ORIGINAL RESEARCH PAPER
Volume - 13 | Issue - 02 | February - 2024 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

A REPORT ON CASE SERIES OF RARE VASCULAR DISEASE: TAKASAYU ARTERITIS

General Medicine

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ABSTRACT

Takayasu arteritis is a rare, systemic, inflammatory large-vessel vasculitis of unknown etiology that most commonly affects women of childbearing age. It is defined as "granulomatous inflammation" of the aorta and its major branches. Takayasu arteritis can manifest as an isolated, atypical, and/or catastrophic disease. It can involve any or all of the major organ systems. The disease has been reported in all parts of the world, although it appears to be more prevalent in Asians. We are reporting a series of three cases who were admitted to our hospital with different presentations and were clinically diagnosed with symptoms of Takayasu. They were further investigated and treated, followed by discharge on clinical improvement.

KEYWORDS

Takayasu Arteritis, Pulselessness, Aorto-arteritis

INTRODUCTION

Takayasu's arteritis is a rare (2–6 cases per million per year), inflammatory large-vessel vasculitis of unknown etiology that most commonly affects women of childbearing age of middle Asian origin. It manifests as large-vessel granulomatous vasculitis with massive intimal fibrosis and vascular narrowing prevalent in the aorta and pulmonary arteries. Takayasu arteritis commonly occurs in women younger than 50 years and can manifest as an isolated, atypical, and/or catastrophic disease. It would be interesting to know about its clinical presentations, investigational procedures, management, and outcomes for the noted conditions.

Case Presentation

CASE 1

A 18-year-old female had been hospitalized with complaints of left hemiparesis, left UMN facial palsy, and seizures that were diagnosed as Rt MCA territory infarcts on MRI. She was treated appropriately and discharged. After four months, she presented with Rt carotid and subclavian bruit and pulselessness in the left brachial and radial arteries, which were investigated using a carotid doppler that suggested a 90–100% block in the Rt CCA and 70–80% block in the Lt CCA and severely reduced flow in the B/L ICA with dilatation and turbulent flow with collaterals in the B/L ECA. (Figure-1). Further patient was investigated with coronary CT angiography that suggested 90% block in Rt CCA, 100% block in Lt CCA, 100% block in Lt SCA, 90% block in brachiocephalic, and 80% block in descending aorta, s/o Takayasu Arteritis. (Figure-2) Where the 2D echo was normal. The patient was medically treated with anti-platelets, statins, steroids, immunosuppressants, IVIG, and tocilizumab. PTA was planned, and stenting was done in DTA, BCT, and Rt VA. The patient responded well to therapy and showed drastic improvement in symptoms, after which she was discharged.

CASE 2

A 32-year-old female has been hospitalized with complaints of pain in the RT hypochondriac region; USG shows cholelithiasis (~18 mm); and no pulse in the RT radial artery. CT angiogram of the aorta shows aorto-arteritis involving the aortic arch, descending abdominal aorta, bilateral subclavian renal arteries, the possibility of Takayasu arteritis, and Thoraco- abdominal aortic dissection (Standford type B,Debakey type III). symptomatic treatment taken but not operated. After 1 year, she had again pain in the abdomen due to cholelithiasis and was revisited in the hospital. 2D echo shows mild MR or mild TR. The aortic dissection flap is visible in the ascending aorta. MDCT aorto-angiography was done, and I got the same finding as before. (Figure-3,4) As a part of treatment 2, PRBC was transfused for anemia, and conservative management was done with NSAIDS and antibiotics. The case was not operated on because of the high risk for surgery. and planned to take a biopsy.

CASE 3

A 51-year-old, chronic smoker male has been hospitalized with complaints of pain in the left side, chest pain, abdominal pain, and shortness of breath for 1 month. And no pulses in the left radial artery. CXR PA: soft tissue density mass with base towards mediastinum seen in Lt mediastinal region. 2D Echo: Grade 1 LV diastolic dysfunction. Contrast CT Thorax: Type B fusiform descending aneurysm with eccentric intramural thrombus. (Figure-5) DOR after 7 days of admission.

