Case Report

Takayasu Arteritis Presenting as Renal Failure in 8 year old female child

Priyanka Poonam*

Tutor, Department of Pathology, Patna Medical College, Patna, India
*Corresponding author email: **ppdoc83@gmail.com**

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Abstract

Takayasu arteritis also known as Aortic Arch Syndrome or Pulseless disease is an uncommon condition mainly affecting aorta and its branches. It is chronic inflammatory disease of unknown cause, so also called as non- specific aortoarteritis. Though it is rarely seen in children, I am hereby presenting a case of 8 year old girl presenting with constitutional symptoms, claudication of lower limbs, anasarca altered sensorium and decreased urinary output.

Key words

Takayasu arteritis, Aortoarteritis, Renal Failure.

Introduction

TA is an inflammatory and stenotic disease of medium and large sized arteries characterized by a strong predilection for the aortic arch and its branches, so also called as Aortic Arch Syndrome [1]. Annual Incidence Rate of TA is 1.2-2.6 cases per million [1]. Although, it is mostly seen in adolescent girls and young women, it is also seen in childhood and rarely seen in infancy [7]. The clinical findings in children are usually constitutional symptoms [7]. Hereby, I am reporting an 8 year old girl presenting with Constitutional symptoms, anasarca, decreased urinary output and altered sensorium.

Case report

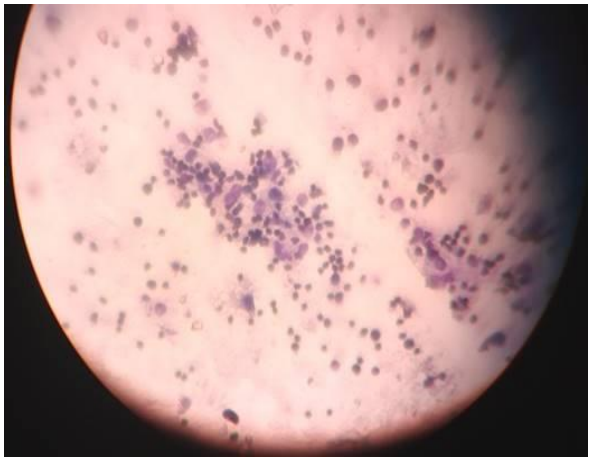
An eight year old girl presented with fever for one month, irregular and feeble pulses in lower extremities associated with claudication, swelling of the whole body starting from both lower limbs on and off vomiting, decreased urinary output, altered sensorium. She also had multiple nodular cervical swellings for 5-6 months. On examination there was anasarca of whole body fever 100Degree F, BP 120/80 mm of Hg feeble pulses in the lower limbs and

tenderness along vessels in the lower limbs. She was sent to the Pathology and department for the FNAC of cervical swellings. FNAC was done and the smears shown epithelioid granuloma as with background showing lymphocytes and plasma cells.

Peripheral blood smear was also made which show microcytic hypochromic RBCs most likely Iron Deficiency Anemia. Her lab findings were Hb. 7.2 g/dl, WBC- 21,700 cells/uL, differential count with all lymphocytes. ASO titre -471TDU raised serum alkaline phosphatase 12.9 K.AU/100ml, SGPT-23U/ml, Reticulocyte count - 2.6%. Total protein - 6.6g/100ml, Albumin-3.7g/100ml globulin -2.9g/100ml Albumin globulin ratio 1:27:1 S. creatinine, 1.7 mg/dl BUN, 60 mg/dl, ESR and CRP raised. CT Angiography report showed aortoarteritis at T4-T6, L2-L3, with semi-stenosis of left renal artery.

Echocardiography report revealed mild atrial regurgitation, mild mitral regurgitation, normal right ventricular systolic, LVEF - 60%. She got admitted in Pediatrics ward and was given oral prednisolone in low doses and acetyl salicylic acid for one week. She became afebrile and acute phase reactants normalized (**Figure – 1 to 3**).

Figure – 1: Smear showed epitheloid granuloma as with background showing lymphocytes and plasma cells.

