

Case Report

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Unusual Case of Annuloaortic Ectasia - A Case Report

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ABSTRACT

Annuloaortic ectasia is a rare cardiovascular condition characterized by aneurysmal dilatation of aortic annulus and ascending aorta resulting in aortic insufficiency. As the ascending aorta enlarges, the risk of aortic rupture or dissection also rises. As per our knowledge, ascending aorta aneurysm more than 9cm without dissection or rupture has scarcely been reported.

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Introduction

Annuloaortic ectasia (AAE) is a rare cardiovascular condition, traditionally accounting for approximately 5-10% of cases with aortic insufficiency [1]. Its signature combination of aneurysmal dilatation of ascending aorta and aortic annulus forms an imminent threat of dissection or rupture that necessitates rapid diagnosis and management. While the most common variant of AAE is idiopathic, there are pathologies of the aorta such as Marfan syndrome, Loeys-Dietz syndrome, aortitis, Turner Syndrome and congenital heart diseases that can have associated aneurysmal root dilatation. It presents in fourth to sixth decade of life as a pear-shaped, symmetric enlargement of proximal ascending aorta that spares the aortic arch, with concomitant aortic regurgitation (AR) [2]. Aortic dissection is a common and dreaded complication of annuloaortic ectasia, seen in up to 44 percent of patients while aortic rupture can occur in up to 74% of patients [3,4]. In addition,

various comorbidities, hemodynamic fluctuations, multiorgan ischemia and postoperative complications make these cases one of the most difficult challenges for an anesthesiologist. We hereby report an unusually dormant form of AAE in which the ascending aorta was found dilated up to 9.0 cm with no predisposing risk factors and no sign of aortic dissection or aneurysm rupture.

Case Report

The patient was a 46 years old male farmer presenting with exertional dyspnea (NYHA Class III), angina and palpitations for two weeks with no history of pre-existing comorbidities. Clinical examination revealed an elevated heart rate and an early diastolic murmur (grade 3/6) along the left parasternal border. Chest radiograph showed significant cardiomegaly with a prominent aortic knuckle. 12 lead electrocardiography (ECG) highlighted the presence of left ventricular hypertrophy (LVH) with ST segment elevation in anterior and inferior leads with atrial fibrillation (AF) [Figure 1].

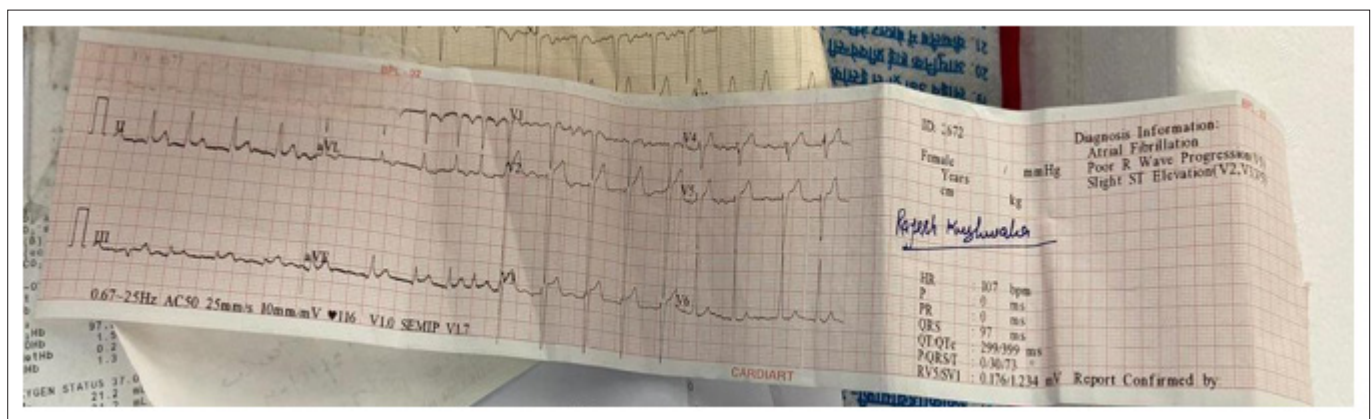


Figure 1: ECG Showing LVH with ST Segment Elevation with AF

2D transthoracic echocardiography (TTE) evaluation showed marked aortic dilatation with maximal ascending aorta diameter of 9.0cm and aortic annulus of 3.5cm with no signs of aortic dissection [Figure 2]. Mild concentric hypertrophy of the left ventricle with an ejection fraction of 56 % was seen. Severe aortic regurgitation with a holodiastolic flow was noted with peak jet width of 10mm.



Figure 2a and 2b: Echocardiography showing annular dilatation and aortic regurgitation

The computed tomography angiography (CTA) assessment revealed a fusiform dilatation of ascending aorta with normal flow of contrast suggestive of aneurysm with a maximum diameter of 9.5cm [Figure 3]. The left main coronary artery appeared to have a short course before dividing into its branches near the aortic root.

