



Interventional Management of High-risk Patients with Takayasu's Arteritis

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Authors' contributions

This work was carried out in collaboration between all authors. Author Sridhar Kasturi conducted the study and procedural work. Author SKB assisted the doctor and data collection. Authors Shivani Kothari and AT performed the technical writing and manuscript preparation. All authors read and approved the final manuscript.

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ABSTRACT

Takayasu's arteritis is a chronic vasculitis affecting aorta and its branches. We report a case series of Takayasu's disease. Percutaneous intervention with stent implantation is better choice for stenosis and occlusion of coronary and carotid arteries as well as peripheral lesions, and is increasingly used for the management of Takayasu's arteritis.

Keywords: Takayasu's arteritis; percutaneous intervention; granulomatous vasculitis; pulseless disease.

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1. INTRODUCTION

TA (Takayasu's arteritis) is a rare, idiopathic, chronic granulomatous vasculitis, which affects aorta and its branches, typically affecting before the age of 40 years. It emerges with insidious onset but has debilitating symptoms. Initially it was believed to involve cervical vessel occlusion [1]. Subsequently this panarteritis, which affects intimal and adventitious region of aorta and its branches became recognized as Takayasu's arteritis. The clinical manifestations are varied and related to the vessel involved, which include aortic arch (pulseless disease), descending thoracic or abdominal aorta (atypical coarction) [2], renal arteries [3], coronary arteries [4] and pulmonary arteries. Some patients are also susceptible to aortic aneurysm [5] and aortic valve regurgitation [6] and ascending aortic dilation. Corticosteroids are usually the first-line treatment. The optimum management is still not known and very few studies have detailed endovascular approach to this disease.

Thus, the aim of our study is to present successful cases of percutaneous intervention in complicated cases of Takayasu's arteritis.

2. CASE SERIES

We present here eight cases of Takayasu's arteritis. The basic details and angiographic profile of patients are given in the (Table 1). The detailed endovascular intervention is described for each patient individually.

2.1 Case Report 1

2.1.1 Interventional management

Shinobi guide wire was crossed and 2.5 mm x 22 mm balloon was introduced in infrarenal portions of aorta for multiple inflations. Shinobi wire was exchanged with 300 cm Terumo 0.35 wire and further dilations of aorta, CIA and EIA was carried out with 6 mm x 38 mm balloon. Abdominal aortic stenting was done with 40 mm x12 mm self expanding Nitinol. Absolute self expanding 8 mm x 100mm stent was implanted in right CIA to EIA. Next day check angio showed well patent aortic stent but slow flow in right CIA stent due to thrombosis and no antegrade flow distally. Proximal portion of right CIA and proximal portion of previously stented portion was implanted with 6 mm x 38 mm Jostent. Right EIA and distal portion of previously stented portion was covered with 6 mm x 58 mm

Omnilink stent to cover dissected flaps. Post procedure angiography (Fig. 1) showed well placed aorta, right CIA, right EIA stents with good antegrade flow without any thrombosis / dissection flaps. Patient was readmitted 9 months later due to massive cerebral infarct and died.

2.2 Case Report 2

2.2.1 Interventional management

Stenting of aorta and right common iliac artery was done with 12 mm x 60 mm and 9 mm x 60 mm self-expanding Cordis smart Nitinol stents through femoral route using stabilizer plus guide wire. Right renal artery stenting was done with 6 mm x 18 mm Genesis stent passed over the stabilizer plus guide wire with the help of 6 Fr right guiding catheter (Fig. 2). Left common iliac bifurcation site was crossed with Shinobi wire through the struts of stented portion of aorta, dilated with 3.5 mm x10 mm and 6 mm x 20 mm balloons and stenting was done with 7mm x 18 mm Genesis stents. Post stenting balloon dilations were carried out with 7mm x 20 mm balloon of bifurcation site. Post stent check aortogram revealed well patent stents and uneventful stay. Follow - up CT angio after 5 years revealed well patent aorta-Iliac stents and right renal artery stent (Fig. 2). Patient is under clinical follow up for the past 5 years without any recurrence of symptoms.

2.3 Case Report 3

2.3.1 Interventional management

Dilations of RCA were done with 2.5 mm x 20 mm balloon using catheter and stabilizer plus guide wire. Subsequently ostium of RCA was stented with 2.5 mm x 8 mm Occam stent and distal RCA with 2.5 mm x 15mm Millennium stent (Fig. 3). Aortoplasty with multiple dilatations were carried out with Mansfield 12 mm and 18 mm balloons. Stenting of thoracic aorta was done with 20 mm x 40 mm BARD Angiomed stent (Fig. 3). A clinical follow up for the past 11 years did not show any recurrence of symptoms.

