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Takayasu Arteritis – A Case Report of Aortic Aneurysm

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ABSTRACT

Aortic pseudo-aneurysm is a rare manifestation of Takayasu arteritis. We present a 16-year-old girl who first complained of multiple arthritis, recurrent abdominal pain and malaise at the age of 15 years. The initial working diagnosis was juvenile rheumatoid arthritis. Follow-up abdominal ultrasonography for her hepatomegaly incidentally revealed an aortic aneurysm. Total aortography showed diffuse aortic narrowing and an infra-renal aortic pseudo-aneurysm. Vascular reconstruction with an interposition Dacron graft was performed with uneventful recovery. Early non-specific presentation of Takayasu arteritis often results in delay of diagnosis. The presence of a vascular bruit in a young female with non-specific symptoms should point to a differential diagnosis of Takayasu arteritis. We review the role of surgery in the management of this condition.

Keywords: Takayasu arteritis, aortic aneurysm, surgical bypass, diagnosis, pseudo-aneurysm

INTRODUCTION

Takayasu arteritis is a chronic inflammatory disease that primarily affects large vessels, aorta and its major branches. It was named after Takayasu, a Japanese ophthalmologist, who reported the ophthalmoscopic findings of retinal arteriovenous shunts and ocular ischaemia around the optic disc in a 19-year-old Japanese female in 1908[1]. This peculiar ocular finding and pulselessness of upper extremity were subsequently described in a number of reports. The etiology of this condition remains unknown. It is typically a disease of young oriental women, who commonly suffer from chronic ischaemia to viscera especially the brain, kidneys and heart. We report here a case of aortic pseudo-aneurysm involving infra-renal abdominal aorta with stenosis of visceral arteries.

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CASE REPORT

A 16-year-old Chinese girl presented to a paediatrician with a six-month history of recurrent abdominal pain, malaise, weight loss, as well as bilateral ankle pain and swellings in January 1995. Physical examination at that time revealed shotty neck nodes, erythema nodosum on the shins and bilateral ankle arthritis. An abdominal bruit was described on one occasion. Blood tests showed an elevated erythrocyte sedimentation rate of 60 mm/hr, mild leukocytosis (WBC 13.4 x 10^9/L) and a haemoglobin level of 13 g/dL. Serology screening revealed negative anti-DNA and rheumatoid factor, normal C3, C4 levels, normal immunoglobulins pattern and weakly positive antinuclear antibody (1:40, speckled). Abdominal ultrasonography showed hepato-splenomegaly. The working diagnosis was juvenile rheumatoid arthritis. She was then treated with non-steroidal anti-inflammatory drugs (NSAIDs) and penicillin. The symptoms subsided within three months and she defaulted follow-up.

The patient was admitted to a regional hospital again in August 1996 because of night sweats, cough and fever for two weeks. Physical examination showed erythema nodosum over both shins. Chest X-ray revealed right hilar shadow. Computed tomography of the thorax showed multiple hilar lymphadenopathy with right middle lobe consolidation. Although culture of sputum for acid-fast bacilli was negative, the patient was treated with anti-tuberculous medications. Follow-up ultrasonography of abdomen for her hepatomegaly incidentally revealed an abdominal aortic aneurysm, which was later confirmed by contrast computed tomography. Total aortography revealed diffuse narrowing in lower thoraco-abdominal aorta with an infra-renal pseudo-aneurysm, measuring 2.4 cm in diameter (Fig 1). The origins of the coeliac and superior mesenteric arteries were also stenosed. The aortic arch and its primary branches were spared. These features were compatible with the diagnosis of Takayasu arteritis.

She was then referred to our unit for further management. On questioning, she had intermittent abdominal pain for 2 years with gradual weight loss of 10 lbs. Physical examination showed a bruit over the epigastrium. All the peripheral pulses were palpable. Blood pressures were normal and equal on both arms. Examination of other systems were unremarkable.

Vascular reconstruction with an infra-renal aortic interposition Dacron graft and a Dacron bypass limb to the superior mesenteric artery was performed on 19 March 1997 (Fig 2). Matted lymph nodes were found encasing the coeliac axis, making access to the coeliac artery hazardous. Infra-renal aorta was approached by an infra-colic transperitoneal approach and a retroduodenal tunnel was created for the bypass graft to the superior mesenteric artery in a reversed C loop fashion. During operation, a pseudo-aneurysm of 2.5cm diameter was found over the left antero-

