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CASE REPORT
Coronary heart disease

Extrinsic tricuspid valve compression due to an aortic aneurysm causing significant right to left shunt via a patent foramen ovale: a case report

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Background Aortic aneurysms are known to cause compression of adjacent structures including the tracheobronchial tree, oesophagus, and recurrent laryngeal nerve. Extremely rarely, they can lead to compression of the tricuspid valve (TV) annulus. We describe a case where aortic aneurysm caused TV annulus compression and persistent right-to-left shunt through a patent foramen ovale (PFO).

Case summary A 75-year-old female was admitted with headache and dizziness. On examination, she had persistent arterial desaturation with oxygen levels reduced to 69% at rest whilst breathing ambient air. Complete blood count demonstrated polycythaemia (Hb 174 g/L). Right to left cardiac shunt was suspected after significant lung and haematologic pathology was excluded. Transoesophageal echocardiography demonstrated a trileaflet aortic valve with an ascending aorta aneurysm and a stretched PFO with persistent right to left shunt across it. The ascending aortic aneurysm was observed coursing superior to and compressing the TV annulus. Invasive haemodynamic data demonstrated prominent 'a' waves in the right atrium, low RV (12/1 mmHg), and pulmonary artery pressures (14/6 mmHg), reduced cardiac output and significant right to left shunt with Qp:Qs 0.6. Computed tomography (CT) angiogram demonstrated a 5 cm fusiform ascending aorta aneurysm that coursed anteriorly causing TV annulus compression.

Discussion Tricuspid valve inflow obstruction associated with a right to left shunt across PFO can be an extremely rare complication of aortic aneurysm. This may result in persistent arterial hypoxaemia and secondary polycythaemia.

Keywords Case report • Right to left shunt • Patent foramen ovale • Aortic aneurysm • Tricuspid valve compression

Learning points

- Right to left cardiac shunting should be considered as a differential diagnosis in cases with persistent arterial desaturation and secondary polycythaemia.
- Aortic aneurysms can cause extrinsic compression of tricuspid valve annulus causing an increase in right atrial pressure and right to left shunting via patent foramen ovale.
- Multimodality imaging and invasive haemodynamics provide complementary datasets that help define cardiovascular pathophysiology.

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Introduction:

Thoracic aortic aneurysms have a variety of genetic and non-genetic aetiologies. The underlying pathology is often cystic medial degeneration that is accelerated by systemic hypertension.[1] Although most patients with thoracic aneurysms are asymptomatic, some become symptomatic due to aortic regurgitation and associated heart failure.[2] Large thoracic aneurysms may produce symptoms due to local compression effects such as cough,[3] breathlessness,[4] dysphagia,[5] hoarseness,[6] and back pain.[7] There is a rare but recognized complication of platypnoea–orthodeoxia syndrome (POS) secondary to distortion of the atrial septal anatomy and episodic right to left shunting via a patent foramen ovale (PFO) by the aortic aneurysm.[8,9] This is characterized by dyspnoea and hypoxia in the upright position relieved by assuming a supine posture.

Here, we describe an unusual presentation of an ascending aortic aneurysm associated with obstruction at the level of tricuspid valve (TV) annulus, causing elevation in right atrial pressures and a persistent left to right shunt through a PFO. Furthermore, persistent systemic desaturation had resulted in secondary polycythaemia, indicating significant volume and duration of the right to left shunting.

Timeline

Index date	Admission with headache and dizziness; found to be severely hypoxic
1 day	Computed tomography (CT) pulmonary angiogram excludes pulmonary abnormality
7 days	Cardiology assessment requested; transoesophageal echocardiography confirms shunt and aortic aneurysm
10 days	Right and left cardiac catheter confirms obstruction at the level of tricuspid valve (TV) and right to left shunt
12 days	CT aorta confirms ascending aorta compresses TV annulus; remaining thoracic and abdominal aorta is tortuous but normal
14 days	Patient discharged from hospital after she declined surgery
170 days	Patient managing limited activities at home, remains hypoxaemic.

Case presentation

A 73-year-old female with a long-standing history of systemic hypertension presented to the Emergency Department with a recent onset of progressive headache, dizziness, and breathlessness. She denied experiencing cough, wheeze, chest pain or ankle swelling. She was a lifelong non-smoker and did not have any history of chronic respiratory illnesses, recurrent urinary tract infections, or long-term use of fluoroquinolones.

