168 Takayasu Arteritis

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Definition

Takayasu arteritis (TA) is a chronic, idiopathic vasculitis of the large arteries. It primarily affects the aorta, its proximal branches, and occasionally the pulmonary arteries, which result in luminal stenosis, occlusion, or aneurysms.

Epidemiology

In children, the mean age of onset is 12 years. The female: male ratio is about 2:1. There are geographic variations in the presentation of TA. While obstructive lesions are frequent in the United States, Europe, and Japan, aneurysms are more common in Africa and India.

Etiology, Pathogenesis, and Genetic Background

Although many studies have proposed microorganisms such as bacteria and viruses as causative agents in TA, no specific infectious agents have been confirmed so far.

Circulating antiaortic endothelial cell antibodies (AAECAs), increased expression of E-selectin and vascular cell adhesion molecule-1, and increased production of interleukin (IL)-4, IL-6, and IL-8 have been reported in patients with TA.

Rare familial cases may suggest the role of genetic factors. Genes of certain activation molecules are upregulated in patients with TA.

Clinical Manifestations

Clinical manifestations are symptoms secondary to ischemia of organs supplied by stenotic vessels and constitutional symptoms such as fever, weight loss. The ischemic symptoms include stroke, visual aberration, angina, and renovascular hypertension, and claudication of extremities. Hypertension is one of the most frequent presenting symptoms in childhood. Headache is also a frequent complaint. Absent pulses and bruits over the stenotic vessel may be detected. Classically, TA follows a number of courses. The monophasic course is limited to 20% of cases. Some patients exhibit progressive or relapsing/remitting course.

Diagnosis and Laboratory Findings

To demonstrate angiographic abnormalitie(s) is mandatory for the diagnosis. Recent proposed classification criteria for TA is given in *Table 168.1*.

Acute phase reactants are elevated in about 2/3 of the patients. Autoantibodies, rhematoid factor, and ANCA are negative; however, anti-endothelial antibodies are frequently present. Electrocardiography may show findings of left ventricular hypertrophy suggesting chronic hypertension.

Radiologic Findings

Since TA involves aorta and its main branches, MR angiogram will be adequate to show the vascular lesions (*Fig. 168.1*). PET scan can be performed in selected cases, especially for differential diagnosis; however, the high cost is of concern.

Four types of involvement have been described in angiography. These include (1) aortic arch only; aortic arch and descending thoracic aorta; aortic arch, thoracic and abdominal aorta; aortic arch and abdominal aorta, (2) descending thoracic aorta only; descending thoracic and abdominal aorta, (3) diffuse aortic involvement, and (4) diffuse aortic and pulmonary artery involvement.

Treatment

Corticosteroids, methotrexate (MTX), azathioprine, mycophenolate mofetil, and cyclophosphamide (CYC) have all been used in the treatment of TA. A high (50%)

Abdelaziz Y. Elzouki (ed.), *Textbook of Clinical Pediatrics*, DOI 10.1007/978-3-642-02202-9_168,

Table 168.1
Classification criteria for Takayasu Arteritis, Ankara 2008 (Ozen S, Pistorio A, Iusan SM et al. (2010) The EULAR/PRINTO/PRES criteria for Henoch-Schönlein Purpura, Wegener Granulomatosis, Takayasu Arteritis and Childhood Polyarteritis Nodosa: ANKARA, 2008. Ann Rheum Dis 69:798–806)

Angiographic abnormalities (mandatory criterion) and 1 out of 5 of the following criteria:
1. Pulse deficit or claudication
2. Four limbs blood pressure discrepancy
3. Bruits
4. Hypertension
5. Increased acute phase reactants

relapse rate is observed in adults treated with corticosteroids only. There is no consensus on follow-up and no A-level evidence-based data on the treatment of childhood patients. In a recent study, patients were allocated to receive (1) oral steroids and MTX if they had disease limited to one side of the diaphragm only, and without pulmonary disease; and (2) oral steroids and oral CYC followed by oral MTX as above if the disease was more widespread. This single-center experience suggests that cyclophosphamide and corticosteroid induction followed by methotrexate is an effective and safe treatment for childhood TA and may prevent relapses. Angiotensin-converting enzyme inhibitors and angiotensin1 receptor blockers should be avoided in the presence of renal artery involvement. In resistant cases, anti-TNF treatment has been successful in case series.

Angioplasty or bypass grafting can successfully be performed when needed. However, this procedure should be done when the patient is in remission.

Prognosis

There is an absence of prospective and randomized controlled trial data in TA in childhood. However, from adult studies, it is estimated that TA has an overall 10-year survival rate of approximately 90%. This rate is reduced by the presence of complications. Optimal management of these factors could reduce the mortality.

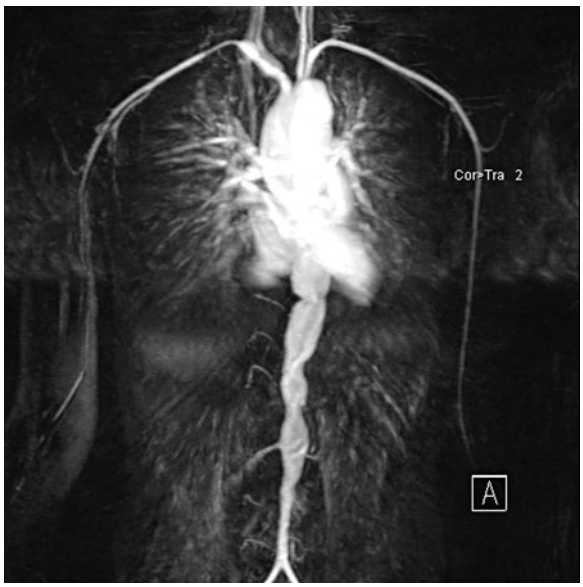

Figure 168.1
Stenosis in right subclavian artery and aneurysmal and stenotic segments in the abdominal aorta (With the courtesy of Dr. Tuncay Hazrolan)

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