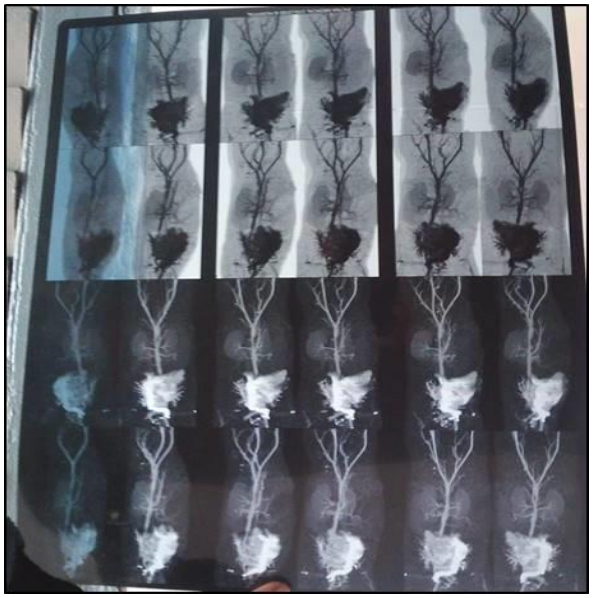


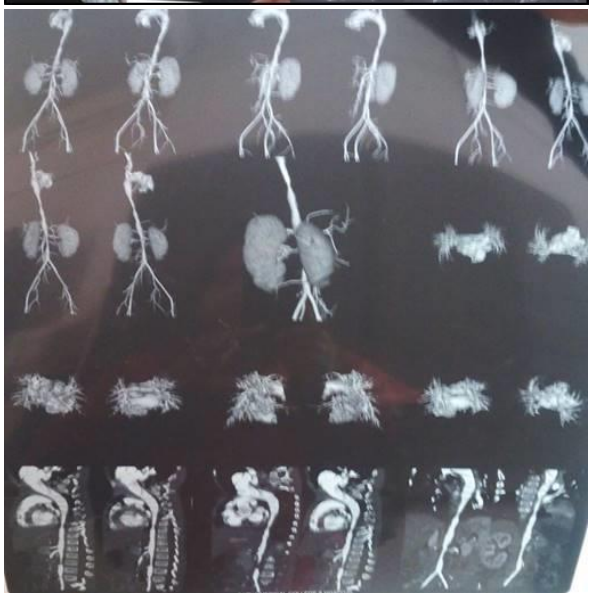
Discussion

TA is a rare large vessel vasculities with an annual incidence rate of 1.2-2.6 cases per million [1]. The American Rheumatologic society considers three of the following six criteria necessary for the diagnosis [9].

- On set of disease before 40 years.
- Claudication of extremities
- Decrease in the brachial pulse in one or both arms
- Difference of 10mmHg/ more in BP measured in both arms.
- Audible bruit on auscultation of aorta/subclavian artery.
- Narrowing at the aorta or its primary branches on arteriogram.

Figure – 2, 3: CT angiography report showed aortoarteritis at T4-T6, L2-L3 with semistenosis of left renal artery.





In this case an 8 year old girl presented with claudication of extremities with feeble pulses in lower limbs and her CT Angiography shown semi stenosis of left renal artery with aortorteritis (**Table – 1**).

Table – 1: Frequency of arteriography abnormalities and potential clinical manifestations of arterial involvement.

Artery	% Arteriography abnormalities	Clinical Manifestation
Subclavian A	93%	-Arm claudication -Raynaud's phenomenon
Common Carotid A	58%	-Visual changes -Syncope -Transient Ischemic Attack -Stroke
Abdominal aorta	47%	-Abdominal pain -Nausea -Vomiting
Renal A	38%	-Hypertension, Renal failure
Aortic Arch/Root	35%	-Visual Changes -Dizziness
Coeliac Axis	18%	-Abdominal Pain -Nausea -Vomiting
Superior Mesenteric A	18%	-Abdominal Pain -Nausea -Vomiting
Iliac	17%	-Leg Claudication
Pulmonary	10-40%	-Atypical chest pain -Dyspnea

Table – 2: Classification of TA according to site of involvement.

Type I	Aortic arch
Type II	Descending aorta
Type III	Aortic arch + Descending aorta
Type IV	Aorta + Pulmoary artery involvement

Three of the above six criteria's is being fulfilled to diagnose her as a case of TA (**Table – 2**).

