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CASE REPORT

Incidental finding of coarctation of the aorta in a Marfan patient: a case report

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Abstract Aneurysm of the ascending aorta with coarctation of the aorta is a rare find in the literature. We report a case of a huge ascending aorta aneurysm in a patient of Marfan's syndrome and associated coarctation of the aorta. The patient underwent staged procedure in the form of balloon dilatation for the coarctation of the aorta and a successful Bentall procedure for the aneurysm of the aorta with a brief period of deep hypothermic circulatory arrest (DHCA). Two-staged approach is the ideal management for such cases amenable for endovascular repair.

Keywords Marfan's syndrome · Deep hypothermic circulatory arrest · Bentall's procedure

Introduction

Marfan's syndrome is an autosomal dominant disorder of connective tissue with a myriad of cardiovascular abnormalities along with other systemic pathologies. The association of coarctation of the aorta is a rare occurrence with Marfan's syndrome and represents an incidental finding in majority of cases reported in literature. Here, we present a 26-year-old male having this combination.

Aim To provide an overview of management for such rare patients using surgical and endovascular modalities.

Case report

A 26-year-old male presented in our emergency department with recent-onset gradually worsening chest tightness and palpitations for the past 2 months.

He was in NYHA-III status at the time of presentation. On clinical examination, he had features of Marfan's syndrome. He was tall with positive thumb and wrist sign and high-arched palate. Ophthalmological examination revealed a dislocated lens in his left eye on slit lamp examination. Ankle brachial pressure index (ABPI) for the right limb was 0.78 and for the left limb was 0.68, signifying moderate disease. Radial pulses were normal but both femoral pulses were weak. Cardiac examination revealed lateral displacement of apical impulse and on auscultation, a grade 4/6 early diastolic murmur was noted along the left sternal border and in the interscapular area. The chest roentegram showed enlarged cardiac silhouette with rib notching. ECG showed features of left ventricular hypertrophy and left axis deviation. No other family member had similar illness.

Echocardiography revealed situs solitus with dilated ascending aorta of size 7.2 cm and aortic annulus of 2.9 cm. Ejection fraction was 50%. There was severe aortic regurgitation with a tricuspid aortic valve morphology. Coarctation of the aorta with a gradient of 80 mmHg was present. There was

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mild eccentric mitral regurgitation with anterior mitral leaflet prolapse.

Computed tomography angiography (CTA) (Fig. 1) revealed fusiform aneurysmal dilatation (7.5 × 7.9 cm) of the ascending aorta. The length of the aneurysm was 11.6 cm with no evidence of intraluminal thrombus. The coronaries appeared normal. Post-ductal short-segment stenosis of 7.9 mm was seen. Pre-stenotic diameter was 20 mm and post-stenotic diameter was 27.7 mm. There were multiple collaterals around the bilateral scapulae. There was no evidence of dissection.

He was planned for a staged procedure after discussion with the cardiologist and the cardiac anaesthetist with the aim to reduce the complexity of the operative procedure and improve patient's outcome. Using a percutaneous retrograde catheterization technique, an end-hole catheter was advanced past the site of coarctation to the aortic isthmus and pullback pressures recorded. The anatomy of the coarctation was noted and balloon dilatation (Fig. 2 pre-procedural coarcted segment) was done. Post-procedural (Fig. 3 post-dilatation of coarcted segment of aorta) gradient came down to 15 mmHg. He was started on antihypertensive (beta blocker) to prevent rupture of the aneurysmal sac. Surgical correction of the aneurysm was planned 5 days post procedure. A median sternotomy was done. Cardiopulmonary bypass was established via right axillary cannulation with end-to-side graft and bicaval cannulation. Aortic clamp was applied proximal to the innominate artery. On inspection, a huge aneurysm was present measuring around 8 × 7 cm extending from the aortic root to the arch of the aorta just proximal to the innominate artery. The aortic valve was tricuspid with non-coapting leaflets. These correlated with CTA and echocardiographic findings. A Bentall procedure was carried out using 27 Saint Jude aortic valved conduit (SJM™ Masters Series Aortic Valved Graft) under moderate hypothermia. After completing the proximal anastomosis and checking