2.4 Case Report 4

2.4.1 Interventional management

Right renal artery was engaged through 6 Fr R Judkins guiding catheter through right brachial approach to get better alignment & support due

to cranial looking origin of right renal artery with total occlusion just after the ostium. Various attempts to cross with 0.014" BMW and pilot guide wires were failed, ultimately lesion was crossed with 0.014" shinobi guide wire, and serial dilations were carried out with 1.5 mm x 10 mm, 2.0 mm x 15 mm & 3.5 mm x 15 mm balloons. Due to tight lesion while crossing with balloon support, shinobi guide wire tip inadvertently migrated distally this resulted in folding of distal tip resulting in perforation of peripheral branches right renal artery.

After balloon angioplasty check angio showed well dilated proximal portion of right renal artery with good antegrade flow with irregular margins of the proximal right renal artery with extravasation of one of the peripheral branches of right renal artery resulting in loin pain (Fig. 4). Stenting of right proximal portion of right renal artery was done with express stent 5.0 mm x 15 mm (balloon mounted) (Boston scientific). In view of persistent extravasation with loin pain, coil embolization of peripheral branches of right renal artery was done using straight coil 0.014 cm x 1 cm (Cock Company). Initially 2 coils were implanted but due to mild residual proximal extravasation, another straight coil was inserted using 0.21 micro catheter and 0.18 guide wire. Post angio views showed well implanted stent with good antegrade flow without any extravasation (Fig. 4).

Patient developed mild hematomas at brachial & left femoral cath sites; dialysis support was given besides abdominal ultrasound showed hypoechoic collection seen in the right renal subcapsular space measuring 10.6 cm x 3.6 cm in size with thick internal echo. A hypoechoic collection was seen in the posterior para-renal space, extending along the psoas muscle. Hb level dropped from initial 9.1 gm% to 5.7gm%. Two units of blood transfusion was given. Next day patient developed abdominal distension with mild ileus repeat ultrasound showed increased in peri-renal & para-renal collection as compared to earlier with mildly elevated serum bilirubin 2.5mg (Indirect 2.0, direct 0.5) with normal liver enzymes but serum creatinine fell down to 2.8mg/dL after post stent dialysis support. Repeat check angio of renal artery showed well patent stent without any extravasations. 36 hours later significantly improved urine output with fall of serum creatinine from initial 7.7gm% to 1.1 % without any dialysis support from second post stent day onwards. Patient also had relief of abdominal pain, recovery of ileus, resumed oral fluids, ambulated from 4th day onwards with

normal serum creatinine & normal urinary output. Medications maintained were Tab. Azoram 50mg daily, Tab. Wyslone 40mg, daily with well controlled Hypertension on metoprolol 100mg and Amlodipine 10mg. Patient was discharged in a stable condition with total relief of symptoms on 6th day of post procedure period.

After one year follow up CT-Aortic angio showed stent in situ in the proximal right renal artery with instant narrowing causing moderate stenosis of about 50% at the ostium (Fig. 4), minimal right posterior perinephric collection, small left kidney with occlusion of ostium of left main renal artery and collaterals from aorta supplying the distal artery, total occlusion of celiac artery at the ostium and proximal superior mesenteric artery with distal reformation by collaterals from inferior mesenteric artery, occlusion of proximal left subclavian artery and collaterals from left internal iliac artery supplying the distal subclavian artery. Patient was under clinical follow up for the past 1 year without any recurrence of symptoms.

2.5 Case Report 5

2.5.1 Interventional management

Stenting of right common carotid artery was performed through femoral route using stabilizer guide wire. Distal protection device (DPD) Spider – FX 6.0 mm (EV3) was placed in distal internal carotid artery. Dilations were carried out with 4.0 mm x 2.0 mm balloon and stenting of right CCA was done with 9.0 mm x 6.0 mm (Fig. 5). Left subclavian artery lesion was crossed with stabilizer guide wire, predilated with 4.0 mm x 20 mm balloon (Fig. 5). Then proximal left subclavian was stented with 5.0 mm x 18 mm Racer (Medtronic balloon mounted) at 12 to 16 atms inflations. Complete SE self expandable stent extending from origin of right CCA to distal bifurcation site of right CCA into right ICA & right ECA (Fig. 5). Post stenting dilations were carried out with 7.0 mm x 18 mm balloon. Post procedure angio view showed excellent end result without any complication. Patient was under clinical follow up for the past 1 year 4 months without any recurrence of symptoms.

2.6 Case Report 6

2.6.1 Interventional management

Long segment lesion of thoracic and abdominal aorta was crossed with Shinobi wire using 2.0 mm x 12 mm balloon support and careful serial

dilations were carried out with 2.0 mm x 12 mm, 3.0 mm x 38 mm, 4 mm x 32 mm and 7.0 mm x 40 mm. Then two 14 mm x 80mm Zilver 635 Vascular Stent (Self-Expandable) deployed in descending thoracic aorta extending from proximal site just distal to left subclavian artery origin to distal thoracic aorta with minimal overlap of the stents. Another 14 mm x 50 mm Zilver 635 Vascular Stent (Self-Expandable) was deployed distal to second stent extending up to abdominal aorta just above the renal arteries (Fig. 6). Post dilatation done with 15 mm x 25mm Advance 35LP PTA balloon at 6-8 atms. Post procedure stay was uneventful and good antegrade flow (Fig. 6). After 1 month follow up CT-angio revealed well patent stents with good antegrade flow. Patient was under clinical follow up for the past 8 months without any recurrence of symptoms.

