Rare Presentations of Takayasu Arteritis: A Case Series

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Abstract

Keywords
► Takayasu arteritis
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► case series

Takayasu arteritis (TA) is a form of large vessel vasculitis that may lead to fibrosis, stenosis, or aneurysm formation of vessels. Its presentation varies depending on the arterial beds involved. We report 3 cases out of around 150 cases of TA with rare initial presentations of brachial plexopathy caused by an axillary artery aneurysm, complicated type A intramural hematoma, and renal artery aneurysms along with abdominothoracic TA presenting as a pulsatile abdominal mass.

Introduction

Takayasu arteritis (TA) is a form of large vessel vasculitis that manifests as limb claudication, stroke, renovascular hypertension, aortic regurgitation, and coronary artery disease, depending on the vasculature involved. However, its diverse and sometimes rare clinical presentation might delay definitive diagnosis and treatment. In this article, we reported three such cases, with the first case presenting as brachial plexopathy due to an axillary artery aneurysm, the second case as a complicated type A intramural hematoma (IMH), and the third case having bilateral renal artery aneurysms presenting as a pulsatile abdominal mass (►Table 1).

Case 1

A 62-year-old male patient with a history of diabetes mellitus, hypertension, and smoking (remote) underwent evaluation for chronic headaches 1 year prior. Imaging studies revealed right vertebral artery occlusion and left subclavian artery occlusion. In the context of coexisting chronic stable angina pectoris, the patient subsequently underwent echocardiography and coronary angiography. These investigations demonstrated annuloaortic ectasia, moderate to severe aortic regurgitation, and near-total occlusion of the left circumflex artery with adequate collateral flow. Considering the patient's preserved left ventricular function and

Table 1 Laboratory parameters of the cases (summary)

	Case 1	Case 2	Case 3
Presentation	Pulsatile mass in axilla Chronic headache	Pyrexia of unknown origin Maculopapular rash	Chronic abdominal pain Pulsatile abdominal mass
Acute ECG changes	No	No	No
Total leucocyte count (/mm^3)	9,200	7,500	19,600
Neutrophil (%)	80	71	91
Erythrocyte sedimentation rate (mm in 1 h)	90	120	10
C-reactive protein	24	16.86	8.36
Blood culture	–	–	–
ANA profile	–	–	–
ANCA	–	–	–
VDRL, RPR, Mantoux test, HIV, HBV, HCV serology	–	–	–
Leptospira serology	–	–	–
Serum protein electrophoresis	Not done	–	Not done
Urinalysis	Normal	Normal	Normal
Ultrasonography of abdomen	Normal	Mild splenomegaly	Aortic aneurysm
PET-CT	+	+	Not done
Immunotherapy	MMF, HCQ	MMF, HCQ	Steroids
Outcome	Improved	Improved	Improved

Abbreviations: ANA, antinuclear antibody; ANCA, antineutrophil cytoplasmic antibody; CT, computed tomography; ECG, electrocardiogram; HBV, hepatitis B virus; HCQ, hydroxychloroquine; HCV, hepatitis C virus; HIV, human immunodeficiency virus; MMF, mycophenolate mofetil; PET, positron emission tomography; VDRL, venereal disease research laboratory.

relatively asymptomatic presentation, medical management was instituted then.

On follow-up, he presented with numbness of the right little and ring fingers with gradually progressive pulsatile axillary swelling (►**Fig. 1A**) for 3 weeks. There was no fever, rash, or other clinical signs of infection. His left brachial pulse was feeble, and his radial pulse was absent. The echocardiogram showed annuloaortic ectasia (ascending aorta 52 mm) (►**Fig. 1B, C**) and severe aortic regurgitation with a mildly dilated left ventricle. Axillary artery Doppler revealed a 42 × 41 mm saccular aneurysm with layered thrombus (►**Fig. 1D–F**) with swirling blood flow within. Computed tomography (CT) aortic angiogram showed a chronically occluded left subclavian artery immediately following the origin of the left vertebral artery (►**Fig. 1G**), the right vertebral artery occluded from its origin (►**Fig. 1H**), and a large 41 mm × 41 mm × 37 mm axillary saccular aneurysm (►**Fig. 1I**) which was partially thrombosed (maximum thickness of 14 mm). Mild focal narrowing of the proximal axillary artery was also noted. Positron emission tomography CT (PET-CT) showed an inflammatory hot spot in the aneurysm wall without any evidence of inflammation in other body parts. His inflammatory parameters are depicted in ►**Table 1**. We advised immunomodulation followed by surgical repair of the aneurysm. However, he was lost to follow-up.