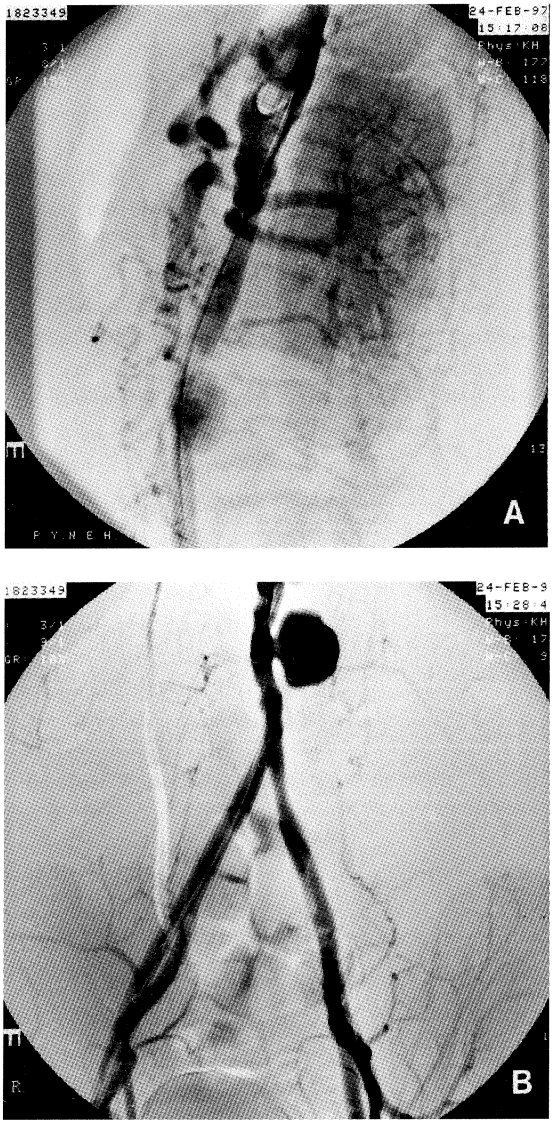

Fig 1 – Aortograms showing diffuse narrowing of abdominal aorta with stenotic origins of coeliac and superior mesenteric arteries (A) and an infra-renal aortic pseudo-aneurysm (B) of 2.4 cm diameter arising from the left lateral aspect of abdominal aorta at level of L3.

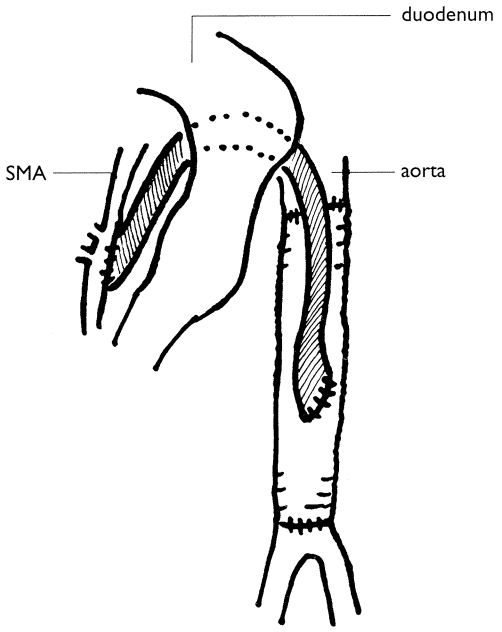

Fig 2 – Illustration of the vascular reconstruction with an aortic interposition Dacron graft and a bypass Dacron graft to superior mesenteric artery through a retro-duodenal tunnel.

lateral aspect of infra-renal aorta, with an aortic orifice of 1cm wide. The pseudo-aneurysm was surrounded by extensive fibrosis, making biopsy of the false aneurysm impossible. Aortic wall was grossly thickened with narrowed lumen. Histopathological examination of the aortic wall biopsy demonstrated segmental thickening of the intima by myxoid fibrous tissue. The elastic fibres and muscle cells in the media were segmentally destroyed. The adventitia was markedly fibrotic with mild lymphocytic infiltration. The features were consistent with that of 'burned out' Takayasu arteritis with pseudo-aneurysm formation. The operative time was five hours and intra-operative blood loss was 0.7 L. The patient was nursed in the intensive care unit after operation and her post-operative course was uneventful. She recovered uneventfully and was discharged on the eighth post-operative day. During follow-up, the patient remained well and stable. A follow-up duplex scan demonstrated patency of the bypass graft.

DISCUSSION

Takayasu arteritis commonly afflicts young Oriental females. Most patients with Takayasu arteritis present with 2 stages of the disease process: acute inflammatory (pre-pulseless) stage followed by a chronic stage[2]. The acute stage of the disease is mainly characterised by non-specific inflammatory constitutional symptoms, such as fever, malaise, arthralgia, myalgia and erythema nodosum. This presentation usually precedes the second stage by a few months to a few years. The manifestations of the chronic stage result from gradual sclerotic stenosis of the involved arteries, leading to arterial insufficiency of the extremities or end organ.

