ORIGINAL RESEARCH PAPER
Volume - 10 | Issue - 06 | June - 2021 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

A STUDY OF A RARE CASE REPORT OF CHILDHOOD TAKAYASU'S ARTERITIS IN A TERTIARY CARE HOSPITAL.

General Medicine

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KEYWORDS

INTRODUCTION:

Takayasu's arteritis is a type of vasculitis disorder- a group of rare disorders – that causes inflammation of large blood vessels, which in turn causes restriction of blood flow and ultimately damage to the vital organs and tissues. The disease commonly affects aorta and its main branches.[1] It can be divided into 6 types, based on angiographic involvement.[2] ~20% patients usually have a monophasic, self-limited disease, while in most other patients, the disease is either progressive or relapsing/remitting and needs immunosuppressive treatment.[3,4,5]

The disease is usually divided into 2 phases:

Systemic phase: in this phase, signs and symptoms of active inflammatory illness are seen. This includes constitutional symptoms- fever, fatigue, weight loss, nonspecific aches and pains, etc. ESR is usually increased in this phase.

Occlusive phase: This phase follows the systemic phase. During this time patients experience symptoms caused by narrowing of the affected arteries-may include claudication, dizziness on standing up, visual disturbances, headaches. Peripheral pulses may not be palpable at this time, and auscultation of great vessels may lead to detection of bruits.

Both these phases are not always distinct, and patients may be suffering from features of both phases at the same time.The current case of Takayasu's arteritis features a young girl aged 12, with seemingly uncommon presentations; namely, involvement of small vessels of the brain thereby defying the standard criteria of the disease.Moreover, the given case shows development of thrombo - embolism in the left pulmonary artery, another uncommon feature early in the disease.

CASE HISTORY:

A 12-year-old Hindu girl belonging to the middle socio-economic class presented with a Total duration of illness = 2 years

The child had presented with constitutional symptoms, namely cough, low grade fever and loss of appetite off and on at presentation The parents had consulted a private hospital where it was treated as fever. Routine blood work up at that time was normal.

During that time (March 2019), patient developed episode of facial deviation to the right and right sided weakness along with involuntary passage of urine and inability to speak for 30 minutes – followed by complete recovery, for which they were transferred to higher centre – Tertiary Care Hospital, Ahmedabad.

Physical examination at Tertiary Care Hospital at that time showed a normal pulse in the right radial artery; while the left radial artery pulsations were not palpable, along with a radio femoral delay with difference in pulse volume Systemic examination showed presence of carotid bruit (more prominent over right carotid) and an abdominal bruit. Cns examination was unremarkable at the time of presentation to the tertiary hospital.

Patient underwent a Ct Brain Plain at that time (April 2019) which was s/o acute infarct in the left GC (ganglio-capsular) region. In view of infarct in a young child, further evaluation was done there.

2D echo done at that time was suggestive of: -
Severe LV dysfunction
EF=15%, along with dilation of all 4 chambers
LA clot of 17*17 mm
Moderate TR + moderate PAH
Thoracic and descending aorta narrowing

A provisional diagnosis of (?) viral myocarditis + DCM + cardiac thrombus was made, and the patient was then started on injectable anticoagulants, diuretics and oral beta blocker therapy, followed by oral anticoagulation.

During the same hospital course at the Tertiary Hospital, patient developed similar episode with complaints of giddiness and transient loss of vision, followed by hypotonia in right upper and lower limbs and a decrease in power to 2/5 in both right sided limbs and absent right plantars.

Therefore, a Ct Angiogram of the Head and Neck and Aorta was done, which showed:

Multi focal intimo- medial wall thickening and luminal irregularities in 2nd part of Left subclavian artery. Mild ostial stenosis in the left vertebral artery.

Intimal-medial thickness of Right osteo proximal common carotid artery with mild luminal narrowing.

Varying degree of circumferential intimo-medial wall thickness with multifocal area of moderate to severe luminal narrowing of descending thoracic and abdominal aorta.

Mild osteo proximal stenosis of superior mesenteric artery. Complete thrombotic occlusion of the left descending pulmonary artery.

MR Angiogram – Brain and Neck was also done, suggestive of-
Acute infarct in left basal ganglia involving caudate head, lentiform nucleus and putamen.Moderate narrowing in distal and proximal segment of left CCA. Moderate narrowing in cervical part of left ICA Mild narrowing in the proximal segment of the right CCA. Complete block in left MCA.