Case 2

A 59-year-old male patient, diabetic, hypertensive, and a reformed smoker, presented with low-grade fever and maculopapular rash for 2 months (►**Fig. 2A**). He had elevated erythrocyte sedimentation rate, C-reactive protein, and other infective workup was negative. Hepatitis and human immunodeficiency virus serology were negative. The autoimmune profile was negative. His blood counts, urinalysis, and blood cultures were negative.

Ultrasound of the abdomen showed mild splenomegaly. Chest X-ray revealed a dilated ascending aorta (►**Fig. 2B**), and an echocardiogram showed a 10- to 12-mm hyperechoic lesion in the ascending aorta wall (►**Fig. 2C, D**). CT aortogram was diagnostic of a supracoronary aortic IMH complicated by Stanford Type A aortic dissection (►**Fig. 2E–G**). PET-CT scan suggested an active inflammatory process involving the ascending aorta, aortic dissection site, aortic arch, and the origin of the right carotid artery (►**Fig. 2H, I**). No other fluorodeoxyglucose-avid loci were noted. He was started on disease-modifying therapy (mycophenolate mofetil 500 mg twice daily and hydroxychloroquine 200 mg once daily), following which fever and rash resolved. Given stable disease, he was managed conservatively and advised to repeat CT after 3 months.

Case 3

A 27-year-old male patient, with recently diagnosed hypertension, on regular medications, presented with chronic abdominal pain for 5 months. The pain was dull aching, localized to the left side of the umbilicus. It was nonradiating and did not aggravate with food intake. There was no associated fever, nausea, vomiting, weight loss, anorexia, jaundice, or dysuria. The patient did not notice any abdominal lump. General physical examination was normal with no pallor, edema, or lymphadenopathy. Blood pressure was
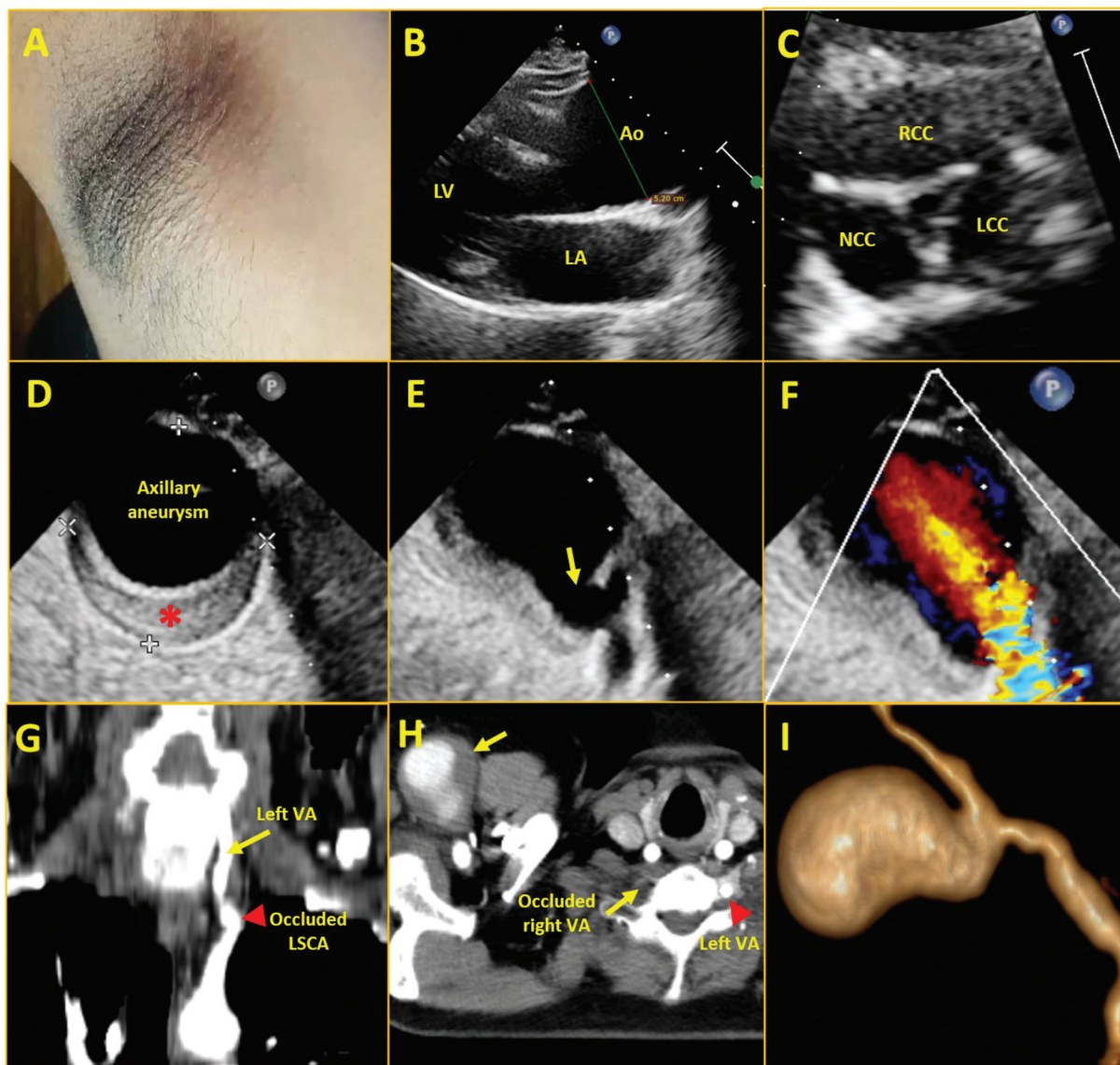
Fig. 1 (**A**) The clinical image of the index patient displays axillary swelling. (**B**) A 2D echocardiogram in the parasternal long-axis view reveals a dilated ascending aorta (52 mm). (**C**) A 2D echocardiogram in the basal short-axis view reveals central noncoaptation of aortic leaflets. (**D–F**) Doppler imaging reveals a 41 mm × 37 mm axillary saccular aneurysm, which is partially thrombosed (asterisk indicates layered thrombus; the arrow indicates the neck of the aneurysm). (**G**) A CT aortic angiogram coronal section showing a chronically occluded left subclavian artery (arrowhead) immediately after the origin of the left vertebral artery (arrow). (**H**) Axial section showed that the right vertebral artery was not visualized, but the counterpart was seen (arrowhead), and a large partially thrombosed axillary saccular aneurysm (arrow). (**I**) 3D volume rendered image was also seen. Ao, aorta; LA, left atrium; LCC, left coronary cusp; LSCA, left subclavian artery; LV, left ventricle; NCC, noncoronary cusp; RCC, right coronary cusp; 2D, two-dimensional; 3D, three-dimensional; VA, vertebral artery.