She was alert, oriented, and only mildly distressed despite severely reduced peripheral oxygen saturation (69%) on physical examination. She was tachycardic (pulse rate 102/min) and hypotensive (92/60 mmHg). Peripheral cyanosis was observed in the absence of clubbing. No change in peripheral oxygen saturation was observed with postural changes. Although saturations improved to 78% when oxygen was administered via high flow nasal cannulae, normoxia was not achieved. Neurological examination did not reveal any abnormality. Given her presentation, initially, respiratory consultation was requested, and she underwent CT angiogram of the pulmonary arteries as well as high-resolution CT which excluded pulmonary thromboembolism and lung parenchymal abnormalities. Ascending aortic dilatation was observed on the initial CT scan, but the contrast was timed to opacify pulmonary arteries. Blood profile demonstrated polycythaemia (Hb 174 g/L) with otherwise normal complete blood count, inflammatory markers and biochemical panel.

Right to left cardiac shunting was suspected on the basis of significant systemic desaturation that was not corrected after administration of high flow oxygen. Transthoracic echocardiography was initially attempted but was unfortunately limited by poor acoustic windows. We proceeded to transoesophageal echocardiography (TOE) with minimal sedation. Transoesophageal echocardiography demonstrated a dilated, unfolded ascending aorta coursing anteriorly and superiorly to the TV annulus (*Figure 1A*, Supplementary material online, *Video*). The aortic valve was trileaflet with mild regurgitation. A stretched PFO with persistent right to left shunt was visualized (*Figure 1B*, Supplementary material online, *Video*). The dilated proximal ascending aorta was seen to course superior to the TV causing significant narrowing (<1 cm) of the TV annulus. Colour flow Doppler demonstrated flow acceleration across the TV (*Figure 1C*, Supplementary material online, *Video*) with a mean gradient of 5 mmHg in the presence of a PFO. No intrinsic TV leaflet abnormality was visualized. Continuous-wave Doppler interrogation of the TV demonstrated prominent 'a' waves.

Following TOE, coronary angiography as well as right and left cardiac catheter was undertaken to exclude significant coronary artery disease and assess haemodynamics. Coronary angiography demonstrated unobstructed epicardial vessels. The data from the haemodynamic study are summarized in Table 1. The right atrial (RA) pressure trace demonstrates a prominent 'a' wave (*Figure 2*) consistent with the TV Doppler on TOE. The mean diastolic pressure gradient between the right atrium and right ventricle was 8 mmHg despite the presence of a decompressing right to left shunt across the PFO which indicated a haemodynamically significant obstruction across the TV. Right ventricular (10/1 mmHg) and pulmonary artery (14/6 mmHg) pressures were significantly reduced. Cardiac index was also reduced (1.8 L/min/m^2) with a Qp: Qs of 0.6 indicating a significant right to left shunt. In summary, the haemodynamic data confirmed significant obstruction at the level of the TV with reduced cardiac index and a right to left shunt across the PFO.

To determine the morphology and extent of the thoracic aortic aneurysm, CT aortogram was obtained. This demonstrated fusiform dilatation of the ascending aorta measuring 5.3 cm in maximum diameter. The aortic root itself was 3.4 cm in maximum dimension. The aneurysmal ascending aorta coursed antero-superiorly to the TV annulus compressing it (*Figure 3*, Supplementary material online, *Video*).