Arterial Doppler of both upper limbs was unremarkable at that time. Ultrasound of the abdomen showed multiple splenic infarcts.

Patient improved on treatment at the tertiary hospital and was discharged on oral steroids, mycofenolate mofetil 360mg twice a day and ecosprin 75mg once a day, along with oral diuretic and ACE inhibitor.

Patient thereafter consulted a private practitioner in the subsequent period, wherein she was started on Inj Tocilizumab (2 doses taken 1 month apart) (given in v/o persistent disease activity – raised ESR, CRP and neovascular involvement on MMF)

Patient then presented to our institute in July 2019 with an episode of tonic posturing of the right upper limb followed by generalised spasm in all 4 limbs a few hours prior to presentation.

Diagnosis of scar epilepsy was then made, and the patient was started

on anti-epileptics. Detailed immunological workup was done and the patient was advised regular tocilizumab injections and follow up.

Patient was then given 4th and 5th doses here 1 month apart, under observation.

During the time of the 6th tocilizumab dose, the patient developed an episode of URTI- due to which the dose was deferred. Thereafter due to constraints in availability of tocilizumab in covid pandemic, the patient was subsequently switched to oral immunosuppressants and maintained on follow up. Patient then came for follow up in March 2021- almost over a year – post covid.

The physical examination at the time of the last presentation (March 2021) showed similar findings like the previous one, with an audible carotid and abdominal bruits, and a residual right claw hand with intact power and reflexes.

2D echo (March 2021):
Normal sized LV with mild systolic LV dysfunction. EF=45%
Global LV hypokinesia.

Suspicion of left subclavian stenosis with (?)Dissection.
Suspicion of stenosis in the suprarenal part of abdominal aorta. Other arteries appear normal.

INVESTIGATION PROFILE:

INVESTIGATIONS	MARCH 2019	JULY 2019	MARCH 2021
HB	10.9	9.2	13
TC	20,000	8350	5790
PC	353	435	338
UREA	62	40	--
CREAT	0.9	0.7	0.5
NA+	130	143	--
K+	4.1	4.3	--
SGPT	43	17	19
SGOT	41	23	--
BILI(T)	0.9	1.6	-
ALP	108	65	--
ESR	80	2	-
CRP	POSITIVE	NEG	0.8(NEG)

INVESTIGATIONS CONTD..	MARCH 2019 CONTD..
TROP I	0.001
PT/INR	19/1.3
D-DIMER	1.3
MP/PC	NEG
DENGUE NS1 AG	NEG
CHICK IGM	NEG
URINE RM	LEUCOCYTE +
BLOOD CS	NO GROWTH
ECG	T INV IN 2,3,AVF
CXR	NAD
SPUTUM AFB	Negative NEGATIVE IN JULY 2019
IMMUNOLOGICAL MARKERS IN JULY 2019	
ANTI CARDIOLIPIN	IgG -3 gpl (NEGATIVE) IgM-<2mpl(NEGATIVE)
B2 GLYCOPROTEIN 1	IgG 1 u/ml(NEGATIVE) IgM-4u/ml(NEGATIVE)
LUPUS ANTI COAGULANT	ABSENT
MPO(p ANCA)	<5 AU/ML(NEGATIVE)
PR3 (c ANCA)	<5 AU/ML(NEGATIVE)

DIAGNOSIS AND DISCUSSION
Based on the history, clinical examination findings and blood and radiological work up, our patient was diagnosed with Takayasu's arteritis.

Takayasu 's disease is a chronic inflammatory disease of large and medium sized arteries, mainly affecting the aorta and its branches. It is a very **uncommon disease**, with an **incidence rate of 1.2-2.6 c**ases per million as per standard texts.[7]

It is most prevalent in adolescent girls and young women.

Criteria to suspect Takayasu's arteritis:
a] A decrease or absence of peripheral pulse
b] A difference of > 10 mm Hg blood pressure in both upper limbs.
c] Arterial bruits.

