Resection and Graft Replacement of Thoracoabdominal Aortic Aneurysm in a Patient with Takayasu’s Disease


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A 39 year old female patient with Takayasu’s disease developed general symptoms of muscular pain, fatigue, general malaise, fever and periods of fainting. She also had an increased erythrocyte sedimentation rate, C-reactive protein and increased gammaglobulin levels. She had received corticosteroid therapy for several years and had developed a thoracoabdominal aortic aneurysm. In 1998 the maximal aneurysmal diameter had increased to 10.2 cm. Using atriofemoral bypass, resection and graft replacement of the thoracoabdominal aneurysm was performed. Six years after the operation the patient is well, working part time as a primary school assistant.

Although, thoracoabdominal aneurysm due to Takayasu’s disease is rare, the condition must be taken into consideration in female patients younger than 40 years of age who have vague general symptoms and increased ESR of unknown cause. The vascular system should then be thoroughly investigated for arterial pathology. Thoracoabdominal aortic resection and graft replacement can be performed in these patients using standard techniques.

Keywords: Takayasu’s disease; Thoracoabdominal aneurysm; Graft replacement.

Introduction

Arterial obstruction due to Takayasu’s disease has a low incidence in the Scandinavian countries. The aneurysmal form of this disease is even more seldom seen and of these only a few cases with thoracoabdominal aortic aneurysms have been reported in the literature. Takayasu’s disease is usually classified from I to V according to the nature and location of the disease. Type V defines the aneurysmal form. Although this disease is rare, it should be taken into consideration in women below 40 years of age presenting with vague symptoms in combination with an increased erythrocyte sedimentation rate (ESR) or C-reactive protein. The intention of this report is to describe a patient with Takayasu’s disease and a thoracoabdominal aortic aneurysm who underwent successful surgical repair.

Patient Characteristics

The patient was 39 years of age at the time of operation in 1998. She had presented with vague symptoms including muscular pain, fatigue, general malaise, fever and periods of fainting. The symptoms started in connection with her first pregnancy and delivery, which was rather traumatic, and she had symptoms for about 10 years before the diagnosis was confirmed. At the age of 32 she delivered her third child and at that time an elevated ESR was confirmed. After delivery by caesarian section she developed signs of endometritis, which was treated with antibiotics. However, she still experienced periods of fever, chest pain and fatigue and was admitted to our hospital for further investigation in 1991. At that point the ESR was 89–78 and C-reactive protein and gammaglobulin levels were elevated. Repeated blood cultures were
negative, and serological tests showed no signs of viral infection. Tests for autoantibodies were performed and were negative including antinuclear antibodies (ANA), anticitrullinated antibodies (ACA) and antineutrophil cytoplasmatic autoantibodies (ANCA). Serological tests for possible infections including anti-HIV antibodies, anti-Hepatitis B antibodies and tests for syphilis were negative. A CT-scan demonstrated aneurysmal dilatation of the descending thoracic aorta and most of the abdominal aorta in accordance with a type I thoracoabdominal aortic aneurysm. There were no signs of aortic dissection at CT, MRI or transthoracic echocardiography. The maximum diameter of the aneurysm was 60 mm. The peripheral pulses were palpable and there were no signs of arterial obstruction. Coronary angiography was not performed since the patient did not have any signs or symptoms of cardiac disease. Dobutamine stress echocardiography and an exercise ECG were both normal. Taking into consideration the age of the patient, the disease history, the elevated inflammatory parameters and the angiographic abnormalities, we concluded that she had a Takayasu’s arteritis type V (aneurysmatic type). The patient received corticosteroid treatment with an initial prednisolone dose of 60 mg daily, gradually decreasing the dose. No other immunosuppressive drugs were used. After a relatively short period her clinical condition improved. She had some side-effects of the corticosteroid medication, mainly characterised by increased body weight and Cushing-like appearance. She still experienced episodic malaise, fatigue, periods of fever, headache and shortness of breath indicating that the disease remained active. In 1998 the maximum diameter of the aneurysm had increased to 10.2 cm on CT as well as on preoperative arteriography (Figs. 1 and 2). She also developed chest pain. She was therefore operated on via a thoracolaparotomy continuing a midline abdominal incision into the 4th intercostal space. The 4th rib was resected. The left atrium was cannulated at the level of the pulmonary veins and aortic bypass was established using a Biomedicus® pump. The arterial cannula was placed in the right femoral artery. An autotransfusion system was used during the operation. The proximal part of the thoracic aorta was double-clamped allowing the upper anastomosis to be performed while the distal part of the aorta and its sidebranches was perfused with blood. A 22 mm vascular graft (Gelweave®, Vascutek Ltd. Inchinnan, Scotland) with a sidebranch was inserted and the proximal anastomosis performed in an end-to-end fashion. A vascular clamp was then placed on the distal abdominal aorta and the aneurysmal sac was opened. The orifices of the coeliac axis, superior mesenteric artery and right renal artery were left on a patch of aortic tissue. The distal part of the vascular graft was cut in an oblique fashion and the distal anastomosis included the orifices of these three arteries. The intercostal arteries were oversewn. Since the distance between the right and left renal arteries was too wide to include them both into the distal anastomosis, the left renal artery was revascularised by an end-to-end anastomosis to the sidebranch of the main graft (Fig. 3). The total perfusion time was 78 min, the operating time 5 h and the total blood loss approximately 4 l. On the day of operation she received 2700 ml of blood, 2000 ml of fresh-frozen plasma, 1000 ml of hydroxy-ethyl-starch and 400 ml of platelets. The postoperative course was uneventful and the patient was discharged to her home on the 13th postoperative day. Microscopic examination of tissue from the aneurysmatic aortic wall demonstrated atherosclerotic plaques and thrombus material, thinning of the aortic wall as well as partial destruction of the media with fibrous replacement, but no active inflammation in the media. In the adventitia chronic inflammation and fibrosis was demonstrated, in addition to scattered groups of histiocytes forming granulomas without necrosis (Fig. 4). From the time of the operation the steroid medication was gradually reduced and it was discontinued 4 months later. She has been followed up at regular intervals at the outpatient clinic and 6 years after the operation she is largely asymptomatic with the exception of a tendency to fatigue and sometimes headache. She is working part-time as a primary school assistant. Annual MR angiography has been performed showing a patent reconstruction without arterial dilatation or obstruction. There are no signs of other vascular involvement.

