

## Retraction of Hypertension in a Young Woman with Middle Aortic Syndrome after Stent Implantation

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DOI: [10.36347/sjmcr.2020.v08i06.003](https://doi.org/10.36347/sjmcr.2020.v08i06.003)

Received: 25.05.2020 | Accepted: 07.06.2020 | Published: 11.06.2020

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### Abstract

### Case Report

A young hypertensive female presented to the cardiology department with complaints of episodic headache of 7 months duration. On examination she had accelerated hypertension and absence of lowerlimb pulses. The electrocardiogram and echocardiogram showed left ventricular hypertrophy. The conventional aortography of the patient revealed atypical coarctation in the descending aorta at the level of diaphragm. The coarctation segment was of  $13.4 \times 3.4$  mm in length and diameter and a luminal diameter stenosis of  $>70\%$  on quantitative analysis. During intervention, the lesion was predilated with a  $7 \times 40$  mm carotid stentballoon at 8 atm for 10 sec and later with  $14 \times 60$  mm TYSHAQ balloon at 6 atm for 15 sec. A  $7 \times 80$  mm Cordis S.M.A.R.T CONTROL self expanding stent was implanted across the lesion. Post stent implantation, the hemodynamic parameters have improved. There was no stent migration or restenosis detected in the follow-up angiogram.

**Keywords:** Accelerated hypertension, atypical coarctation, aortogram, descending aorta, TYSHAQ balloon, Cordis S.M.A.R.T CONTROL.

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## INTRODUCTION

Middle Aortic Syndrome is a rare condition with a frequency of 0.5-2% in the general population [1]. The characteristic feature of this clinical entity is the narrowing of the abdominal or distal thoracic aorta. Of the branches of the aorta, the renal and visceral arteries are commonly involved followed by the celiac and superior mesenteric arteries. The inferior mesenteric arteries are rarely involved [2, 3]. The symptoms vary based on the severity and the extent of narrowing of the aorta and visceral vasculature [4]. It usually manifests as refractory hypertension, or with the claudication of the lower limbs, mesenteric angina [2]. On literature review very few cases have reported the survival age of patients beyond 40 years with this clinical entity [5]. If left untreated, this condition leads to life-threatening complications like renal or cardiac failure or intracerebral haemorrhage [6]. The etiology of this syndrome is congenital, genetic with mendelian inheritance i.e., neurofibromatosis, Williams syndrome, or acquired secondary to takayasu arteritis and fibromuscular dysplasia [7].

## CASE

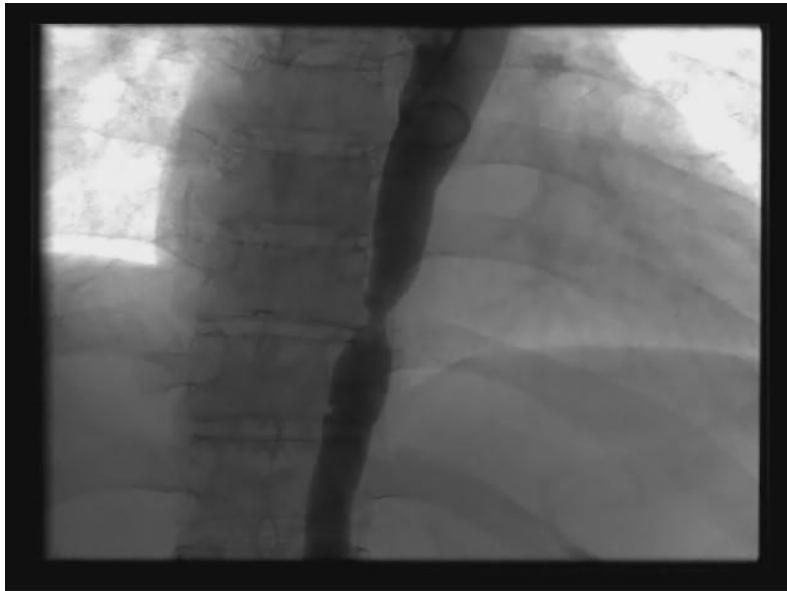
A 27 years old female hypertensive for three years with episodic headache and a 7-month history of amenorrhea and accelerated hypertension on triple antihypertensive drugs, who had delivered a stillborn baby, was referred to us for control of hypertension. On physical examination, there was no evidence of facial dysmorphism and lower limb pulses were absent. There was a difference of 10mmHg of SBP between right and left upper limb. Fundus examination suggested stage I hypertension-induced retinopathy. Blood and urine profiles were normal. Ultrasound evaluation revealed no organomegaly and ruled out the tumor and renal parenchymal disease. Electrocardiogram showed left ventricular hypertrophy. Echocardiogram confirmed left ventricular hypertrophy with grade II diastolic dysfunction. Aortogram of the patient through the right radial approach with 6F pigtail catheter revealed the coarctation in the descending thoracic aorta at the diaphragm (atypical site) with a diameter of 3.4mm and a length of 13.4 mm and luminal diameter stenosis of  $>70\%$  (Figure-1). On Computed tomography (CT)

angiography, there was coarctation at atypical site, diffuse renal artery disease, total block of the mid segment of superior mesenteric artery, and mild disease of the proximal part of the iliac arteries.