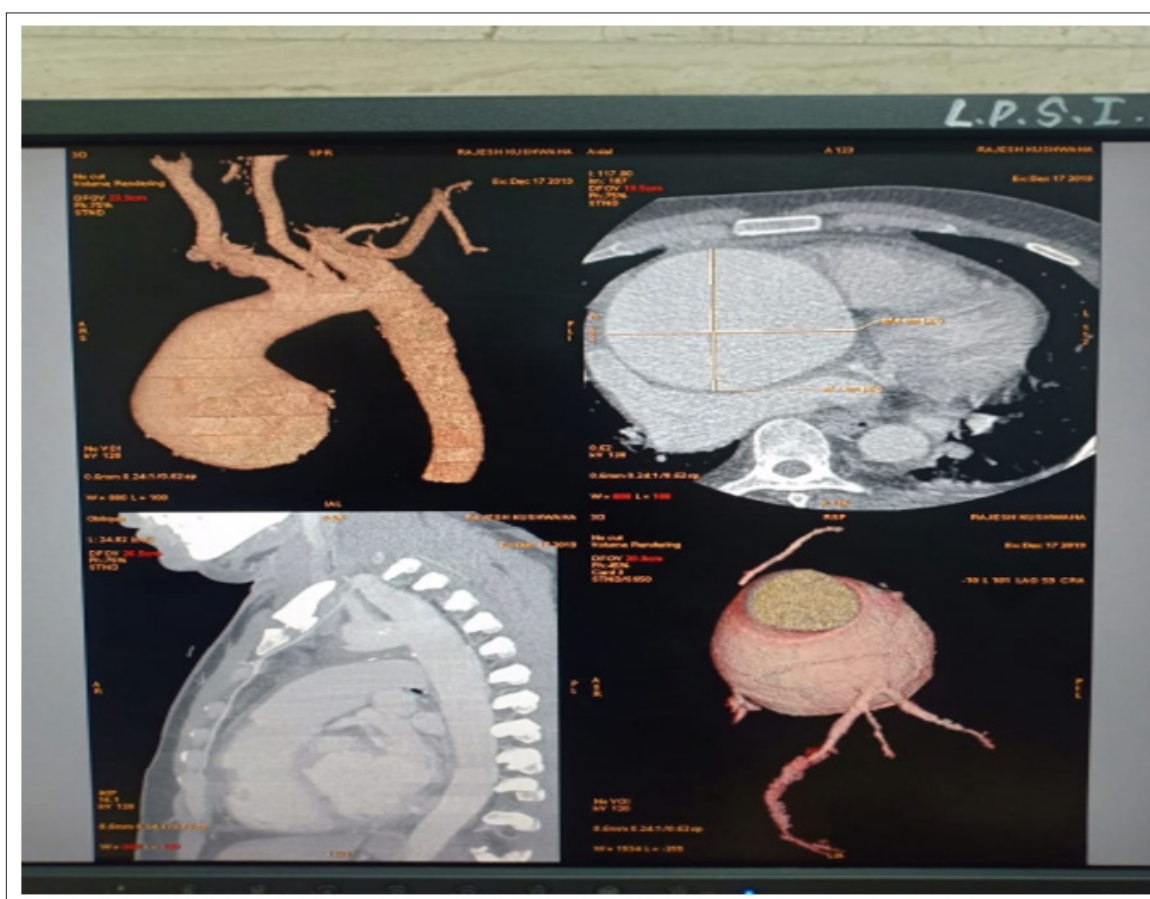


Figure 3: Ascending Aorta Dilatation and Short Course of left main Coronary Artery

The patient was planned for modified Bentall's surgery, after a thorough preoperative assessment, serum investigations and informed consent. As per standard monitoring guidelines, pulse oximetry probe, non-invasive blood pressure monitoring cuff and ECG electrodes were attached to the patient. Large bore peripheral venous intravenous access was secured with a 16G cannula under local anesthetic coverage. Premedication, as per institutional protocol, was given intravenously in the form of glycopyrrolate 0.2mg, fentanyl 1mg/kg and midazolam 1mg. Cannulation of left radial artery for invasive pressure monitoring and right internal jugular vein for central venous pressure monitoring was ensured under local anesthesia. Intravenous (i.v) etomidate 0.3mg/kg along with rocuronium 1mg/kg and fentanyl 5 mg/kg was used injected slowly for a smooth and uneventful induction. Denitrogenation by intermittent positive pressure ventilation using 100% inspired oxygen for three minutes was followed by endotracheal intubation with an appropriately sized tube. Other standard monitors such as end tidal capnography (EtCO₂), left femoral arterial cannula, transesophageal echocardiography (TEE) probe and nasopharyngeal temperature probe were inserted. The patient was ventilated intraoperatively using volume-controlled

ventilation tailored to result in an EtCO₂ level of 30-35 mmHg with a conventional mixture of oxygen and air (fractional inspired oxygen concentration of 0.6). Total i.v anaesthesia was used with continuous infusion of vecuronium and propofol to maintain anesthetic depth with intermittent fentanyl bolus of 1 mg/kg every 30 minutes for analgesia. The hemodynamic parameters were maintained within 20% of the baseline and the intraoperative blood glucose level was maintained between 140-180 mg/dL. Heparin 300U/kg was given for anticoagulation to achieved an activated clotting time (ACT) \geq 400 seconds.

