Extensive Myocardial Infarction in a Child with Takayasu Vasculitis: Report of a Case and Literature Review

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Abstract

Objective: Takayasu arteritis (TA) is an inflammatory arteritis involving large vessels, predominantly the aorta and its main branches. Angina pectoris or myocardial infarction may occur in 3-5% of patients due to coronary artery ostial narrowing from aortitis or coronary arteritis.

Case presentation: We describe the case of an 11-year boy presented with hypertension and severe abdominal pain. After treatment he developed extensive myocardial infarction and died.

Conclusion: Takayasu’s disease should figure prominently amongst the causes of coronary artery disease in children and coronary arteriography should be undertaken more often during investigation of the arterial lesions of these patients.

Key Words: Takayasu; Vasculitis; Myocardial infarction; Coronary arteritis

Introduction

Takayasu arteritis is a chronic vasculitis of unknown etiology. Worldwide, Takayasu arteritis is the third most common vasculitis of childhood with the greatest prevalence in Asians¹, but it is relatively uncommon in Europe and North America. About 150 new cases per year are witnessed in Japan²; while it has been estimated that only 1 to 3 new cases per million population occur in the United States and Europe³.

The aorta and its primary branches are primarily affected by Takayasu arteritis. Although the inflammation could involve entire vessels, it may also be localized to a portion of the thoracic or abdominal aorta and branches. Myocardial infarction may occur...
because of angina pectoris which may be caused by coronary artery ostial narrowing from aortitis or coronary arteritis. Coronary artery's involvement in Takayasu's disease may occur approximately in 9-10% of cases and appears to be relatively unappreciated[4]. In this paper we report a boy with Takayasu and extensive myocardial infarction.

**Case Presentation**

An 11-year old boy suffering from severe abdominal pain, flashing and palpitation since twenty days before admission was admitted to Pediatric Nephrology ward with high blood pressure. Hypertension was found in the patient during evaluation of abdominal pain. In his medical history, he had developmental delay (his speaking was impaired), was hospitalized in the age of 9 months and 5 years because of pneumonia. His mother told us about his exertional dyspnea since 4-5 years ago.

In physical examination he looked smaller than his real age, he had mild mental retardation and his communication was not good. Blood pressure measured in his four extremities was:

- Right arm: 150/70 mmHg; Left arm: Undetectable; Right leg: 190/100 mmHg; Left leg: 160/90 mmHg.

Other features in physical examination contain: high jugular vein pressure, heart murmur (II/VI) in left sternal border, and a palpable spleen 3-4 cm under rib border. No abdominal vascular bruist was heard in physical examination. Pulses in the right upper extremities were normal, in the left side, radial and brachial pulse was not detectable and left axillary pulse was weak.

**Routine laboratory tests:** Cell blood count (CBC): leukocytes 12300 (PMN 61%), Hemoglobin 12.5mg/dl, platelets 475000, erythrocyte sedimentation rate (ESR) 60, CRP negative, Sodium 139 meq/l, Potassium 4.2 meq/l, BUN 18 mg/dl, Creatinine 0.6 mg/dl, arterial blood gas (ABG): HCO₃ 19.8, pCO₂ 38, pH 7.45.

**Specific laboratory tests:** Normal C₃, C₄, CH₅₀, ANA, c-ANCA and p-ANCA, Anti ds DNA.

**Doppler sonography of abdominal vessels:** Aorta diameter in upper part of abdomen was 15 mm and decreased gradually to 10 mm in lower part of abdomen. In celiac, superior mesenteric and both renal arteries origin, blood flow velocity increased to 3-4 m/s. Velocity of blood flow in proximal to this vessels was approximately 1 m/s (more than 70% stricture in origin). In lower abdomen before bifurcation of aorta many collateral vessels were seen. Liver and spleen were larger than normal. Portal and spleen vein diameters were normal. There was no portal hypertension.

**ECG:** Normal pattern.

**Chest Xray:** Prominent pulmonary artery, cardiothoracic (CT) ratio 65%.

**Echocardiography:** Stenosis of the origin of brachiocephalic artery and severe stenosis of the origin of left subclavian artery, left ventricular hypertrophy, mild aortic regurgitation.

**Angiography of aorta and its branches:** The findings of angiography are shown in fig 1 and 2.

The patient received atenolol and nifedipine for hypertension control. The abdominal pain was severe. We believed this pain was due to stenosis of celiac, superior mesenteric ostia. Therefore, we started corticosteroid pulses and then oral prednisolone (2 mg/kg/day). The blood pressure was controlled. Twenty days after discharge from hospital, he developed severe chest and abdominal pain and was referred to hospital. ECG showed extensive myocardial infarction (anterior and inferior wall), cardiac enzymes confirmed the diagnosis. In CCU clinical and paraclinical manifestation of pulmonary edema was eminent. Finally, he died because of myocardial infarction complications.
**Discussion**

In 1908, Mikito Takayasu, a Japanese ophthalmologist, reported the association of retinal arteriovenous anastomoses and absent upper extremity pulses. Takayasu arteritis (TA) is a chronic inflammatory disease of the aorta and its major branches of unknown origin. After Henoch-Schoenlein purpura and Kawasaki syndrome, it is the third most common form of childhood vasculitis worldwide. Women are affected in 80 to 90 percent of cases, usually between 10 and 40 years of age[3]. Although TA may occur in all races, it is more frequently found in Asians[5].