Balloon angioplasty was done through the right femoral approach using a 7F long femoral sheath. The lesion was crossed with 0.035 extra stiff wire. The lesion was dilated with a 7x40mm carotid stent balloon at 8 atm for 10 sec and later with a 14x60 mm TYSHAQ balloon (BVM Medical limited, Leicestershire, Leicester, UK) at 6 atm for 15 sec. CORDIS S.M.A.R.T. CONTROL (Cordis Vascular Stent system, Santa Clara, California, U.S.) self expanding stent was deployed (Figures 2 and 3). After this procedure, the peak systolic gradient decreased from 100 to 40mmhg, Coarctation site diameter

increased from 3.4 to 7.4mm and percentage stenosis decreased from 70 to 50%. Despite the residual gradient, no further dilatation was done as deployment to normal size may result in aortic rupture and also because staged correction with further dilatation at a later date after healing may be a better approach. After at two months of follow-up, both lower limb pulses were felt and blood pressure was controlled on triple antihypertensive drugs Follow up aortogram (figure 4) showed increase in diameter of the coarctation segment to 8.4 mm from 7.4 mm and percentage stenosis decreased from 50 to 42% on using quantitative analysis software.

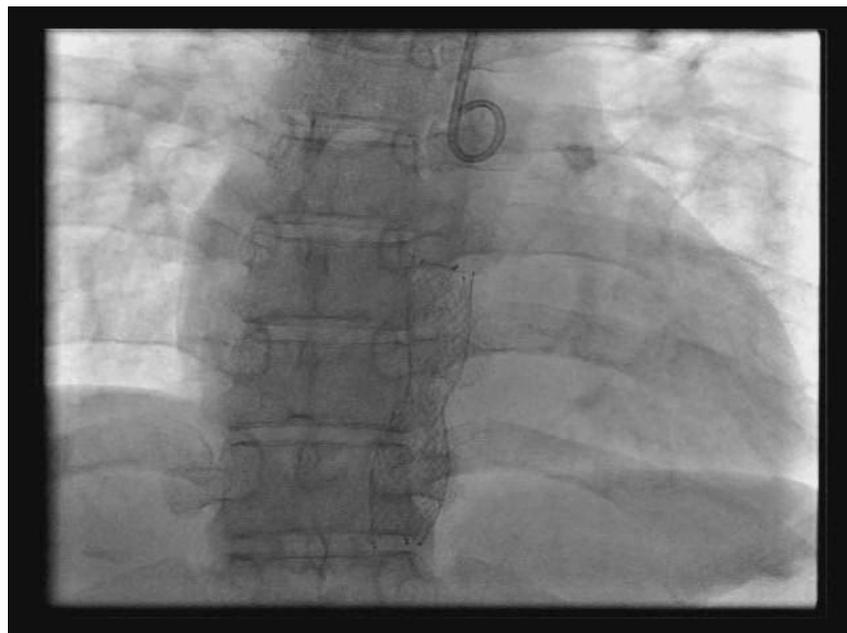
The automated pressure gradient calculated showed decreased in gradient to 36mm Hg. No stent migration and no restenosis were detected.



**Fig-1: Aortogram showing significant stenosis in the thoracic aorta**



**Fig-2: Aortogram showing stent deployment**



**Fig-3: Aortogram showing the deployed stent**



**Fig-4: Aortogram showing good result**

## DISCUSSION

Onset before 40 years, abnormal arteriogram, absence of inflammation or symptoms of myalgia, pleuritis, pericarditis, fever and rashes, and observation of no neurological and dermatological abnormalities, and, normal range of CRP and ESR levels of our patient suggest a congenital type of etiology [8, 9]. Management modalities of this syndrome include aorto-aortic bypass, prosthetic or autologous venous graft vascular reconstruction, or percutaneous transluminal angioplasty. In the Indian context, Sen *et al.*, [10] from Bombay in 1964 have reported 16 patients with the middle aortic syndrome in whom blood pressure was controlled by surgical bypass and

nephrectomy. As 75% had a history of tuberculosis, these patients were also maintained on antituberculosis and corticoid regimen. From Calcutta in 1979 Gupta has informed the survival of five patients out of seven who undergone corrective surgery [11]. Difficulty in fashioning an appropriate graft and extensive nature of vascular abnormality may be responsible for high surgical mortality [9]. Unresponsiveness of the hypoplastic segment to balloon dilatation, aneurysm formation, dissection of the aorta, pseudoaneurysm, and rupture of aorta leading to death are the disadvantages in balloon dilatation [9]. Controlled dilatation of stenotic segment and limited intimal injury are advantages reported for stent implantation [9]. In our

case, stent implantation after balloon dilatation improved hemodynamic parameters and controlled blood pressure. Further, observation of no stent migration and restenosis as revealed by follow-up angiogram, suggested the successful management of aortic syndrome with endovascular stent implantation. To the best of our knowledge, this case serves as a reference for aortic syndrome managed invasively with stent implantation in the Indian context.

## CONCLUSION

Middle Aortic Syndrome is rare in the general population which can be either congenital or acquired. It manifests as uncontrolled hypertension with varying involvement of the renal and superior mesenteric arteries. Surgery is the usual treatment of choice, though individual cases like the one reported can be managed successfully by percutaneous approach.

**Acknowledgements:** We acknowledge the cath lab, echo room and nursing staff of ICCU in the Department of Cardiology, Osmania General Hospital.

No funding sources for any of the authors.

No conflicts of interest by the authors.

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