2.7 Case Report 7

2.7.1 Interventional management

Patient underwent PTA with stenting of RVA and left ICA (Fig. 7). Planned FFR guided PCI with stenting of LAD. The procedure was performed through left femoral artery with guiding catheter and guide wire. DPD was kept distal to the lesion to prevent further ischemic events. Left internal carotid artery (LICA) stenting was done with 6-9 mm x 40mm Crisallo Ideale self expandable stent INVATEC (Medtronic). After stenting distal protection device was removed (Fig. 7). Right vertebral artery (RVA) lesion predilatation done with 3 mm x 15 mm balloon, stenting done with 5.0 mm x 18 mm x 130 mm Racer stent (Medtronic) (Fig.7). Post angio showed good TIMI-III flow. Patient was under clinical follow up for the past 7 months without any recurrence of symptoms.

2.8 Case Report 8

2.8.1 Interventional management

Procedure was performed through right femoral approach using 6 Fr sheath. Aortogram was performed using 6 Fr pig tail catheter introduced over the exchange length 0.035 Terumo wire which revealed significant narrowing of thoracic aorta and long segment narrowing of entire abdominal aorta with severe stenosis just above the renal artery level. Terumo wire exchanged with 0.035 cm x 260 cm amplatz guide wire, serial dilatations of thoracic and abdominal aorta were carried out with 8 mm x 59 mm balloon and 10 x 40 mm Mustang balloon (Boston scientific) (Fig. 8.2). A 6 Fr pig tail catheter was re introduced through left radial route for check angio views. After serial balloon dilatations through femoral approach 12 Fr long sheath (Cook Company) was introduced over amplatz wire. Thoracic aorta was stented with Wallstent-uni Endoprosthesis (Boston scientific) 22 mm x 45 mm x75 cm (Fig. 8.2). After stenting of thoracic aorta long sheath was pulled down to just below the level of superior mesenteric and celiac artery and check angio was performed prior to abdominal aortic stenting. Stenting of entire abdominal aorta starting from the level of superior mesenteric artery and extending in to right common iliac artery using 14 mm x 90 mm x 75 cm Wallstent-uni Endoprosthesis (Boston scientific) (Fig. 8.2). Post stenting aortogram was performed with pig tail catheter placed from right radial route showed well patent thoracic, abdominal aorta and aorta-iliac bifurcation. Post stenting balloon dilatations were carried out with 12 mm x 40 mm Mustang balloon (Boston scientific) (Fig. 8.2) and 14 mm x 4 cm balloon (XXL Vascular large diameter).Final check aortogram revealed well expanded thoracic and abdominal aortic stent with good antegrade flow both iliac arteries.

Table 1. Case details

Case no.	Basic demographic details	Baseline characteristics including laboratory investigations	Angiographic and other diagnostic reports
1	35-year- old male Comorbidities and symptoms: DM, claudication of both legs, impalpable lower limb pulses	HR - 86/min BP - 118/86mmHg Hb - 11gm S Cr - 0.9mg%	4 cm long occlusion of right CIA (Fig. 1A), thrombus in left SCA, calcified thrombus in LV, total occlusion of aorta ECG - partial RBBB, 2D echo - RWMA of anterior wall and LV clot

Table 1 continued.....

