P-ISSN: 2664-3685
E-ISSN: 2664-3693
www.paediatricjournal.com
IJPG 2022; 5(1): 39-40
Received: 21-11-2021
Accepted: 23-12-2021

Dr. Haripal Kashyap
Department of Pediatrics and Neonatology, Faridabad, Haryana, India

Dr. Shreyansh Valjiyani
Department of Pediatrics and Neonatology, Faridabad, Haryana, India

Dr. Vineetranjan Gupta
Department of Pediatrics and Neonatology, Faridabad, Haryana, India

Dr. Seshendra Sharma
Department of Pediatrics and Neonatology, Faridabad, Haryana, India

Dr. Kanika Sain
Department of Pediatrics and Neonatology, Faridabad, Haryana, India

Dr. Srishti Gautam
Department of Pediatrics and Neonatology, Faridabad, Haryana, India

Corresponding Author:
Dr. Shreyansh Valjiyani
Department of Pediatrics and Neonatology, Faridabad, Haryana, India

Takayasu arteritis: A surgical case

Dr. Haripal Kashyap, Dr. Shreyansh Valjiyani, Dr. Vineetranjan Gupta, Dr. Seshendra Sharma, Dr. Kanika Sain and Dr. Srishti Gautam

DOI: https://doi.org/10.33545/26643685.2022.v5.i1a.164

Abstract
Takayasu's Arteritis (TA), also known as pulseless disease, is a chronic large vessel vasculitis of unknown etiology that predominantly involves aorta and its major branches. Diagnosis of TA remains challenging due to non-specific symptoms in childhood. We report a 9 year old girl presenting with complaints of fever of short duration, associated with severe pain abdomen. Clinical examination revealed decreased pulses (right radial and right brachial), hypertension, hepatomegaly, increased ESR and high sensitive CRP. CT Angiography was suggestive of TA. TA must be considered in children who present with Fever and Severe Pain abdomen with nonspecific findings on abdominal examination, Decreased peripheral pulses, hypertension, and increased acute phase reactants.

Keywords: Abdominal pain, Takayasu's arteritis, large vessel vasculitis

Introduction
TA is a chronic inflammatory disease that involves aorta, its branches, and pulmonary artery. Inflammation results in varying degree of stenosis, occlusion or dilation of involved vessels. The etiology and precise pathogenesis of TA is still unknown. This article reviews the salient features of TA in children.

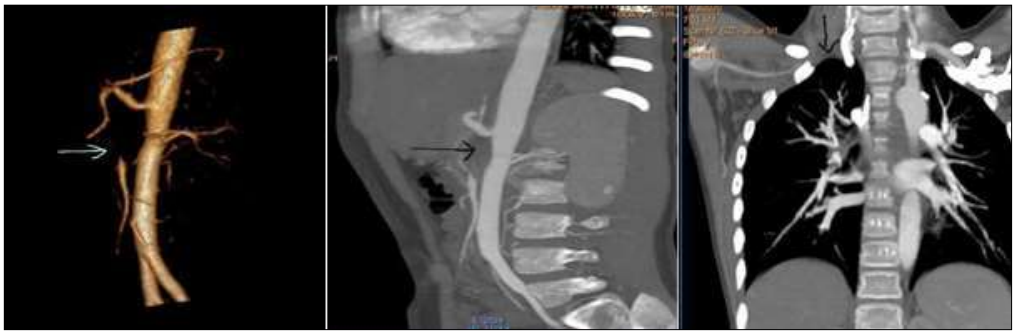
Case report
Young girl aged 9 years, presented to our hospital with complaints of fever since one week associated with severe intermittent pain abdomen. There was history of her having nonspecific symptoms such as malaise, weakness, fatigue, pain abdomen and fever off and on in past over the last one year or so.

Examination revealed pulse rate of 100/min with significantly diminished right radial and right brachial artery pulsations as compared to left side. BP was 160/100 mm Hg in left arm and 130/80 in right arm. She had mild fever and Tachypnea Respiratory rate-35/minute, temperature-99.9 F. Per abdomen examination was revealed generalised tenderness and guarding. Positive laboratory findings include elevated erythrocyte sedimentation rate (25 mm/1st hour) and high sensitivity C reactive protein (104 mg/dl). Blood culture, viral markers and autoimmune serological markers were negative. Angiographic findings included plaque in descending aorta with occlusion of subclavian and proximal axillary arteries& occlusion of SMA and right renal artery from origin. All other abdominal arteries were normal.

