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Case Report

Asymptomatic Takayasu Arteritis Disease in a 13-Y-Old Girl: A Case Report with a Short Review

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Abstract

A 13-y-old girl presented with complaints of cough, cold, and fever for 3 days; she was considered as a case of viral rhinitis. As her pulses were feeble on examination, she was admitted and was found to have weak pulses all over. Prior to this she was asymptomatic. Her aortogram revealed multiple stenoses of major arteries of thorax and upper limbs. She was diagnosed to have Takayasu arteritis.

***Keywords:** Takayasu, arteritis, pulseless disease, weak pulses, aortic stenosis, adolescent girl, Large vessel arteritis.*

Case History

A 13-year-old girl presented with fever, cold, cough, and headache for three days. On examination she was conscious, oriented and had runny nose with clear discharge. Her pulse was 80/ min respiration was 24/ min, and blood pressure was 96/72 mm of Hg, her weight was 29 kg, height was 136 cm, and BMI was 15.7 underweight, thin built. The resident doctor in outpatient admitted her as she found weak pulse and difficulty in measuring blood pressure in the girl. She was found to have weak pulse in both entire upper limbs; and the femorals at groin, and dorsalis pedis at foot, were not easily palpable. The heart sounds were normal and there was no murmur. Rest of the clinical exam was normal including the fundus exam, [except for the upper respiratory infection part, considered as viral origin].

Investigations

She was mildly anemic (Hb 10 g/dl), her ESR was 17 and CRP was 6.6 mg/dl.

Pulseless or Takayasu arteritis was suspected and a CT aortogram (MDCT angiography of aorta and neck vessels) was carried out.

The aortogram 3D pictures revealed (Figures 1 - 3)

- Brachio-Cephalic artery <50% luminal narrowing minimal circumferential hypodense wall thickening
- Right proximal common carotid artery (CCA) and subclavian marked stenosis >90% narrowing with marked hypodense non enhancing circumferential wall thickening
- Proximal left CCA with significant (near total block) stenosis hypodense non-enhancing circumferential wall thickening
- Left distal CCA upto bifurcation mild circumferential wall thickening
- Proximal both vertebral arteries significant (near total block) occlusion non-enhancing circumferential wall thickening
- Proximal left subclavian artery near total occlusion non-enhancing circumferential wall thickening
- Aortic arch with <50% luminal narrowing extending into proximal descending thoracic aorta with focal segmental >70% luminal narrowing at the level of left atrium mild non-enhancing circumferential wall thickening