2	14-year- old male Comorbidities and symptoms: Severe HTN, severe headache, convulsions, claudication of both legs, impalpable right radial and both lower limb pulses	BP - 220/130mmHg	Proximal occlusion of right CIA and total of left CIA (Figs. 2A, 2B), 70% stenosis of right RA including ostium, total blockage of left RA, 70-80% narrowing of abdominal aorta
3	25-year- old male Comorbidities and symptoms: Cardiorespiratory arrest, Severe LV dysfunction with moderate MR, TR, PAH		Total occlusion of distal RCA (Fig. 3A), totally occluded right SCA, 50% occluded left SCA, 40% left CCA, 70% narrowing of thoracic aorta 2D echo - RWMA of inferior and posterior lateral wall and LV clot
4	18- year-old female Comorbidities: Renovascular HTN, ARF with acidosis	BP - 210mmHg Left upper limb BP- 170/100mmHg	Totally occluded celiac axis, superior mesenteric left SCA and both RA, Doppler study- total occlusion of left subclavian (Fig. 7B), celiac axis (Fig. 7C), superior mesenteric and both renal arteries (Fig. 7A), left kidney is non functional
5	20-year- old male Comorbidities and symptoms: Transient ischemic attack, syncopal attacks, giddiness, loss of vision in left eye	ESR - 120mm/hr	Totally occluded Left CCA (Fig. 5A), 99% tight lesion of Left SCA (Fig. 5B), long segment lesion of right CCA (Fig. 5C)
6	47-year- old male Comorbidities and symptoms: HTN, DM, alcoholic, ARF, oliguria, puffiness of face, paraparesis of both limbs	Hb - 8gm% ESR - 65mm/hr CRP - 42 mg/L S Cr - 1.2 mg/dL Serum urea - 33mg/dL	Long thrombus from distal arch of aorta, descending thoracic aorta and upper abdominal aorta (Fig.6A)
7	46-year- old female Comorbidities and symptoms: HTN, DM, posterior cerebral artery stroke, giddiness and vomiting since a week	PR - 107/min, BP - 210/120mmHg RBS - 325mg/dL	CAG - double vessel disease, RAG – stenosis of left renal artery, right vertebral artery- 70%, LICA 90% lesion (Fig. 7B), Carotid doppler - plaque in left carotid bulb (Fig. 7A), No flow in left vertebral artery (Fig. 7C)
8	54-year- old female Comorbidities and symptoms: HTN, episodes of chest pain, shortness of breath, headache, claudication and dragging of both lower limbs, feeble lower limb pulses	Hb - 10.4 gm% RBS - 108mg/dL Serum urea - 34 mg/dL	Calcified plaques in arch of aorta (Fig. 8.1 A), mild narrowing of left SCA, proximal descending aorta (Fig. 8.1 B), mild stenosis of descending thoracic aorta (Fig. 8.1 C), multiple calcified plaques in abdominal aorta (Fig. 8.1D)

DM - Diabetes Mellitus, HR - Heart rate, BP - Blood pressure, Hb - Haemoglobin, S Cr - Serum creatinine, CIA - Common iliac artery, SCA - Subclavian artery, LV - Left ventricle, RBBB - Right Bundle branch block, RWMA - Regional wall motion abnormality, RA - Renal artery, MR - Mitral regurgitation, TR - Tricuspid regurgitation, PAH - Pulmonary hypertension, RCA - Right coronary artery, CCA- Common carotid artery, ARF- Acute renal failure. ESR- Erythrocyte sedimentation rate, HTN - Hypertension, CRP - C-reactive protein, LICA - Left internal carotid artery, RBS - Random blood sugar, PR - Pulse rate