The diagnosis is mainly confirmed by characteristic pattern on arteriography, which include: Stenosis, aneurysm, irregular vessel wall, post stenotic dilation. Overall survival rates are estimated to be ~>90%.[7] Standard 2d-echocardiography as well as transthoracic echocardiography play a significant role in screening and characterization of arterial lesions in patients of Takayasu's arteritis.[8]

Aortic arch branch vessels and the carotid artery are commonly affected and show diffuse, homogenous wall thickening called the "Macaroni Sign"- visualised on echocardiogram. The descending aorta can also be involved, visualised as stenotic with "pseudo coarctation".[8] Moreover, involvement of pulmonary vessels can also be delineated from echocardiographic studies.[9]

CONCLUSION:
Takayasu's arteritis is also called the pulseless disease due to the difficulty in detection of peripheral pulses that occurs occasionally due to the vascular narrowings. The disease commonly affects women aged <40; with a 9:1 female predominance worldwide. The disease is commoner in Asian women compared to other races.[6]

In the given case scenario, the development of Takayasu's arteritis at a young age, along with significant involvement of cerebral circulation is an intriguing and somewhat rare finding. The development of cardiac thrombi and narrowing of the left pulmonary artery- is another relatively rare documented evidence at young age in the disease. The development of subclavian artery dissection documented echocardiographically-not so uncommon in Takayasu's presenting in the later age groups- in the early decades of life, is another noteworthy finding at the patient's age.

ADDITIONAL INFORMATION:
1] NO CONFLICT OF INTEREST.
2] CONSENT WAS OBTAINED FROM THE PATIENT'S PARENT REGARDING PUBLISHING THE CASE DETAILS AND RELEVANT SCAN IMAGES FOR PURPOSE OF THE STUDY.

*American College of Rheumatology Criteria for Takayasu's arteritis [10]

TABLE II
- CRITERIA OF AMERICAN COLLEGE OF RHEUMATOLOGY FOR THE CLASSIFICATION OF TAKAYASU'S ARTERITIS (Ref. 27)

Criteria	Definition
Age at disease onset in year	Development of symptoms or findings related to Takayasu's arteritis at age <40 years.
Claudication of extremities	Development and worsening of fatigue and discomfort in muscles of one or more extremity while in use, especially the upper extremities.
Decreased brachial artery pulse	Decreased pulsation of one or both brachial arteries.
Blood pressure difference >10 mmHg	Difference of >10 mmHg in systolic blood pressure between arms.

RADIOLOGICAL IMAGES-PATIENT BASED-2019

Image 1

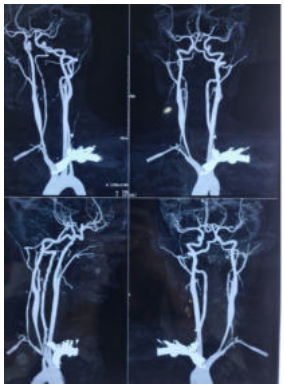


Image 2

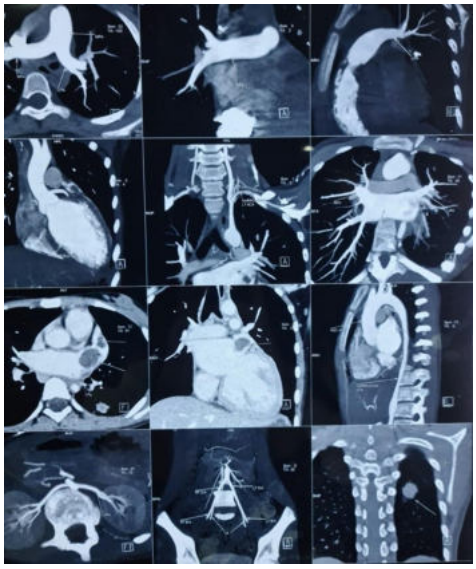
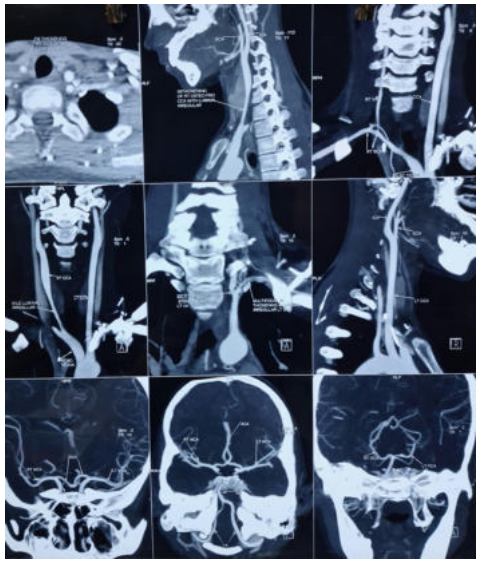


Image 3



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