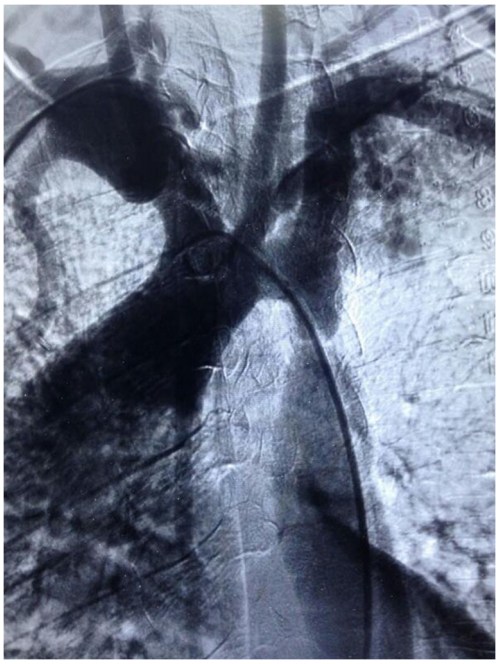

Fig. 2 Pre-procedural coarcted segment

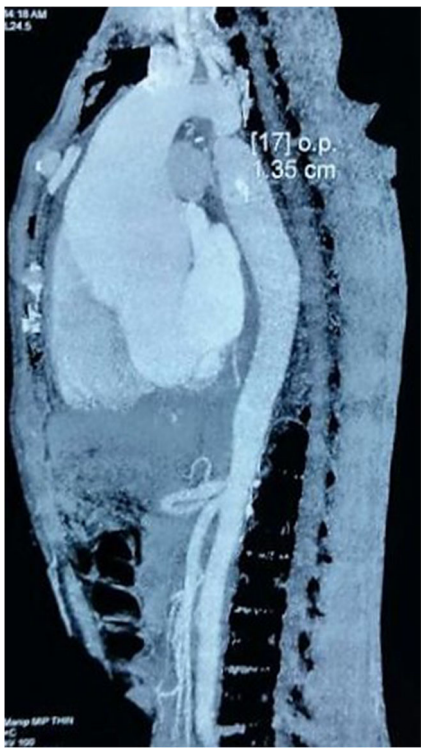

Fig. 1 CTA showing coarctation with ascending aorta aneurysm

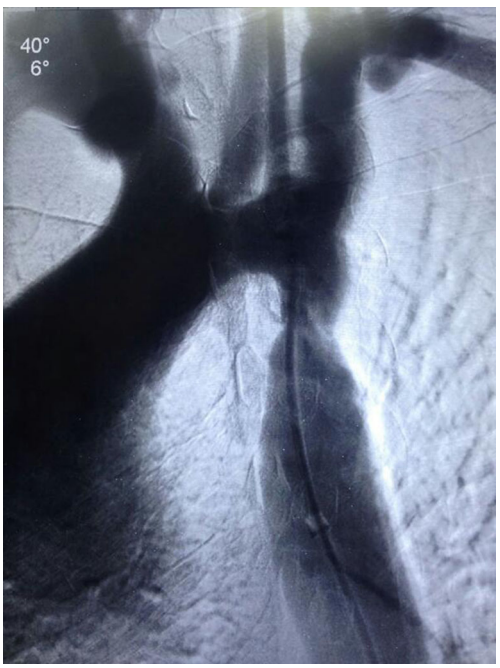

Fig. 3 Post balloon dilatation of coarcted segement

the adequacy of coronary buttons, we proceeded with the distal anastomosis. Cross clamp was applied across the brachiocephalic, common carotid and left subclavian arteries and selective antegrade cerebral perfusion through the axillary artery was started. As the aneurysm was extending up to the brachiocephalic artery, and we needed adequate distal stump for open distal anastomosis, so deep hypothermic circulatory arrest (DHCA) was initiated. Aortic cross clamp was removed and open distal anastomosis was done using prolene 3-0 suture in two layers. The distal anastomotic suture line was reinforced with Teflon felt due to poor tissue strength of the aortic wall in Marfan's syndrome. The aortic cross clamp was 120 min and total bypass time was 145 min. The DHCA time was 9 min.

Post-operatively, he required support in the form of adrenaline at 5 mic/kg/min and nitroglycerin at 1 mic/kg/min, which were gradually tapered. Peripheral pulses were good. Gradient was 10 mmHg at the time of discharge. The patient is in regular follow-up in our outpatient department and presently is in NYHA class I.

Discussion

Marfan's syndrome is a genetic disorder of connective tissue with marked abnormalities of ocular, skeletal and cardiovascular systems. Genetic mutation occurs in FBN1 gene coding fibrillin on chromosome [1]. This leads to inability of fibrillin to bind calcium and resultant weakness. Major and minor criteria have been described in order to decide which patients are at risk and need surveillance. Great majority with Marfan's syndrome are familial. The presence of a dilated aorta plus ectopia lentis is sufficient to give an unequivocal diagnosis of Marfan's syndrome. In the cardiovascular system, the most common features include mitral valve prolapse, mitral regurgitation, aortic insufficiency and aneurysms of the ascending and descending aorta which may progress or be complicated by dissection [2]. Our patient fulfils the criteria for Marfan's syndrome and fortunately he presented early with good ventricular function.