Cardiopulmonary bypass (CPB) was instated using Maquet HL20 (Maquet, Wayne, NJ) after priming with 1000 ml lactated ringer solution, 500 ml of plasma expander, 5000 IU heparin, 20 gm mannitol, 50 ml sodium bicarbonate, and furosemide as a diuretic as per institutional protocol. Aorta was cannulated distal to aneurysm with a metal tipped 20Fr and venous cannulation was performed using 30Fr and 34Fr for superior and inferior vena cava, respectively. The pump flow rate was selected according to body surface area and hematocrit was maintained between 20-25%. Adequate depth of anaesthesia and muscle relaxation was maintained throughout the CPB. Aorta was cross clamped and aneurysm sac opened after venting the heart via cannula placed in the left ventricle through left atrium. After ostial cardioplegia and cooling of patient to temperature 28-30°C, modified Bentall's procedure were performed with aortic valve replacement with a 25mm St. Jude Medical® Mechanical Heart Valve (SJM; St. Jude Medical Inc.; Minneapolis, Minn) and an aortic graft conduit with coronary button implantation. CPB time was 292 min and aortic cross clamp time was 246 min. during which ACT, ABG and hematocrit were maintained within normal limits.

Rewarming at a rate of \leq 0.5°C/min was initiated after distal aortic anastomosis. Vasodilator and vasopressors were used as required to achieve adequate haemodynamics. After meticulous de-airing and restoration of adequate cardiac function, as assessed by TEE, patient was slowly weaned off CPB with inotropic support titrated to a systolic blood pressure of 90-110 mmHg. Following venous decannulation, protamine was given after confirmation of no adverse reaction to the test dose. After ensuring hemostasis with a normal ACT and ABG, chest was closed with mediastinal drains in place and patient was shifted in postoperative intensive care unit for weaning and extubation.

Discussion

Ever since a combined replacement of the aortic valve and ascending aorta with a composite graft was first described by Hugh Bentall and Anthony De Bono in 1968, it has been used for various pathologic conditions involving aortic valve, aortic root and ascending aorta [5]. Later modified by Kouchoukos et al. this technique has become the gold standard for patients with AAE and various degrees of AR, even if the aortic valve may be structurally unaffected. Development of more conservative modalities such as replacement of aortic root with an adequately fashioned graft or valve-sparing procedures restoring adequate valvular function by resuspension of AV within a graft tube have altered the precept for management of AAE [6-8]. However, given the beneficial long-term outcomes of modified Bentall's operation, whether conservative surgeries are superior to composite graft replacement of the aortic root remains inconclusive [9-15].

Our case presented a rare challenge where the ascending aorta was massively dilated (approximately 9.15cm by CTA) which put him an imminent risk of rupture. Therefore, a thorough baseline

preoperative assessment was performed. Pulses in all extremities were palpated and patient was auscultated for decrescendo early-diastolic blowing murmur which indicated AR and hence valvular involvement. A thorough baseline neurologic assessment was conducted, as well as an evaluation for evidence of malperfusion. As for any major cardiovascular surgery, baseline serological investigations, including a complete blood count, hepatorenal metabolic panel, coagulation studies, and blood grouping/cross-matching, as well as a baseline ECG and chest radiograph was obtained [16]. However, a dearth of high-quality data regarding ideal anesthetic protocol for proximal aortic surgery has led to high inter-institutional variability regarding monitoring, anesthetic techniques, CPB management and other aspects.

Our patient had a smooth induction of anesthesia with no hemodynamic instability (within 20% of baseline value). Patients with AR tend to have left ventricular dysfunction and need a low afterload while avoiding bradycardia [17]. These patients have been noted to have depleted intramyocardial catecholamines, thus requiring higher inotropic support during weaning from CPB [18].

In this case, the patient did not require deep hypothermic circulatory arrest (DHCA). For a median sternotomy approach, a single lumen endotracheal tube was preferred. Giant aortic root aneurysm may have mass effect resulting in compression of the trachea and right main stem bronchus, which can result in stridor, wheezing, cough or tracheal deviation on X-ray (19,20). Our patient did not have such compression related symptoms. Intraoperative arterial blood gas (ABG) analysis, activated clotting time (ACT), temperature and urine output were continuously monitored.

In modified Bentall procedure, haemostasis is difficult to achieve and postoperative bleeding is a major problem. Myocardial ischemia occurs in approximately 6% of patients who underwent root replacement [21]. The incidence of clinically overt stroke is approximately 3.5–5%, but may be as high as 13% for type A dissection [22]. No such complications were encountered in our case.

Conclusion

Annuloaortic ectasia with an aortic root diameter of >9.0 cm with no associated etiological syndromes or aortic dissection is an extremely rare and challenging case for anesthetic management during aortic repair. The successful outcome of this case was manifested through a well-organized perioperative multidisciplinary approach. This case underscores the significance of regular follow-up and monitoring of aortic diameter in suspected AAE patients.

Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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