TA is the commonest cause of Reno vascular hypertension in India [2]. TA produce narrowing of the renal arteries, resulting in renal ischemia and hypertension [3].

Thoracoabdominal aortic involvement is commoner in children [4]. TA is a chronic sclerosing aortitis of unknown aetiology that can cause renal artery stenosis from narrowing of the ostium [3]. The underlying mechanism involves cell mediated immunity pathways with activation of inflammatory cells such as macrophages, cytotoxic T-cells, TNF- α, NK cells and increased levels of cytokines such as IFN γ and IL-6 [9]. The main mediator for the inflammatory cascade is T-cell mediated immune response [9]. In TA, histological sections of the artery shows chronic inflammation and fibrosis of the arterial wall which has a predilection for the aortic arch branches [3]. This leads to absence of pulses in the upper extremities, ocular and neurological symptoms. The disease is a panarteritis with inflammatory cell infiltrates and occasionally giant cells. There are marked intimal proliferation and fibrosis, scarring and vascularization of the media and disruption and degeneration of the elastic lamina [1]. The evolution from normal renal function to symptomatic chronic renal failure progresses through 4 stages (**Table – 3**).

Table – 3: Evolution from normal renal function to symptomatic chronic renal failure progresses through 4 stages.

Stage	GFR	C/F
Diminished renal reserve	50% of Normal	Asymptomatic
Renal insufficiency	20-50% of Normal	- Azotemia - Anemia - Hypertension - Polyuria -Nocturia
Renal Failure	<20-25% of Normal	- Oedema - Metabolic Acidosis - Hypo Calcaemia
End Stage Renal Disease	<5% of Normal	-Terminal Stage of Uremia

When arteriosclerotic changes are seen at unusual site in young or middle age individuals, TA should always be considered as differential diagnosis [3]. A high frequency of haplotype A24-B52-DR2 has been found in Japanese patient [2].

Many children with TA have associated tuberculosis and strongly positive Monteux Reaction [2]. In this case, the girl presented with multiple cervical lymph nodes enlarged associated with fever for 5-6 month which on FNAC, the smears shown epithelioid granulomas with lymphocytes and plasma cell's in the back ground.

TA is a systemic disease with generalized as well as vascular symptoms [1]. Malaise, fever, night sweats, arthralgia anorexia and weight loss are the constitutional symptoms which occur months before vessel involvement [1]. Evidence of vessel inflammation such as tenderness along arteries, bruit and aneurysm may point to the diagnosis of TA [4, 6] vascular manifestation are decrease or absence of peripheral pulses, discrepancies in blood pressure and arterial bruits [1] lab findings include elevated ESR, mild anemia, elevated immunoglobulin levels [1]. Later manifestation includes claudication, Raynaud's phenomenon, renal failure, splenomegaly and symptoms of pulmonary or cardiac ischemia or stroke [5].

Aortography is diagnostic and reveals narrowing or occlusion of entire aorta and its branches or large arteries in proximal upper and lower extremities. Magnetic resonance angiography has better resolution [2]. Arteriography also shows irregular vessel walls, stenosis poststenotic dilatation, aneurysm formation, occlusion and evidence of increased collateral circulation [1]. The course of the disease is variable. In some persons there is rapid progression but in other a quiescent stage, is reached in 1 or 2 years permitting long term survival sometimes with visual or neurological deficits [10].

Death of TA patients occurs due to congestive cardiac failure, cerebrovascular accidents, myocardial infarction, aneurysmal rupture or renal failure.

Treatment includes low dose prednisolone. Remission is achieved is 60% patients treated with glucocorticoids alone although relapses may occur with dose reduction [7].

Unless it is urgently required, surgical correction of stenosed arteries should be undertaken only when the vascular inflammatory process is well controlled with medical therapy [1].

Cyclophosphamide and Methotrexate can be added if the patient is unresponsive to glucocorticoids alone [9]. Surgical treatment is angioplasty. The 5 year survival rate of the patients is >90% [2].

Conclusion

Although rare, childhood TA must be considered in children presenting with non-specific systemic symptoms, hypertension and increased acute phase reactants. High index of suspicion is required in children presenting with constitutional symptoms with limb claudication/ Raynaud's phenomenon so as to reduce significant morbidity associated with disease.

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