CASE REPORT

Takayasu's Disease

by

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Abstract

A fourteen years old girl developed Takayasu's arteritis (pulseless disease) since six months prior to investigation. This unusual form of arteritis is common in Japan and Korea but has rarely been reported in individuals born in the United States. In Indonesian literature it has never been documented so far. The etiology is unclear. The literature currently but forward the hypothesis of an autoimmune basis and treatment with steroid. Although a tuberculin sensitization pathogenesis has been suggested, a close temporal relationship with the onset of a tuberculous process has not previously been documented. The likelihood of uncovering tuberculin sensitivity or active tuberculosis in patients with Takayasu' arteritis is substantially higher than in the general population in all countries analyzed. The natural history of his arteritis is highly variable. The adolescent described in this paper has demonstrated no response either to antituberculosis therapy, or to heparin and corticosteroid as suggested by Ishikawa, 1987. The patient died on the 59th day of hospitalization after getting syncopal attacks followed by shock.

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Introduction

Takayasu's disease, also known as non-specific arteritis or pulseless disease, is a chronic inflammatory arteriopathy of unknown cause, affecting the aorta and/or its branches and often the pulmonary artery. Most reports of this condition have originated from Japan, where Takayasu, an ophthalmologist first described the peculiar vascular funduscopic changes in a young woman in 1908. Since then, this disease has been recognized worldwide in patients of various racial origins, mainly young woman (Taieb et al., 1987).

These pathological events lead to occlusive changes in the lumina, often combined with dilation and secondary thrombosis formation. Major complications attributed to the disease are Takayasu's retinopathy, secondary hypertension, aortic regurgitation and aortic or arterial aneurysm. There are geographical variations in the clinical aspect of this disease. The following terms may be identical with what is known as Takayasu's disease or arteritis entity: Takayasu's arteriopathy, occlusive thromboaortopathy, aortitis syndrom, and non-specific aorto-arteritis (Ishikawa, 1987).

Initially thought to be confined to Japan and Korea, it has now been reported in India, Israel, South America, Mexico, South Africa, Italy, Spain, Scotland and Scandinavia (Pantell and Goodman, 1981). It is not frequently seen in the United States, with less than 2 dozen cases documented in the literature for individuals born in this country (Warshaw and Spach, 1966). It is not well documented so far in the Indonesian literature. This is the first case reported in Indonesia. Its possible etiology and management are discussed in this paper.

Case report

T., a girl fourteen years old, the second of four normal children, was admitted to the hospital on June 16th, 1989 because of visual disturbance and syncopal attacks.

She had been well until 6 months ago, when she began to experience fatigue, occasional dull occipital headaches, mild fever and loss appetite. Four months later she had syncopal attacks and her vision was blurred. The patient denied exertional dyspnea, abdominal pain, joint pain or erythema of the external ear or nasal bridge. Her grandfather who stayed together in the same small house had tuberculosis and took medicament regularly, and she had never had BCG vaccination.

On examination she appeared weak and her face showed mild cyanosis. Her height was 153 cm and weight 31.5 kg (66 per cent P50 Harvard). The head and the neck were normal. Body temperature was 36.9°C. The left carotid, subclavian and radial pulses were not palpable; the right carotid pulse was slightly palpable, whereas the femoral, popliteal and dorsal pedal pulses were fully palpable bilaterally; the pulse rate was 100/minutes. Bruits were present over the right carotid artery. The blood pressure could not be measured over the upper extremities, whereas it revealed 210 mmHg systolic and 180 mmHg diastolic pressure over her right and left thigh.

The lungs were clear, and the heart was not enlarged; the rhythm was regular and neither systolic nor diastolic murmurs were present. No collateral vessels could be palpated at the thoracic wall. There was no peripheral edema, clubbing nor cyanosis. Neurologic examination was normal. An ophthalmologist noted papil edema and occlusion of retinal vessels, exudation and slight hemorrhage in the retina.

Laboratory data were as follows: the urine was normal, the hematocrit 34 per cent; the white blood-cell count was 6.700/ul with 52 per cent polymorphonuclear leucocytes, 1 per cent eosinophils; the platelet count was 280.000/ul and the sedimentation rate 101 per hour. The prothrombine time and partial thromboplastin time were normal. The glucose 84 mg/dl and the total protein 8.4 g/dl, the albumin 3.9 g/dl and the globulin 4.5 g/dl. Test for rheumatic factor, C reactive protein, ASTO, Takahashi and VDRL were negative. The BCG test was strong positive (20 × 20 mm^2).

All electrocardiogram demonstrated abnormal rhythm at rate of 94 with normal inteval and axis; the pattern was considered normal. X-ray film of the chest revealed a normal appearance of the lungs, heart and mediastinum; no rib notching was observed. A percutaneus retrograde thoracic aortography examination and cineangiographic study (Fig. 1 A & B) disclosed an occlusion and a narrowing in the brachiocephalic branches, occlusion in the right subclavian and carotid arteries, and rowing in the left subclavian and carotid arteries, a narrowing in the branches of abdominal aortae especially both renal and mesentery arteries. There were numerous collateral vessels to the brachiocephalic branches via the intercostal arteries (Fig. 1 A & B).

On the basis of the physical and laboratory findings she was diagnosed as suffering from pulseless disease or arteritis disease or Takayasu's disease or Takayasu's arteritis, with systemic hypertension but without cardiac and renal abnormalities.