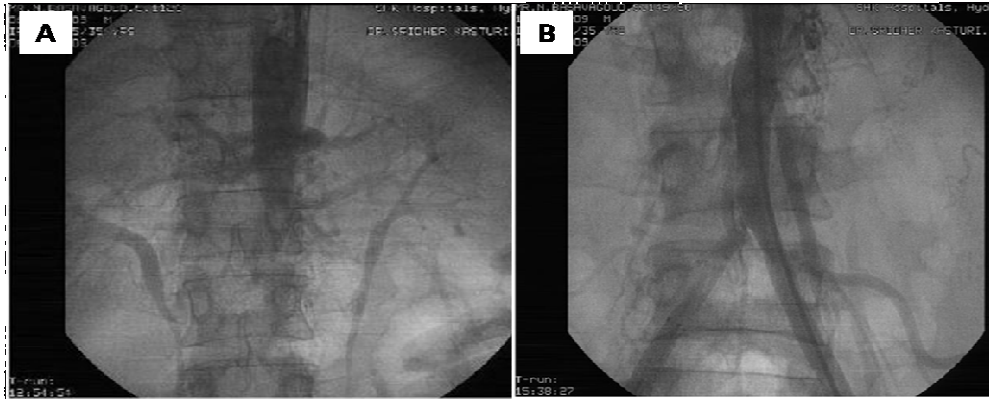


Fig. 1. Pre - and post - procedural angiogram of case 1

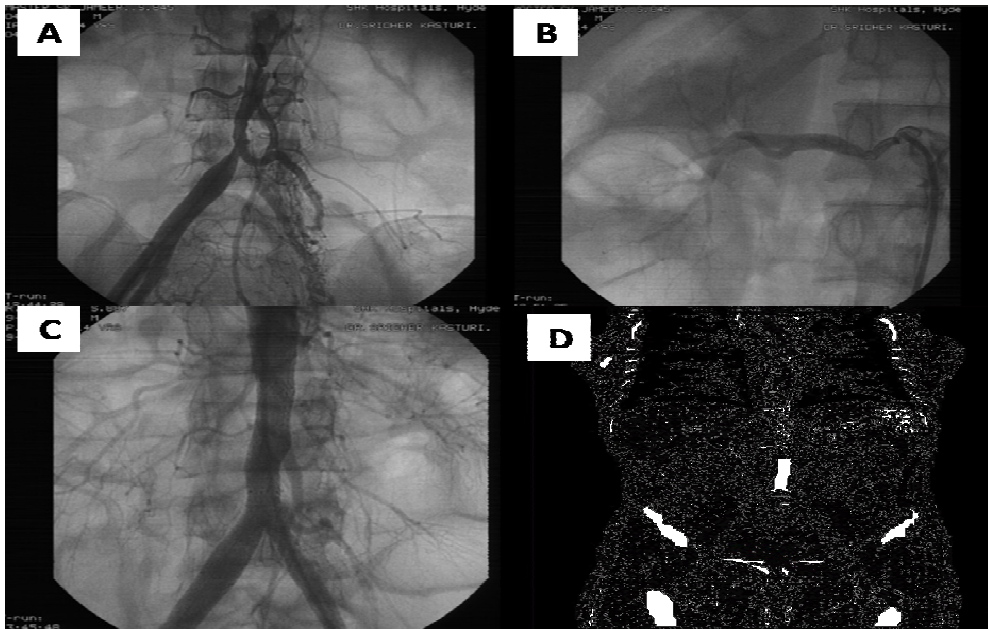


Fig. 2. Pre - and post - procedural angiogram of case 2

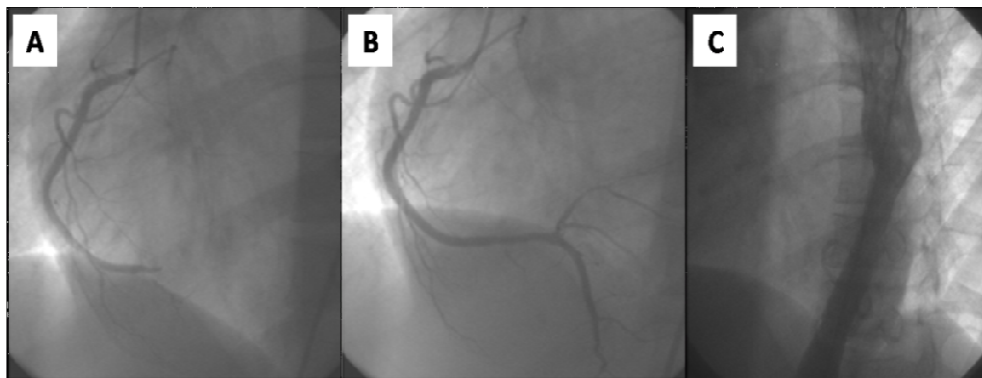


Fig. 3. Pre - and post - procedural angiogram of case 3

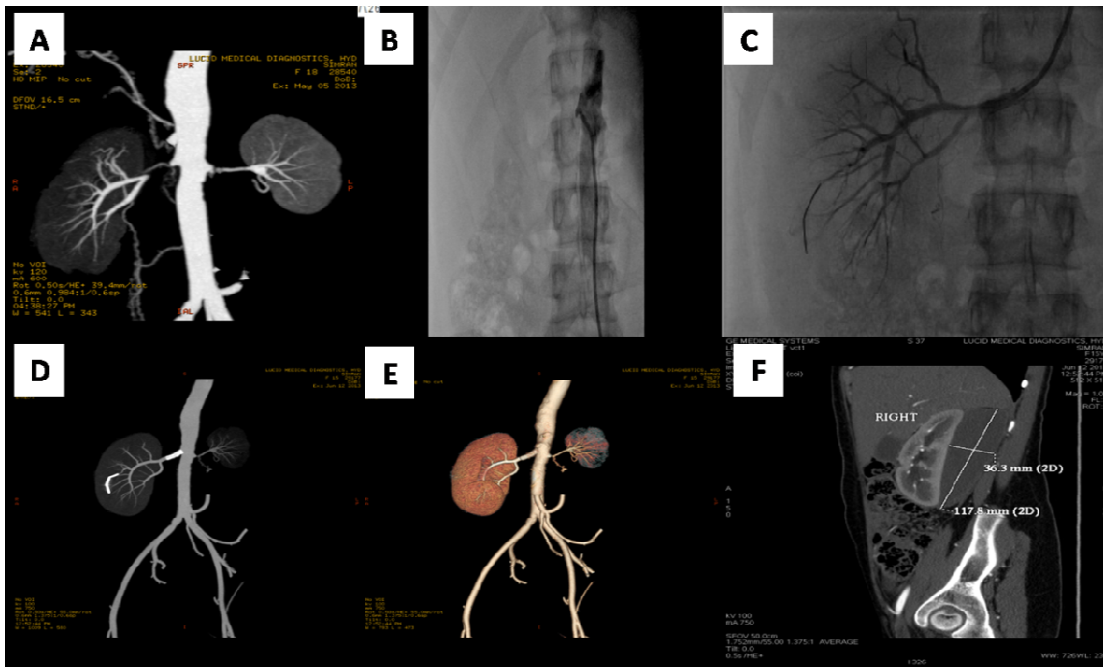


Fig. 4. Pre - and post - procedural angiogram of case 4

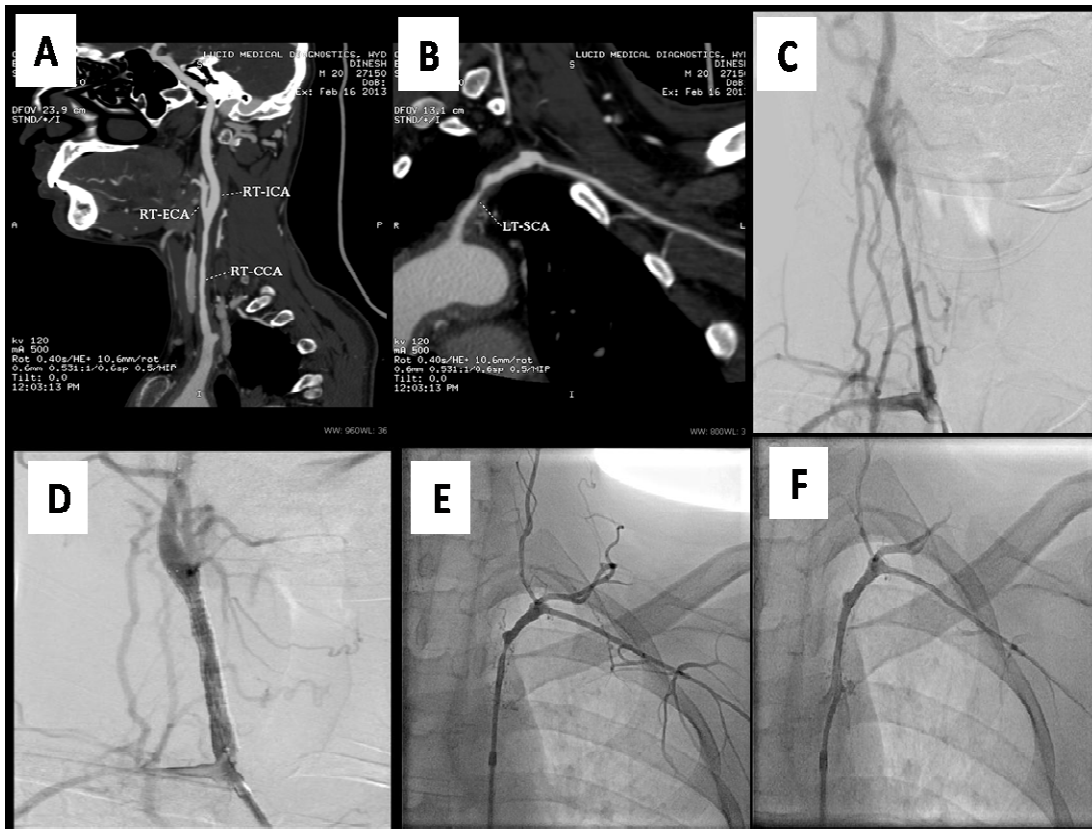


Fig. 5. Pre - and post - procedural angiogram of case 5

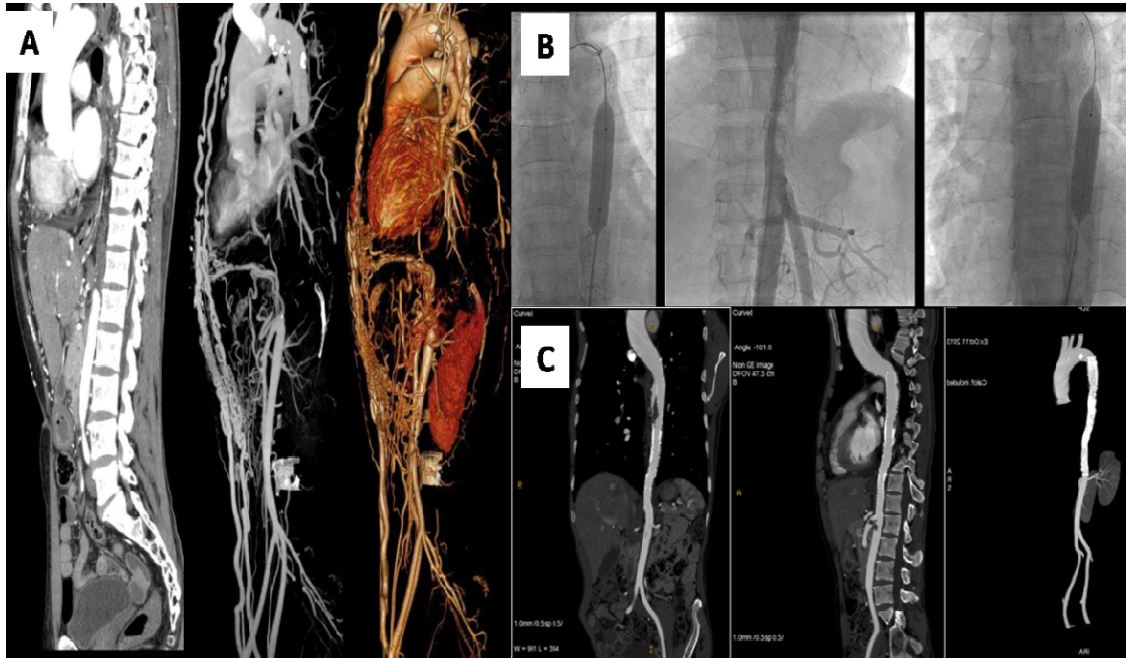


Fig. 6. Pre - and post - procedural angiogram of case 6

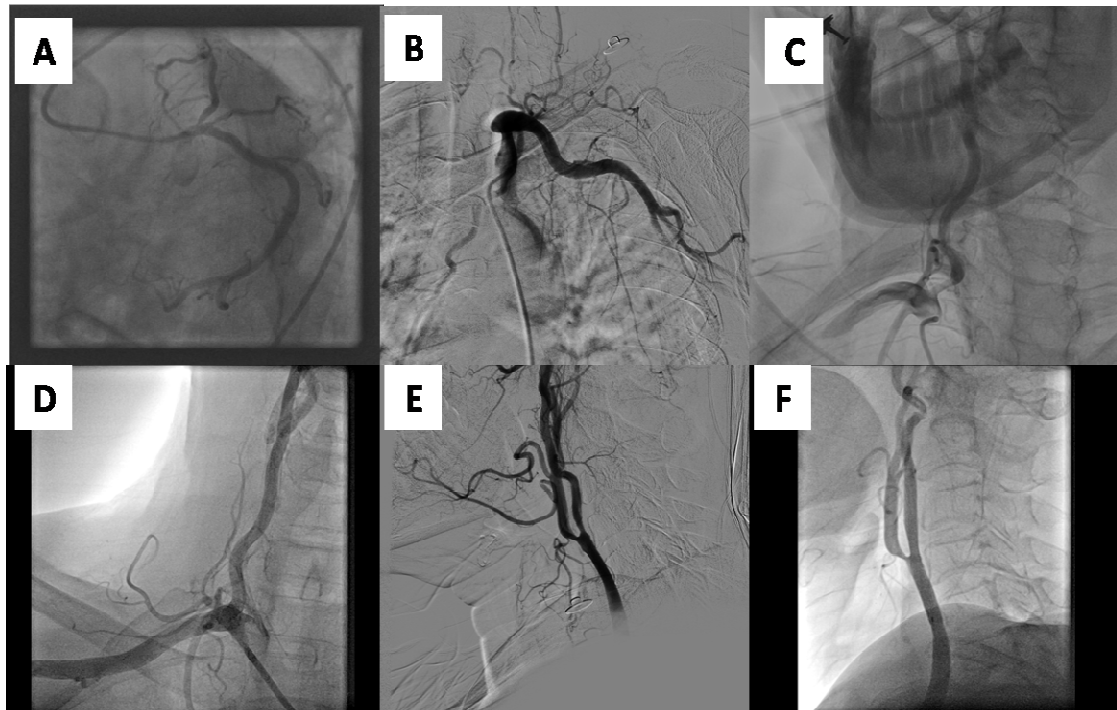


Fig. 7. Pre - and post - procedural angiogram of case 7

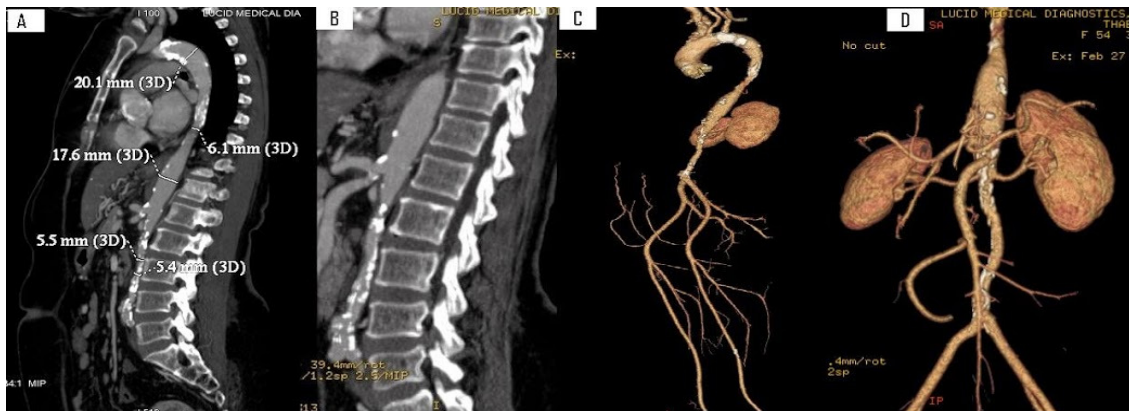


Fig. 8.1. Pre - procedural angiogram of case 8

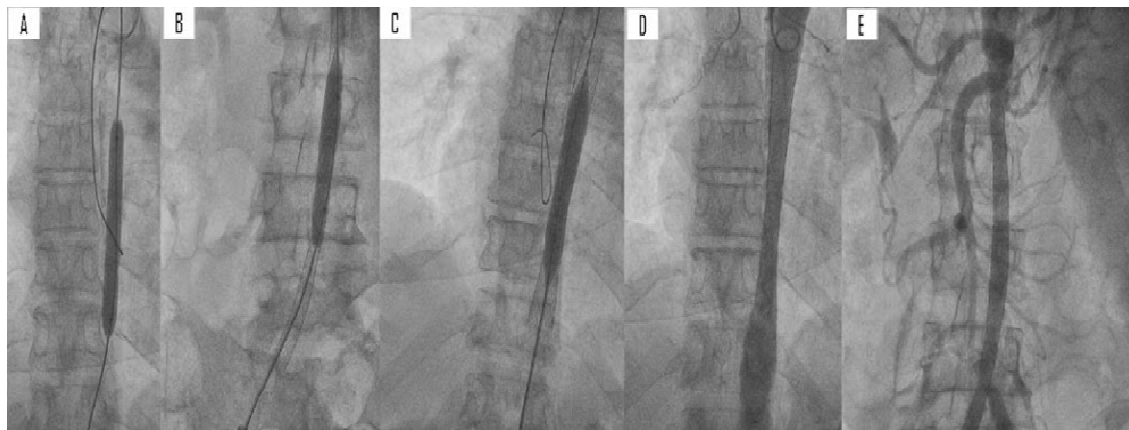


Fig. 8.2. Post - procedural angiogram of case 8

3. RESULTS AND DISCUSSION

Takayasu's arteritis is also known as "pulseless disease" and involves the aortic arch and its branches and presenting symptoms are based on involved vasculature. Symptom onset is insidious and two-third of patients lack significant symptoms early in the disease. Cardiac features such as aortic regurgitation and heart failure occur in 40% of all patients but angina is less common. A long-term follow-up study from India reported survival rate of 91% at 5 years and 84% at 10 years, with event free survival of 74.9% and 64% [7].