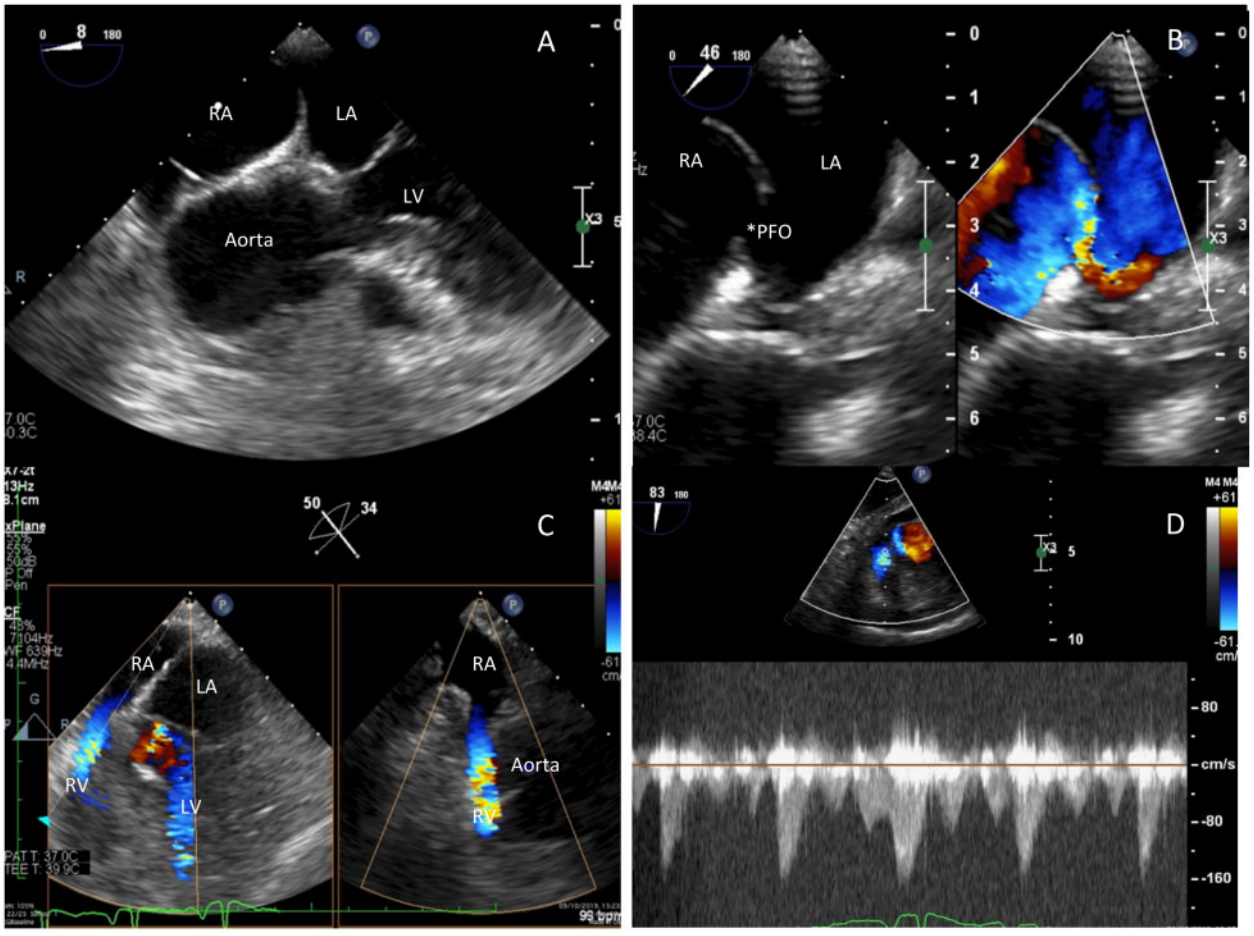

Figure 1 (*A*) The mid-oesophageal long-axis view with the ascending aorta crossing over the tricuspid valve annulus; (*B*) A colour comparison image of stretched patent foramen ovale with right to left shunt; (*C*) Flow acceleration across the tricuspid valve annulus due to extrinsic compression by the dilated aorta; (*D*) A continuous wave Doppler of the tricuspid valve and shows prominent a waves. *LA, left atrium; *LV, left ventricle; *RA, right atrium; *RV, right ventricle.

Table 1 Haemodynamic data from right and left heart catheter

Site	Pressure (mmHg)	Oxygen saturation (%)
Superior vena cava		38
Inferior vena cava		41
Right atrium	*a* = 13, *v* = 8, mean = 9	43
Right ventricle	10, EDP = 1	44
Main pulmonary artery	16/6; mean = 9	44
Left atrium	*a* = 5, *v* = 4, mean =4	62
Left upper pulmonary vein	—	97
Left ventricle	80, EDP = 4	—
Femoral artery	88/62	69

Note elevated right atrial '*a*' wave and low right ventricular and pulmonary artery pressures.

The aortic arch and descending aorta were normal in calibre with mild calcification. There was no haematoma, thrombosis, or dissection. Furthermore, tortuosity of the descending aorta and kyphoscoliosis were also observed.

Based on the echocardiographic, cardiac catheter and CT data, it was clear that the dilatation and configuration of ascending aortic aneurysm caused extrinsic compression of the TV annulus and persistent, significant right to left shunting via the PFO. The patient was advised to have the ascending aorta replaced and the PFO closed but after careful discussion of the risks and benefits, she declined the procedure. The patient continues to be followed up in the clinic and despite restricted mobility and hypoxaemia, continues to manage her daily activities inside the house.