Coarctation of the aorta is presumed to be a less frequent condition in Marfan's syndrome and it is rarely accompanied by true aneurysm in the proximal part of the aorta [2]. After extensive search, only 15 cases were found to have been reported in literature till date. Their prevalence in isolated forms is 3 per 10,000 live births. Though there is coexistence by chance, associations have been suggested. According to an old concept [3], there may be a special tissue in the ductus arteriosus that leads to obliteration of the lumen after birth. Abnormal spread of this tissue involving the isthmus of the aorta causes coarctation. In Marfan's syndrome, defective connective tissue formation accounts for non-limitation of this specialized tissue into the isthmus, leading to coarctation of the aorta. In an another theory, there is disorder of involution at the junction of the embryonic left fourth and sixth foetal arches and together with the traction by fibrosing ductus arteriosus on the aortic arch causes a deleterious effect upon the aortic arch containing abnormal tissue [4]. Coarctation superimposed on the Marfan's syndrome predisposes the fusiform aneurysm to rupture and further dilates the aorta to dissect. In a clinicomorphological study of 18 necropsy patients by William C. Roberts et al. [5], out of 16 patients with fusiform ascending aorta aneurysm, 1 patient had coarctation of the aorta and of the previously reported 110 necropsy patients, 10 had aortic isthmic coarctation of mild degree. In adults with Marfan's syndrome, aortic regurgitation is rarely present when aortic diameter is less than 40 mm and almost always present when it is more than 60 mm [6].

The treatment of fusiform ascending aneurysm in Marfan's syndrome is aimed at elimination or prevention of aortic regurgitation and in preventing aneurysmal rupture, not at preventing dissection, because the latter is an infrequent complication. In a series of 212 patients of Marfan's syndrome with fusiform aneurysm of the aorta by Gott and his colleagues [7], it was inferred that modified Bentall's procedure remains the operation of choice with valve sparing surgery in selected patients. The histological analysis of excised aortic leaflets in the same study revealed diminished fibrillin and microfibril degeneration adding caution to valve sparing surgery, thus adding proof to their conclusion. In another study [1] of 56 patients with aortic root replacement in such patients, it was deduced that successful employment of the Bentall operation is considered being the single most important factor for the dramatic improvement in prognosis of such patients as seen over the past 3 decades. In their study, the mean size of the ascending aorta was 6.6 ± 1.4(3.8–12). Our patient had a diameter of 7.5 cm which was strikingly on the higher side. Gott and his colleagues [7] reported a mortality of 1.5% in elective surgery versus 2.5% in urgent surgery. The emergency mortality was 11.7% despite surgical correction.

In a study carried out in four patients of ascending aorta aneurysm with coarctation by Sampath et al. [8], three of them underwent staged surgical procedures for correction. The early and late results of balloon coarctation angioplasty have been described as successful in different studies. Also, operating on cardiac defect without addressing the significant coarctation may lead to significant underperfusion of organs distal to the coarcted segment, leaving the left ventricle with severe pressure load because of congestive heart failure and hypertension [9]. Nonetheless, staged surgical procedures increase the morbidity, post-operative stay, post-operative pain, lung issues and bad cosmesis. Single-staged repair has also been devised by Vijaynagar et al. [10] in 1980 where the ascending aorta was bypassed to the descending aorta by placing the graft to the left margin of the heart. But these extensive procedures require stringent cannulation and perfusion

measures to prevent neurological problems and the dreaded spinal cord ischemia. Also, kinking and stenosis may occur with such long grafts. With the advent of interventional cardiology, we avoid such potential complications where coarctation can be dealt by balloon angioplasty/stenting and the aneurysm can be repaired surgically in the second stage.

Conclusion

A more aggressive surgical approach is required when coarctation with aneurysm of the aorta is present in a Marfan patient as they are not without life-threatening complications of dissection and rupture owing to their poor tissue strength. We believe that initially treating with balloon dilatation/stenting followed by a Bentall procedure is the optimal surgical management in such patients. Since they are at risk of developing new aneurysms in the future, long-term clinical and radiological follow-up with yearly measure of aortic dimensions is deemed necessary.

Compliance with ethical standards

Funding This study has not been funded by any party whatsoever.

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval This article does not contain any studies with animals performed by any of the authors.

All procedures performed in study involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from all individual participants included in the study. Additional informed consent was obtained from all individual participants for whom identifying information is included in this article.

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