DTPA renal scan done showed mild to moderate impaired cortical function (lower half) in right kidney. Differential renal function showed decreased GFR in right kidney compared to left kidney. In view of involvement of large vessels, diagnosis of Takayasu's arteritis with multiple vessel occlusion (Subclavian artery, Superior mesenteric artery, Right renal artery) with impaired right renal function was made.

She was treated with high dose IV methyl prednisolone for 5 days along with antihypertensive drugs. Despite high dose steroid therapy, severe pain in abdomen persisted. Parents were explained pros and cons of medical versus surgical management. In view of unrelenting severe pain abdomen with suspicion of ongoing bowel ischemia and poor response to medical management, Aorta to SMA bypass grafting using reverse saphenous vein was done by CTVS team. Intra-operatively bowel looked pale and ischaemic. Postoperatively there was significant improvement in symptoms and inflammatory markers stabilized gradually. She was discharged on oral steroids, antihypertensive drugs, methotrexate and folic acid.

Although surgical management is not the 1stoption of management in TA, It should be considered as such in cases where symptoms are severe and do not respond to adequate medical management, as was done in our case.



(A) (B) (C)

Fig 1: (A) and (B) showing occlusion of proximal SMA and (C) showing occlusion of subclavian artery.

Discussion

TA is a chronic inflammatory disease of large and medium sized arteries; involving aorta and its major branches [1]. Although TA occurs worldwide, the disease is most common in Asian and Indian population [2].

Most children are diagnosed on an average at the age of 13 years with a reported F:M ratio of 2-4:1 in children and 9:1 in adolescents. Patients usually have no risk factors for atherosclerosis and yet have atheromatous aorta [3].

Vascular changes leading to main complications include hypertension, most often due to renal artery stenosis. Fever, malaise, weight loss, headache, myalgia, dizziness and abdominal pain are the complaints in pre pulseless stage of disease. Later manifestations include diminished pulse, asymmetrical blood pressure, claudication, Reynaud's phenomenon, renal failure and cardiac/pulmonary failure. Inflammation may cause aortic insufficiency [4].

Proposed classification criteria for Pediatric onset Takayasu's arteritis is as follows:

Angiographic abnormality plus any one of the following [4]:

- Decrease arterial pulse and/or claudication of extremities
- Blood pressure difference of arms or legs >10 mm Hg
- Bruits over aorta or major branches
- Elevated acute phase reactants (ESR and CRP)

Progressive vascular damage can result in arterial stenosis, aneurysm and occlusion which produce ischemic symptoms. Potential ischemic complications include stroke, renal impairment/failure, myocardial infarction, mesenteric ischemia and limb threatening arterial disease [5].

The indications for considering surgical intervention include uncontrolled hypertension as a consequence of renal artery stenosis, severe symptomatic coronary artery or cerebrovascular disease, severe aortic regurgitation or coarctation, stenotic or occlusive lesions resulting in critical limb ischemia, and aneurysms at risk of rupture. In these cases the risk benefit ratio for surgery is good [6, 7].

Miyata *et al*. demonstrated that surgery increases the long-term survival of patients in late stages of TA (major complication and progressive disease) while conversely survival is decreased in early stages of TA patients (no major complications and no evidence of progressive disease) due to surgery-related complications [7].

Conclusion

Delayed diagnosis and lack of specific treatment might explain the extent and severity of the disease at the time of hospital admission. Hypertension and headache are the main presenting manifestations of TA and should prompt consideration of TA when present without alternative explanation.

References

1. Ohta Ein Seltener K, Fall Von Beiderseitigem. Carotissubclavia verschluss: Ein beitrag zur pathologie der anastomosis peripapillaris des auges mit fehlendem. Trans Soc Pathol Jpn. 1940;30:680-90.
2. Kliegman, Blum. Nelson textbook of pediatrics, 1320.
3. Koide K. Takayasu's arteritis in Japan. Heart Vessels Suppl. 1992;7:48-54.
4. Kliegman, Blum. Nelson textbook of pediatrics, 1320.
5. Kliegman, Blum. Nelson textbook of pediatrics, 1321.
6. Fields CE, Bower TC, Cooper LT, *et al*. Takayasu' sarteritis: Operative results and influence of disease activity, Journal of Vascular Surgery, 43(1), 64-71, 206.
7. Miyata T, Sato O, Koyama H, Shigematsu H, Tada Y. Long-term survival after surgical treatment of patients with Takayasu's arteritis, Circulation. 2003;108(12):1474-1480.