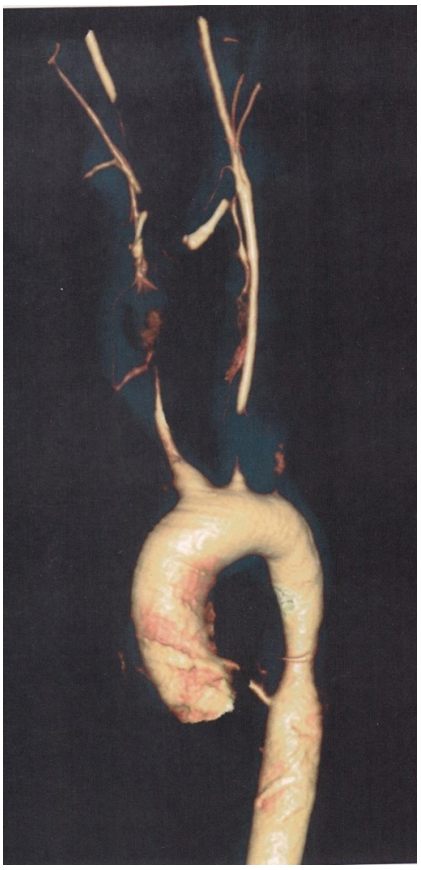

Fig 1: *Aortic Carotid involvement.*

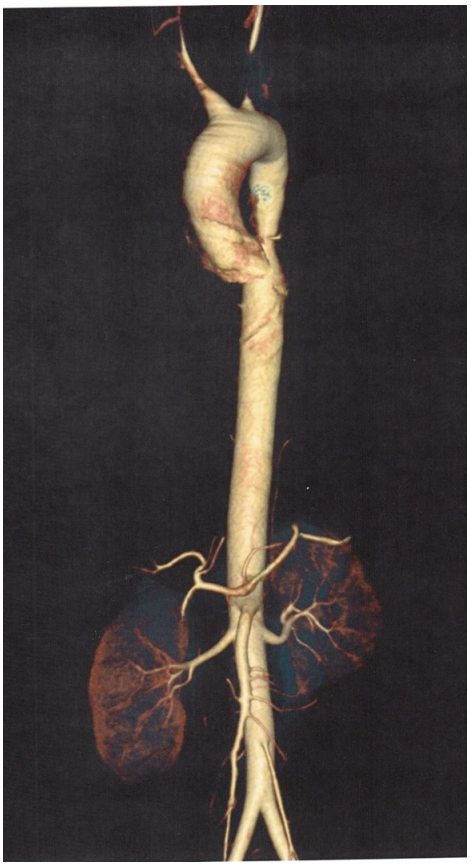

Fig 2: *Preserved Renal arteries.*

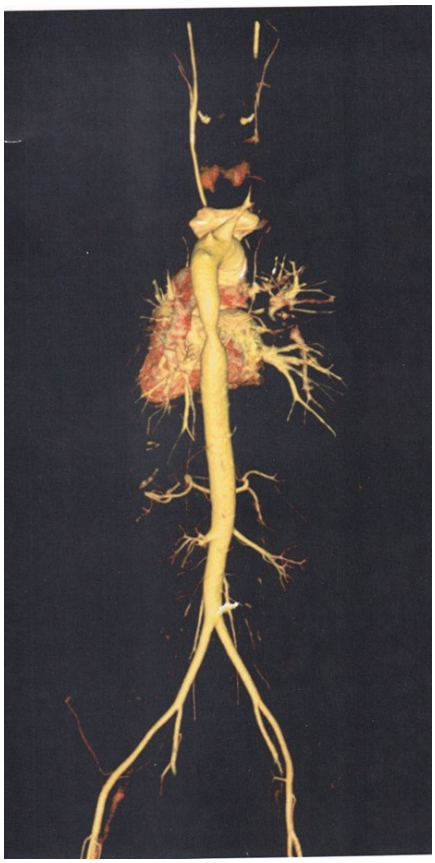

Fig 3: *Abdominal arteries much less involved.*

Abdominal aorta, aortic bifurcation, celiac trunk; hepatic, splenic, superior mesenteric, proximal common iliac, and both renal arteries were normal (fig. 2, 3)

Findings suggest "Changes of Arteritis involving aortic arch, descending thoracic aorta, and major neck and upper limb arteries; Takayasu Arteritis; correlate clinically."

Treatment

Paracetamol and nasal saline drops were given for viral infection.
For Takayasu disease she was started on low-dose steroids oral prednisolone 0.5 mg/kg/day.
She recovered in three days and was feeling better, and as she was asymptomatic before the viral rhinitis presentation also, she was discharged with advice to visit after 4-weeks. However, she did not return for follow-up.

Discussion

Takayasu Disease is a large vessel arteritis. [1]
Takayasu's arteritis
Blood vessel inflammation, autoimmune disease, affects girls and women < 40 years.
The disorder occurs worldwide, but is mostly seen in Asia.

Proposed Classification Criteria for Pediatric-onset Takayasu Arteritis [2, 3]
Angiographic abnormalities (conventional, CT, or magnetic resonance angiography) of the aorta or its main branches and at least one of the following criteria:

- Decreased peripheral artery pulse(s) and/or claudication of extremities
- Blood pressure difference between arms or legs of >10 mm Hg
- Bruits over the aorta and/or its major branches
- Hypertension (defined by childhood normative data)
- Elevated acute phase reactant (erythrocyte sedimentation rate or C-reactive protein)

Sometimes the condition runs in families. *HLA-B*52* locus has many of the genes. Non-HLA loci are *FCGR2A/ FCGR3A*, *IL12B*, *IL6*, *RPS9/LILRB3*, and a locus on chromosome *21* near *PSMG1*. [4]

Clinical features [1] *(present in this case are)*

Stage 1

- Fatigue
- *Unintended weight loss*
- Muscle and joint aches and pains
- Mild fever, sometimes accompanied by night sweats

Stage 2

- Weakness or pain in limbs with use
- *A weak pulse*, *difficulty getting a blood pressure reading* or a difference in blood pressure between arms
- Light-headedness, dizziness or fainting
- Headache or visual changes
- Memory problems or trouble thinking
- Chest pain or shortness of breath
- High blood pressure
- Diarrhea or blood in stool
- *Anemia*, low RBC count

Complications [1]

- Hardening and narrowing of blood vessels, High blood pressure, kidney disease.
- Myocarditis, Inflammation of the heart, heart muscle / valves.
- Heart failure, Stroke,
- Transient ischemic attack (TIA), mini-stroke. TIA serves as a warning sign, does not cause permanent damage.
- Aortic Aneurysm, Myocardial infarction.

Different types of arteritis are small, middle, large vessel [2, 3]

I. Predominantly *Large Vessel* Vasculitis

• Takayasu arteritis

II. Predominantly *Medium Vessel* Vasculitis

• Childhood polyarteritis nodosa

• Cutaneous polyarteritis nodosa

• Kawasaki disease

III. Predominantly *Small Vessel* Vasculitis

[** with antineutrophil cytoplasmic antibody ANCA*].

A. *Granulomatous*:

• Granulomatosis with polyangiitis (Wegener granulomatosis) * [ANCA]

• Eosinophilic granulomatosis with polyangiitis (Churg-Strauss syndrome) * [associated with leukotriene antagonist like monteleukast, ANCA].

B. *Non-granulomatous*:

• Microscopic polyangiitis* [ANCA]

• Henoch-Schönlein purpura

• Isolated cutaneous leukocytoclastic vasculitis

• Hypocomplementemic urticarial vasculitis

IV. *Other* Vasculitides

• Behçet disease

• Vasculitis secondary to infection (including hepatitis B associated polyarteritis nodosa), malignancies, and drugs, including hypersensitivity vasculitis

• Vasculitis associated with connective tissue disease

- Isolated vasculitis of the central nervous system
- Cogan syndrome
- Unclassified

Treatment

Anti-hypertensive drugs if hypertension and stenting for major stenosis.[5]

Prognosis

About 20% has full-time remission.[3] A healthy pregnancy is possible.[1]

Conflict of Interest

The authors declare no conflict of interest. Consent was obtained from the parents to publish this case.

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