The treatment was given according to the standard therapy proposed by Ishikawa (1987) i.e. 50 mg prednison per day which was then gradually reduced until 10-20 mg per day. Due to the history of tbc contact, nutritional status 66 per cent standard (P_{50} Harvard) and strong positive of BCG test the patient was treated with triple tuberculostatic drugs consisting of rifampicin, isoniazid and ethambutol. Owing to Ishikawa (1987) that occlusive changes in the lumina of the arteritis are often combined with dilatation and secondary thrombus formation, heparin was given to this patient intravenously every day. Following institution of heparin therapy, the patient's blood pressure decreased to 140 mmHg systolic and 90 mmHg diastolic. Heparin therapy was continued until for weeks intravenously, but despite decrease in blood pressure, no clinical improvement occurred. On the 59th day of hospitalization she got syncopal attacks, femoral and dorsal pedal pulses were gradually becoming smaller and then became even not palpable. Thus the blood pressure could not be measured. She was given Ringer lactate infusion and dopamin but inspite of all effects she died three hours after falling into a shock.

Discussion

A form of vasculitis, Takayasu's arteritis is generally classified along with temporal arteritis as one of the giant cell arteritides (Fauci et al., 1978). However, giant cells are found far less frequently in Takayasu's arteritis than in other common forms of arteritis. The prevalence of this form of aortitis may be greater than a few case reports indicate. In an autopsy series in New Orleans, 0.1 per cent of cases had nonspecific aortitis of the descending aorta (Restrepo et al., 1969). As a cause of renovascular hypertension Takayasu's arteritis ranks first in non white children (Wiggelinkhuizen and Cremin, 1978). The occurrence in a set of identical twins

suggests a genetic predisposition (Numaro et al., 1978).

The presenting symptoms in our patient were similar with the reported series in which 67 to 90 per cent of the cases occurred in females with onset generally in the second and third decades (Lupi-Herrera et al., 1977). She exhibited a number of symptoms and signs as found in the majority of patients including decreased pulse, malaise, headache, fever, easy fatigability of the extremities, dizziness, transient visual disturbance, nausea and syncopal attacks (Ishikawa, 1987). She did also demonstrate certain common features which usually accompany renovascular involvement such as hypertension (Wiggelinkhuizen and Cremin, 1978).

Increased sedimentation rates were documented in 82 per cent and 100 per cent of two series; Nakao et al. (1967) demonstrated the evidence of the tendency of the sedimentation rate and c reactive protein to decrease years after onset of the symptoms and signs of Takayasu's arteritis. Nonspecific autoimmune investigation (rheumatoid factor, anti nuclear antibodies, lupus erythematosis) has not uncovered a clear mechanism.

To evaluate the disease states and for better understanding of the clinical profile in an individual patient, it is pertinent to clarify where the patient belongs in each of the varied classifications according to the following three factors: inflammatory activity of the disease as determined by the erythrocyte sedimentation rate (ESR), site of arterial lession and complications attributed to Takayasu's disease.

When the erythrocyte sedimentation rate is consistently 20 mm/hour or more (ESR Westergen), is particularly over 40 mm/hour, or less than 20 mm/hour, the inflammatory activity may be defined as the active and inactive stage respectively (Ishikawa, 1987).

According to the location of arterial lesions, the disease is anatomically classified into three types: (1) the arch type involving the aortic arch and its branches; (2) the descending type involving the descending thoracis and abdominal aorta and its branches; (3) the extensive type which describes the combined arch and descending type (Ishikawa, 1987).

The patient's arterial lesions belonged to the extensive type. All branches of the arcus aorta and lower abdominal aorta were stenotic or almost obstructed. Other disorders that can involve the aorta, such as syphilis, and coarctation had been excluded.

The prognosis is generally favourable although characteristics that offer assistance in predicting the course of the illness have not been delineated (Sunamori et al., 1976). Steroid have been heralded as being therapeutic without having been subjected to a clinical trial. Nakao et al. (1967) administered corticosteroid to 29 patients; they claimed "remarkable" clinical remissions in five patients in whom the pulse became palpable and "sufficient" remission in 13. None of the 15 patients receiving antituberculosis treatment improved.

The association between tuberculosis and Takayasu' arteritis has been suggested by several authors (Zerpa et al., 1966; Ueda et al., 1968). Tuberculin sensitivity was documented in 81 to 100 per cent of patients in the six studies reporting this data. Only one study provides comparative figures for the general population. Tuberculin (PPD-S) conversion occurred in 81 per cent of patients and 66 per cent of the general population (Lupi-Herrera et al., 1977). In India where prevalence of active tuberculosis figures were available, patients with Takayasu's arteritis were 46.6 times as likely to have had active tuberculosis as general population (Kinare, 1970).

In our patient the evidence of tbc contact, low nutritional state and strong positive BCG test suggested reactivation of a tuberculosis process. The precise timing of the tuberculosis reactivation was uncertain; whether the tuberculosis infection occurred prior to the development of the arteritis was speculative. However, the close temporal relationship between these two events is clearly documented and suggest an association. Two alternative hypotheses exist, the first is that a stressful event or infection, such as streptococal pharyngitis, triggered reactivation of the tuberculosis. Considering the documented association of the two conditions in the literature, we favor the former hypothesis.

For that reason, she was treated with intensive tuberculosis therapy. The result of this therapy, however, was not satisfactory. No improvement had ever been found in this patient.

Surgical treatment due to the so many aortic branches affected (Fig. 1 A & B) was not possible. Corticosteroid and heparin were not helpful, and the patient died on the 59th day of hospitalization.

In conclusion, patients with Takayasu's disease present challenging clinical problems for the clinician, both in the selection of appropriate surgical candidates and in the aspects of medicamentous treatment. The authors believe that histological identification of a granulomatous inflammatory process or fibrosis of the external part of the arterial wall is required before the diagnosis of Takayasu's disease can be confirmed. Finally, since Takayasu's disease is characterized by a progressive involvement of multiple vessels that involve vital organs, early diagnosis, long term follow up and repeated evaluation are required to provide the best prognosis for long term survival.

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