DISCUSSION

In our hospital, we have a series of three cases over a period of six months that were identified and evaluated for the rare disease Takayasu Arteritis. This case series is reported, considering its academic value as a potentially life-threatening condition. As its exact etiology is unknown, few studies suggest the role of genetics in the pathogenesis of this condition. The diagnosis of Takayasu arteritis should be suspected strongly in a young woman who develops a decrease or absence of peripheral pulses, discrepancies in blood pressure, and arterial bruits. The disease involves medium- and large-sized arteries, with a strong predilection for the aortic arch and its branches; the pulmonary artery may also be involved. The involvement of the major branches of the aorta is much more marked at their origin than distally.

Pulses are commonly absent in the involved vessels, particularly the subclavian artery. The frequency of arteriographic abnormalities and the potential for hypertension occur in 32–93% of patients and contribute to renal, cardiac, and cerebral injury.

Characteristic laboratory findings include an elevated ESR and/or CRP, mild anemia, and elevated immunoglobulin levels. The diagnosis is confirmed by the characteristic pattern on arteriography, which includes irregular vessel walls, stenosis, post stenotic dilation, aneurysm formation, occlusion, and evidence of increased collateral circulation. Complete imaging of the aorta and its major branches by magnetic resonance or computed tomography arteriography should be obtained to fully delineate the distribution and degree of arterial disease.

ARTERY	PERCENTAGE OF ARTERIOGRAPHIC ABNORMALITIES	POTENTIAL CLINICAL MANIFESTATIONS
Subclavian	93	Arm claudication, Raynaud's phenomenon
Common carotid	58	Visual changes, syncope, transient ischemic attacks, stroke
Abdominal aorta*	47	Abdominal pain, nausea, vomiting
Renal	38	Hypertension, renal failure
Aortic arch or root	35	Aortic insufficiency, congestive heart failure
Vertebral	35	Visual changes, dizziness
Coeliac axis*	18	Abdominal pain, nausea, vomiting
Superior mesenteric*	18	Abdominal pain, nausea, vomiting
Iliac	17	Leg claudication
Pulmonary	10–40	Atypical chest pain, dyspnea
Coronary	<10	Chest pain, myocardial infarction

CONCLUSION

Therefore, based on the evaluation of three case series in our hospital,

PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr

it is recommended that patients present with non-specific symptoms of vascular disease. So, we should be more suspicious of rare diseases like Takayasu arteritis.