Discussion

Ascending aortic aneurysms are often asymptomatic but can cause acute symptoms in cases of catastrophic complications such as

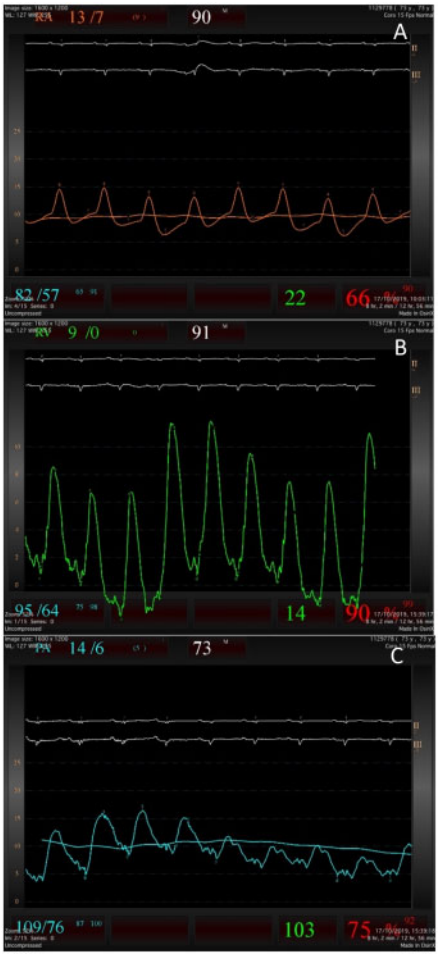

Figure 2 Selected pressure traces from haemodynamic study. (*A*) Right atrial pressure trace with prominent a wave (slightly precedes each QRS complex). (*B,C*) Reduced right ventricular and pulmonary artery pressures respectively. Prominent respiratory swing is also noted in the ventricular and arterial traces.

dissection or rupture or chronic symptoms related to pressure effects on adjacent structures. They may also be associated with POS. In the case of our patient, arterial desaturation and secondary polycythaemia with normal pulmonary investigations alerted us to the possibility of a right to left cardiac shunt. Although TOE confirmed our diagnostic impression of a right to left shunt across the PFO, it also demonstrated a dilated ascending aorta encroaching on the TV annulus and associated flow acceleration across the TV. However, confirmation of this physiology and quantification of associated significant right to left shunt required an invasive cardiac catheter study. Furthermore, CT aortogram displayed the extent and three-dimensional morphology of the aortic aneurysm. Our impression is that right to left shunt through the PFO allowed maintenance of her cardiac output at the expense of systemic desaturation.

To our knowledge, significant right to left shunting across a PFO due to TV compression by an aortic aneurysm has not been reported before. Compression of the TV by the aortic root[10] and the association of POS with an aortic aneurysm[11] have been previously reported separately. Our case demonstrates that extrinsic compression of the TV by the aortic root can be associated with significant right to left shunt through a PFO due to a combination of all these lesions. This requires careful multimodality assessment for an accurate diagnosis.

Lead author biography

Dr Rizwan Ahmed is a consultant cardiologist at the Institute of Cardiac Sciences, Sheikh Khalifa Medical City, Abu Dhabi, United Arab Emirates. He completed his training in Adult Congenital Heart Disease and Cardiovascular MRI in London, UK. He undertook post-doctoral research in Cardiovascular genomics at Imperial College, London. His research interests

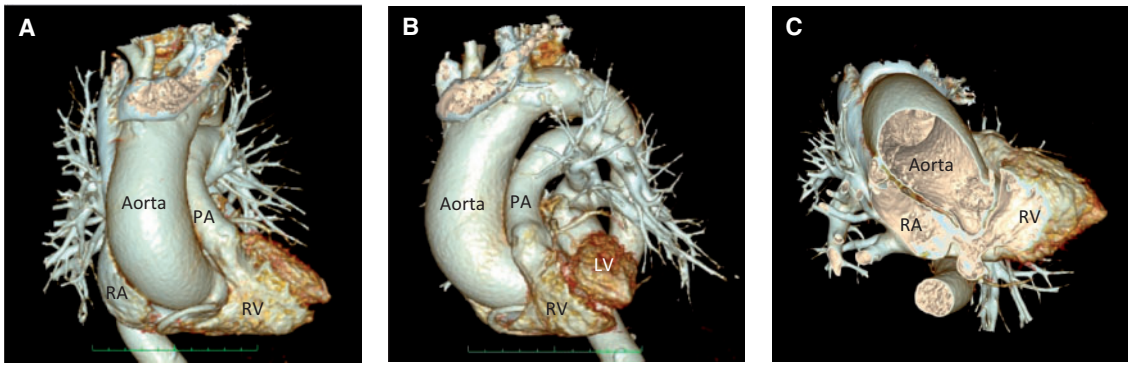

Figure 3 Computed tomography aortogram demonstrating the ascending aortic aneurysm in AP view (*A*) and left anterior oblique (LAO) view (*B*). (*C*) An endoluminal cross sectional view at the level of the tricuspid valve to demonstrate the extrinsic compression caused by the aortic root.

include cardiovascular developmental biology and cardiovascular haemodynamics.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author confirms that written consent for submission and publication of this case report including images and associated text has been obtained from the patient or next of kin in line with COPE guidance.

Conflict of interest: none declared.

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