease, avoid suturing in a pathological wall and, if not avoidable, use large bites to make the anastomoses, and avoid post-operative corticotherapy. Associated renal arterial stenoses should always be corrected; if not, the prognosis of hypertension is poor [5].

In the presented case, ascending aorta to descending aorta bypass graft could have been another technical choice. This technique should be considered if redo surgery is judged to be necessary.

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References