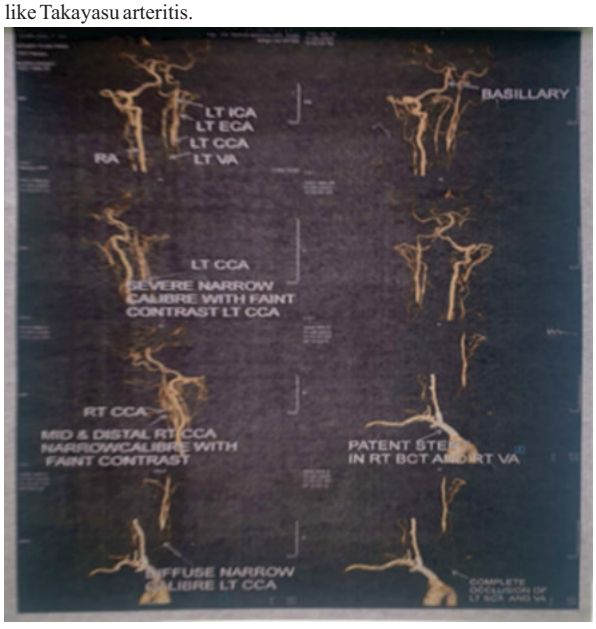


Figure-1

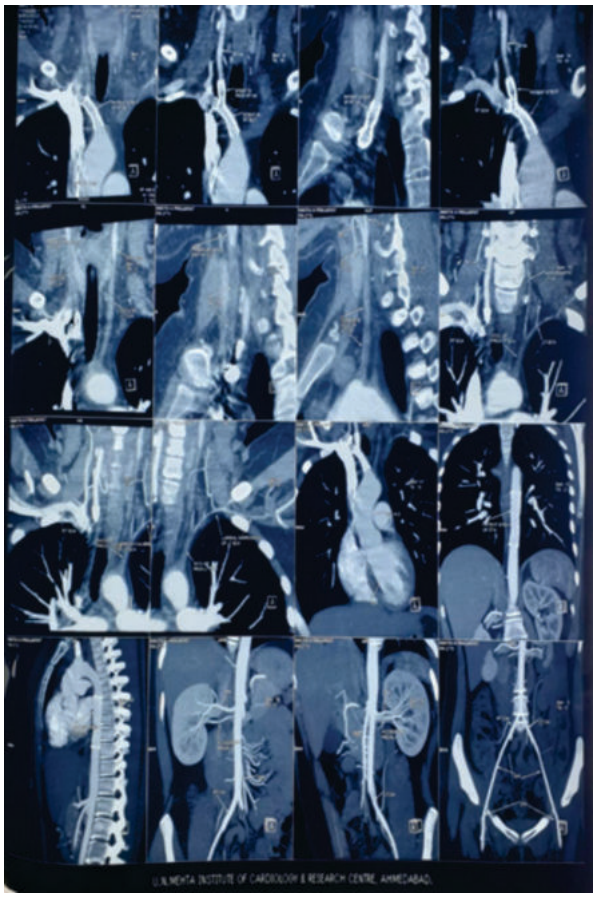
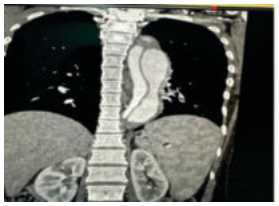


Figure-2



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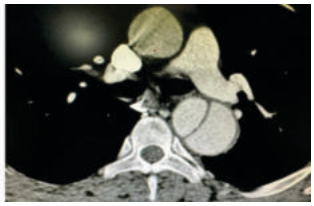

Figure-4

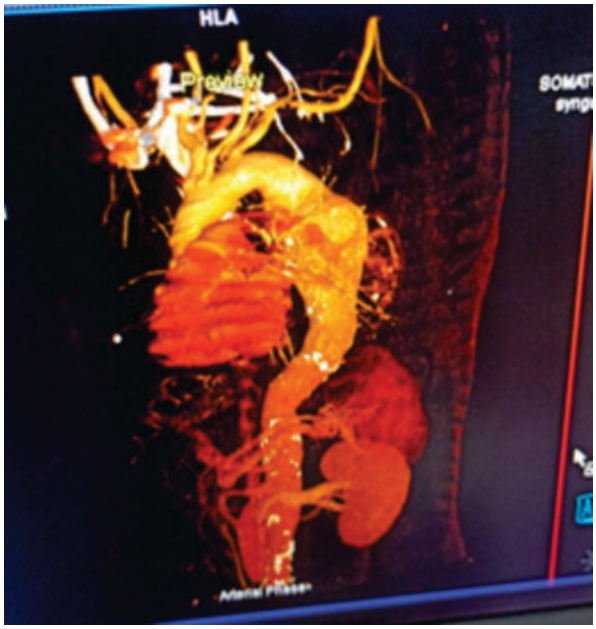

Figure-5

(UMN- Upper Motor Neuron, MCA- Middle Cerebral Artery, CCA- Common Carotid Artery, ICA- Internal Carotid Artery, ECA- External Carotid Artery, IVIG- Intravenous Immunoglobulin, DTA- Descending Thoracic Aorta, BCT- Brachiocephalic Trunk, VA- Vertebral Artery, PRBC- Packed Red Blood Cell, NSAIDS- Non-Steroidal Anti-Inflammatory Drugs, ESR- Erythrocyte Sedimentation Rate, CRP – C-Reactive Protein)

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