The three major clinical diagnostic criteria for the disease includes lesion in left mid subclavian artery, right mid subclavian artery, and signs and symptoms for duration greater than one month [8]. The ten minor criteria are increased erythrocyte sedimentation rate, pain or tenderness in the carotid artery, high blood pressure, aortic regurgitation or ectasia of the

aortic ring, lesion in the pulmonary artery, lesion in the left mid common carotid artery, distal lesion in the brachiocephalic trunk, lesion in the descending thoracic aorta, lesion in the abdominal aorta, and lesion in the coronary artery [9]. In our study, the diagnostic criteria were longer duration lesion of subclavian artery and other minor criteria like increased ESR, lesion in carotid artery, lesion in brachiocephalic artery and lesion in abdominal aorta. The ESR is a proven test for detecting disease [10] but still question arises regarding its efficacy because of histological findings of specimen obtained during surgery and of sequential angiographic studies [11].

The treatment aspect of Takayasu's arteritis is not yet established and still faces vagueness to some extent. High dose corticosteroids are the mainstay of treatment. Steroids taper the systemic symptoms and acts as barricade for disease progression. Adjunctive immunosuppressant including cyclophosphamide

and azathioprine can be given if there is difficulty in the withdrawal of steroids. However in chronic stage, the clinical treatment does not suffice and creates room for surgical treatment. The ability of guide wire to cross the lesion determines the rate of success. Many of the cases reported in our study also faced difficulty in crossing of the lesions. The chronic process induced and diffused disease associated with inflammatory process is culprit for failure of recanalization. The diffuse, multifocal and ostial vessel involvement makes percutaneous intervention method of choice rather than surgical revascularization. Percutaneous angioplasty with stent implantation has been successful in treating obstructive carotid, renal and peripheral lesions in TA especially in patients at high surgical risk [12]. In our study also we present cases of successful percutaneous intervention. PTA results in good TIMI-III flow and well patent stents in post angio check. The first case of multiple stenting in all branches of supraortic arch without the involvement of renal arteries with a two year follow had a favorable outcome [13].

There have been few long term studies of PTA and most studies used balloon angioplasty and not stents. A Restenosis rate with angioplasty alone approaches 20% with initial success rate greater than 85%. Although there have been limited studies, angioplasty with stent placement has a restenosis rate closer to 10% and success rate approaching 100%. Small discrete lesions, after inflammation has subsided, are most favorable for PTA. Thus interventional procedures should not be taken lightly in patients with TA. Surgery can increase long term survival of patients with complicated (comorbidities and progression of disease) disease of Takayasu's arteritis [14,15].

4. CONCLUSION

Takayasu's disease is a slowly progressive form of large vessel vasculitis that can lead to severe complications including strokes, angina, renal failure and limb ischemia. The optimal management of symptomatic stenosis is not clear. Outcome of patients with complicated Takayasu's arteritis can be improved by endovascular approach.

CONSENT

All authors declare that 'written informed consent was obtained from the patient (or other approved

parties) for publication of this case report and accompanying images.

ETHICAL APPROVAL

All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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