Discussion

In our series of operated aortic aneurysms during a 20-year period, which now includes more than 1200 patients and about 120 descending thoracic-or thoracoabdominal aneurysms, this is the only one who had aneurysmal formation due to Takayasu’s disease. Some patients with obstructive disease, mainly of the brachiocephalic arteries, due to Takayasu’s arteritis have been described from our health region. We consider the presenting symptoms and clinical findings in this patient to be clearly compatible with the diagnosis of Takayasu’s arteritis. She had a history of constitutional symptoms with fever, general malaise and muscular pain. There were no signs of infection, and no serological or clinical findings suggestive of
other vasculitides, systemic lupus or rheumatoid arthritis. The histopathological changes were also suggestive of the diagnosis, but the biopsy was taken 8 years following suspicion of the diagnosis and start of corticosteroid treatment. At that time there were no signs of active inflammation, which is in accordance with previous publications.8,9

It was decided to treat the patient with corticosteroid treatment to control the active phase of the disease and to postpone surgery to a later stage when there would be less inflamed tissue in the aortic wall.11 There could have been a risk that further aneurysmal dilatation and rupture might have occurred12 and this represented a dilemma. However, we had close contact with the patient and when she developed chest pain she was operated on without further delay. The technique of leaving the coeliac axis, superior mesenteric artery and right renal artery on a patch of aortic tissue is an issue since one cannot rule out the presence of arteritis in the aorta near these orifices. However, as little of the aorta as possible was included in the anastomosis. Furthermore, it was felt of importance to make the operation as simple as possible. Finally, the distal part of the abdominal aorta, which had a near-normal diameter was not replaced to minimise aortic resection and probably reduce the risk of neurologic complications.7 Because of the relatively long distance between the two renal arteries the sidearm of the main graft was anastomosed to the left renal artery. Postoperative

Fig. 1. Arteriogram indicating an aneurysm starting in the proximal descending thoracic aorta.

Fig. 2. Distal part of the arteriogram shown in Fig. 1. The thoracoabdominal aorta including the coeliac axis, superior mesenteric artery and both renal arteries is affected by the aneurysmal dilatation. Only the distal part of the abdominal aorta has a near-normal diameter.

Fig. 3. Schematic illustration indicating the technique of operation using atriofemoral bypass. A separate bypass was made to the left renal artery using a sidearm of the main graft.
MR-angiography has shown that the circulation to both kidneys, the coeliac axis and superior mesenteric artery is satisfactory and that the vascular graft with its sidearm is patent. There is no further aneurysmal degeneration or signs of pseudoaneurysm, which has been reported in other series of patients operated on for aneurysms due to Takayasu’s disease.2,13

In conclusion, although thoracoabdominal aneurysm due to Takayasu’s disease is rare, the condition must be considered in female patients younger than 40 years of age who have vague general symptoms and an increased ESR of unknown cause. The vascular system should then be thoroughly evaluated for arterial obstruction or aneurysmal dilatation.4,5,14 Thoracoabdominal resection and graft replacement can be performed in these patients using standard techniques.1

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