The early non-specific presentation of Takayasu arteritis often causes delay of the correct diagnosis. This problem is particularly prominent in juvenile patients[3]. This patient was initially treated for seronegative juvenile rheumatoid arthritis, a similar finding observed in other previous series. Increased ESR and anaemia are common findings during the active stage of disease. However, laboratory investigations are often non-specific and unhelpful to diagnosis.

The pathologic features of active Takayasu arteritis are granulomatous panarteritis, characteristically with a heavy cellular infiltration of the media and adventitia by lymphocytes, plasma cells and giant cells[4]. Nonetheless, biopsy is rarely obtained during this period. The pathological findings from aortic biopsy in the current patient showed sclerosing arteritis, characterised by fibrous intimal hyperplasia, medial degeneration and adventitial fibrosis. It indicated that the present patient was in chronic 'burned out' stage. Gradual fibrosis and thickening of the walls of blood vessels led to formation of stenotic lesions with ischaemia of end-organs. Pseudo-aneurysm is a rare manifestation of Takayasu arteritis[5]. When there is deficient production of supporting fibrous tissue in the presence of extensive destruction of media during the acute inflammatory stage, the weakened aortic wall

may perforate to form a pseudoaneurysm.

Takayasu arteritis cannot be confidently diagnosed by biopsy alone, especially at this chronic phase of the disease. Clinical suspicion of Takayasu arteritis is therefore mainly dependent on clinical criteria, as suggested by the American College of Rheumatology in 1990 (Table I)[6]. To confirm the diagnosis, total aortography is indispensable because of the protean clinical manifestations of Takayasu arteritis[7]. The characteristic aortographic fndings are diffuse narrowing of the aorta and severe stenosis of the origins of the major branches. It also delineates the extent of the disease process and allows follow-up of the progression of the disease process.

Table I – Diagnostic criteria of Takayasu arteritis suggested by American College of Rheumatology[5]

1.	age < 40
2.	claudication of extremity
3.	diminished brachial artery pulse
4.	> 10 mmHg difference in SBP between arms
5.	bruit over aorta or subclavian artery
6.	aortography showed narrowing or occlusion of aorta or its primary branches

Presence of 3 or more of these 6 criteria gives a sensitivity of more than 90%

Common causes of death include stroke, rupture of aneurysm, heart failure and renal failure secondary to renovascular hypertension. Aneurysmal formation in the infra-renal aorta is rare. Robbs et al[8] reported the largest series of patients (n = 18) undergoing vascular reconstruction for infra-renal aorto-iliac aneurysm. There was no hospital mortality following elective reconstruction but 2 patients died after emergency operative procedures for ruptured aneurysm. Regular surveillance for this complication with early surgical intervention may be able to avoid unexpected early mortality. Other indications for surgery comprise renovascular hypertension, incapacitating claudication, extra-cranial cerebrovascular insufficiency and ischaemic heart disease[9]. It is imperative to avoid operation during active disease as anastomotic aneurysm, pseudoaneurysms and post-operative haemorrhage are prone to develop at anastomosis[10]. It has been suggested to schedule the operation when the disease is quiescent with ESR < 15mm in the first hour. With recent advances in vascular surgery, most patients have satisfactory results from vascular reconstruction and the long term patency rates of bypass grafts are satisfactory[11,12].

This patient had 2 surgical conditions, viz. abdominal aortic pseudo-aneurysm and chronic mesenteric ischaemia, that required operative intervention to prevent rupture and catastrophic bowel infarction respectively. Antegrade bypass to both coeliac and superior mesenteric arteries was our initial plan[13]. Unfortunately, the presence of matted coeliac lymph nodes made the dissection of coeliac artery too hazardous. Retrograde bypass to the superior mesenteric artery alone was therefore performed. The Dacron graft was looped through a retro-duodenal tunnel so as to avoid kinking. Isolated bypass to superior mesenteric artery had been demonstrated to produce a comparable patency and recurrence rates to extensive revascularisation of multiple vessels[14]. Percutaneous transluminal angioplasty is an alternative treatment for focal stenotic arterial lesions[15]. Owing to the high incidence of restenosis, its role and long term result remain to be proven.

The management of this patient requires a multidisciplinary approach involving the rheumatologist, vascular surgeon and pediatrician. Regular follow-up is necessary to look for progression of stenosis of visceral arteries and recurrence of symptoms. The primary medical therapy of Takayasu arteritis is immunosuppressive treatment with glucocorticoids. Significant palliation of symptoms and successful induction of remission have been reported to follow its use[1]. The real benefit of corticosteroids in Takayasu arteritis has not been well proven because of conflicting reports on its efficacy. Above all, serious side effects, like poor wound healing and susceptibility to infection, have significant implications. Immunosuppressive treatment would only be considered in this patient in case of relapse.

The clinical finding of a vascular bruit in a young woman who complained of non-specific systemic symptoms should alert clinicians to the possible diagnosis of Takayasu arteritis. Increased awareness can lead to early recognition of this condition.

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