within normal limits, and all peripheral pulses were well felt. Abdominal examination revealed a 40 × 40 mm nontender and pulsatile mass in the umbilical region to the left of the midline.

The patient underwent an abdominal CT examination for the pulsatile mass. The CT scan revealed an ectatic descending thoracic aorta, a fusiform abdominal aortic aneurysm with wall calcifications from the origin of the superior mesenteric artery to the aortic bifurcation, along with eccentric thrombosis, bilateral renal artery aneurysms, a right subclavian artery aneurysm, and a complete occlusion of the celiac artery (►Fig. 3A–C). He was diagnosed with TA and was started on steroids and other supportive management.

The patient underwent a staged aneurysm repair. The first stage involved a right subclavian artery interposition graft and a polyester graft from the left common iliac artery to the right renal artery. This was followed by an open repair of the aortic aneurysm using a polyester tubular graft, which included the reimplantation of the superior mesenteric artery and an interposition bypass to the left renal artery. Significantly elevated serum amylase and lipase levels prompted a repeat CT, revealing patent grafts and complete exclusion of the aneurysm sac with a normal anastomotic site without contrast leak or narrowing (►**Fig. 3D**). The patient improved symptomatically postsurgery and is currently doing well.
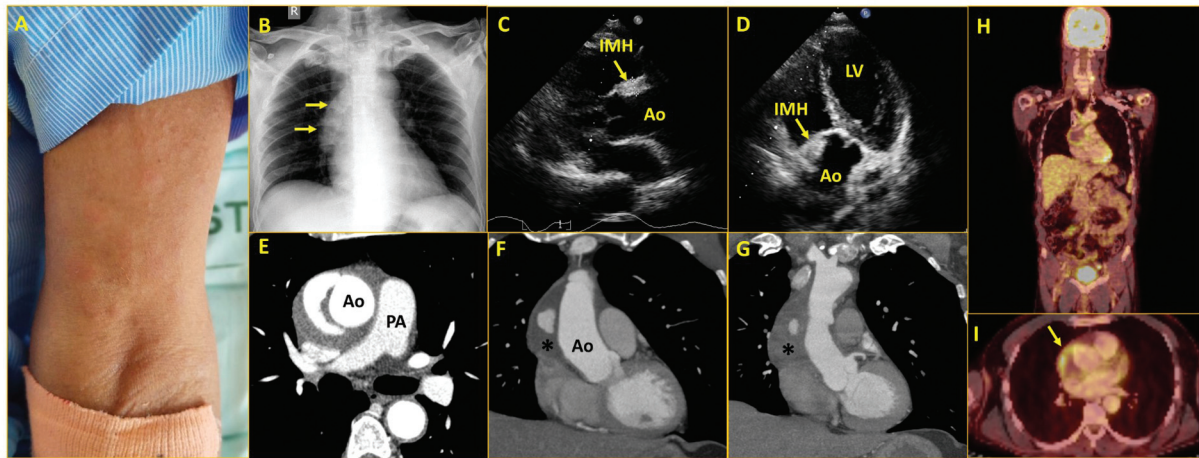
Fig. 2 (**A**) The clinical image of the index patient showing maculopapular rash on the left arm and elbow; (**B**) chest X-ray (posteroanterior view) showing dilated ascending aorta (arrows); (**C**) 2D echocardiogram in parasternal long axis and apical five-chamber (**D**) view showing grossly thickened aortic wall (arrow); (**E, F**) computed tomography aortic angiogram in axial and coronal views showing intramural hematoma (asterisk) complicated by aortic dissection; (**H, I**) positron emission tomography scan suggested an active inflammatory process (hotspots) involving the ascending aorta, aortic dissection site, aortic arch, and the origin of the right carotid artery. Ao, aorta; IMH, intramural hematoma; LV, left ventricle; PA, pulmonary artery; 2D, two-dimensional.
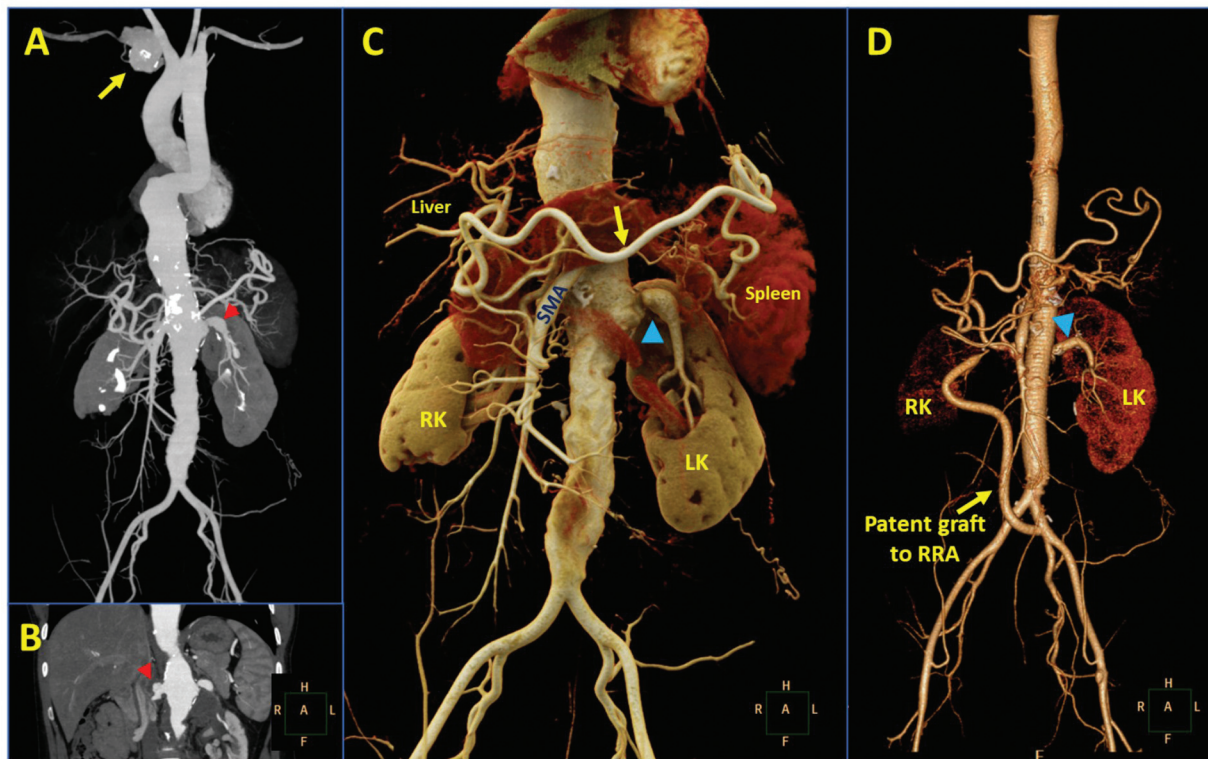