Takayasu vasculitis is characterized by granulomatous inflammation of the aorta and its major branches, leading to stenosis, thrombosis, and aneurysm formation. The disease progresses at variable rates to a late sclerotic stage in which there is intimal hyperplasia, medial degeneration, and adventitial fibrosis[6].

Early manifestations include fever, night sweats, anorexia, weight loss and arthralgia. Subsequently, features of hypertension, heart failure and pulse deficits become apparent.

Despite the term pulseless disease, which is a synonym for TA, the predominant finding in individuals with TA is asymmetric pulse. Absent peripheral pulses occur late in the course of the disease[5-7]. Hypertension may be paroxysmal and develops in more than one-half of cases due to narrowing of the renal artery, or narrowing and decreased elasticity of the aorta and its branches. However, narrowing or occlusion of the arteries in the arms may make it difficult to assess the blood pressure. Since hypertension typically results from renovascular compromise, this is a high-renin hypertension[8].

The reported incidence of 9-10% for coronary artery involvement is chiefly found in autopsy cases. This may be due to the late evidences of coronary artery disease such as angina pectoris, myocardial infarction[4]. Nowadays, a few children-aged Takayasu cases with coronary artery involvement and serious cardiac complications are reported. Vos et al reported Takayasu’s disease as the cause of myocardial infarct in an infant for the first time in 1987[9]. Another report by Basso et al in 1994 presented a 14-year-old girl who was rescued from a sudden cardiac arrest at...
school. Aortography disclosed mild aortic root dilation with aortic valve incompetence and subocclusion of coronary ostia and left common carotid artery. An emergency aortocoronary bypass operation was undertaken, but the patient did not recover from cardiopulmonary bypass. Autopsy disclosed massive myocardial infarction. The microscopic feature of arterial involvement was consistent with giant cell Takayasu's arteritis[10]. Similar to this case, angiography in our patient showed dilatation of the aortic arch and descending aorta, large areas of irregularities, occlusive and aneurismal formation of aortic artery and its branches.

Recently Limsuwan reported an 8-year-old girl with chest pain and the presumptive diagnosis of pneumonia and sepsis. She had myocardial infarction and her chest pain was attributed to myocardial infarction secondary to coronary occlusion from TA[11]. She developed refractory heart failure and died. It is possible that the coronary lesion was the cause of the heart failure and consequently, of death. Limsuwan's patient presented with chest pain; whereas our patient had severe abdominal pain, flashing and palpitation.

Generally arterial biopsy is not performed due to central location of the arteries involved by arteritis. Therefore, in order to confirm the diagnosis of Takayasu arteritis, imaging techniques of the aorta and major arteries is a diagnostic milestone.

Showing dilation of the aorta or increased mediastinal widening, chest X-ray may be useful to detect aneurysmal dilation of the great vessels. Additionally, using contrast angiography, although involvement may be most significant or limited to the more distal aorta and its branches, the arteriographic changes are seen to be most accentuated in the region of the aortic arch and its primary branches.

Smooth-walled, tapered, focal, or narrowed areas with some evidences of dilation are the primarily observed abnormalities. However, collateral circulation is often prominent due to chronicity of the disease. Beside defining the location and appearance of the arterial lesion, arteriography may also have a therapeutic role by allowing an intervention such as angioplasty and/or stenting of a stenotic area.

Treatment is mainly initiated with daily administration of high-dose corticosteroid. Additional agents including weekly infusions of methylprednisolone (30 mg/kg, not to exceed 1 g/wk), weekly methotrexate, daily or monthly intravenous cyclophosphamide, cyclosporine and mycophenolate mofetil are needed when a patient do not respond to corticosteroids or relapse during corticosteroid tapering. Another therapeutic choises like TNF inhibition with etanercept or infliximab has also been used while relapsing or glucocorticoid-dependency occur in a patient. Even more, surgical treatment of symptomatic stenotic or occlusive disease by means of percutaneous angioplasty, stenting or, in severe cases, by resection and placement of a manmade graft is required in patients with fibrotic changes following the acute phase.[12]

The patient reported here is an example of extensive Takayasu vasculitis. The angiography showed stenosis and dilatation of the aortic arch and descending aorta, large areas of irregularities, occlusive and aneurismal formation of aortic artery and its branches. Multiple large vessels such as common carotid, left subclavian, celiac, superior mesenteric and renal arteries were involved in imaging studies. In this case, the diagnosis of coronary involvement was initially missed due to the low index of suspicion for coronary involvement by TA causing subsequent myocardial infarction in such a young boy. Ultimately, the patient developed myocardial infarction secondary to coronary occlusion from TA in spite of treatment. We recommend performance of coronary angiography in all children with Takayasu arteritis.

**Conclusion**

The relatively high incidence of coronary involvement in Takayasu's disease and its often unexpected revelation by myocardial
infarction or sudden death, suggest that coronary arteriography should be undertaken more often during investigation of the arterial lesions of these patients. Takayasu’s disease should figure prominently amongst the causes of coronary artery disease in children.

References