Fig. 3 (**A**) Computed tomography angiogram showing right subclavian artery (arrow in **A**), ectatic descending thoracic aorta and a fusiform abdominal aortic aneurysm with wall calcifications from the origin of the superior mesenteric artery to aortic bifurcation, aneurysms of bilateral renal arteries (arrowhead—left renal artery, [**B**] arrowhead—right renal artery), and celiac artery complete occlusion; (**C**) 3D volume-rendered imaging shows the spatial relationships of major branches of the abdominal aorta. Note that the superior mesenteric artery is providing the collaterals to hepatic, gastric, and splenic bed with prominent gastroepiploic artery (arrow in **C**); (**D**) Postoperative 3D computed tomography showing the patent left common iliac artery to right renal artery bypass (arrow), good flow to superior mesenteric artery, and patent interposition bypass to left renal artery (arrowhead). LK, left kidney; RK, right kidney; RRA, right renal artery; SMA, superior mesenteric artery; 3D, three-dimensional.

Discussion

Our patients met the modified Ishikawa and Sharma et al diagnostic criteria for TA. TA, known as pulseless disease, is a chronic granulomatous inflammatory arteritis that falls under large vessel vasculitis as per the latest revised Chapel Hill Consensus Conference criteria (2012) for vasculitis.[1–3] Patients may present with constitutional symptoms such as fever, malaise, or weight loss early in the disease course. Acute inflammation can destroy arterial media and if left untreated can lead to aneurysm formation.[4] Vascular fibrosis, wall thickening, stenosis, and thrombus formation can occur in the later phase of the disease. Myocardial involvement in TA is rare.[4,5] The median age of TA disease onset is 25 years and is more common in female gender (female to male ratio 8:1).[6] However, the ratio is 1.6 to 2:1 in the Indian population, and older age at presentation is not uncommon. In a group of 88 patients who were followed up on average for 7 years, Subramanyan et al found that 5.7% of them had aortic regurgitation, 15.9% had aneurysm formation (any vascular bed), and 6.8% had a congestive cardiac failure.[7] Arterial aneurysms complicated by compressive neuropathy are rare. One case of brachial plexopathy due to an axillary artery aneurysm managed by resection was reported in a 23-year-old woman.[8] Our patient is a sexagenarian with a much larger aneurysm.

Dermatological manifestations such as erythema nodosum, pyoderma gangrenosum-like ulcers, inflammatory nodules, and rarely purpura have been reported. In a series by Rocha et al, cutaneous involvement was as high as 32% of cases.[9]

Aortic involvement is very frequent. Wu and Zhu presented seven cases of aortic dissection in TA, all being type B aortic dissection. One underwent surgery, and two medically managed patients died at 1 and 5-month follow-up.[10] An autopsy series of 10 patients by Sharma et al comprised one type A aortic dissection patient.[11] However, acute IMH is rare in TA. IMH can be complicated by penetrating aortic ulcer or dissection.[12] A meta-analysis by Chow et al revealed that the 1-year survival rate in type A IMH is similar for patients under medical first versus surgical management, with patients in the former group more likely to require follow-up intervention if their maximum aortic dimension is more than 45 mm.[13] We preferred to follow-up with our patient medically because of the resolution of symptoms and no disease progression.

In TA, steno-occlusive lesions are common, whereas dilatation and aneurysm are extremely rare clinical presentations due to end-organ ischemia and arterial claudication.[14] Renal artery involvement in TA is associated with increased morbidity (chronic renal failure, hypertension, heart failure) without affecting mortality.[15] Although steno-occlusive disease is more common, renal artery aneurysms are extremely rare with only a few cases reported to date.[16,17]

Conclusion

Rare presentations of TA often baffle clinicians, masquerading as life-threatening presentations. Knowledge of these manifestations, early detection, and immunotherapy are critical to improved outcomes in such patients.

Learning Points

- *Accurately diagnose Takayasu arteritis at its earliest stages*, thereby preventing potential delays in treatment and improving patient outcomes.
- *Proactively identify critical complications*, allowing for timely intervention and minimizing the risk of life-threatening events.

Authors' Contributions
S.M. and S.S. contributed to conceptualization; formal analysis; writing—original draft; and writing—review and editing. S.S.K. contributed to conceptualization; investigation; and supervision. N.N., J.R., and S.P. contributed to conceptualization; formal analysis; and writing—review and editing.

Conflict of Interest
None declared.

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