Indian Journal of
Applied Radiology

Open Science Publications

Volume 11, Issue 1 - 2025

www.opensciencepublications.com

Spontaneous Aorto-Pulmonary Artery Fistula Secondary to Ruptured Aortic Arch Aneurysm Presenting as Acute High Output Cardiac Failure

Case Report

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Article Information: **Submission: 22/09/2024; Accepted: 10/02/2025; Published: 15/02/2025**

Abstract

Aorto-pulmonary artery fistula is an uncommon consequence of chronic thoracic aortic aneurysm and is mostly caused by pressure erosion. This can be aggravated by infection, trauma, surgery and is rarely spontaneous. Cross-sectional imaging in patients with unexplained acute onset high output cardiac failure is a warranted investigation to diagnose fistulous communication between great vessels i.e. aorta and pulmonary arteries, outline the anatomy, diagnose secondary complications and in effectively guiding the management.

Introduction

Aorto-pulmonary fistulas are rare complications of usually thoracic aortic aneurysms and can present with sudden onset high output failure. The modality of treatment is usually surgical with endovascular treatments coming up in the recent past. It is an emergency and has high mortality rates if left untreated.

Case Presentation

A 55-year-old male, a known hypertensive on medication, was brought to the emergency department with sudden onset of breathlessness, sweating and dry cough. The breathlessness had worsened to grade IV NYHA over a period of 4 days. There were no complaints of chest pain, palpitations and swelling of lower limbs. Examination revealed pulse rate of 90 beats/minute and blood pressure of 110/70mmHg. Hoarseness of voice was noted with engorged neck veins. Fine crepitations were heard in the right infra-scapular region. Continuous murmur was noted on auscultation in the aortic area. Left vocal cord palsy was noted on indirect laryngoscopy which was performed to evaluate hoarseness of voice.

ECG showed T-wave inversion in the lead I and avL, reflecting left atrial enlargement.

Chest X-ray **(Figure 1)** performed revealed mediastinal widening, cardiomegaly with right chamber enlargement with an upturned cardiac apex. The right hilar and perihilar vascular markings were prominent with ipsilateral peribronchial cuffing. There was blunting of right costophrenic angle suggestive of pleural effusion. These features with clinical correlation represented congestive cardiac failure with unilateral cardiogenic pulmonary oedema.

Laboratory investigations revealed Hb of 13.6mg%, total

ISSN: 2581-3919

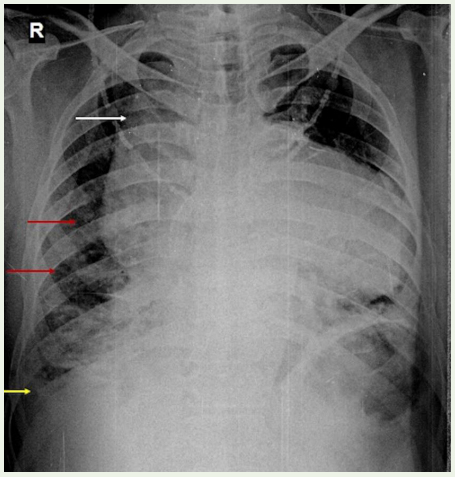

Figure 1: Chest X-ray PA view shows mediastinal widening (white arrow). Cardiomegaly with right chamber enlargement with an upturned cardiac apex seen. The hilar and perihilar vascular markings are prominent on the right side suggestive of right sided pulmonary oedema (red arrows). The right costophrenic angle is blunted representing pleural effusion (yellow arrows).

leucocyte count 5700mg/ml, platelet count of 1.3 lakhs, urea 34mg/dl, creatinine 1.0 mg/dl, troponin I 0.085ng/ml (normal upto 0.04ng/ml), serum CK-MB 1.3ng/ml (normal upto 5ng/ml) and positive D-dimer test.

2D echocardiography revealed dilated right atrium and right ventricle with severe pulmonary arterial hypertension (PASP 80mmHg) and moderate tricuspid regurgitation. IVC was dilated and non collapsing. LVEF 60%. No regional wall motion abnormalities were seen. No pericardial effusion was present. However, orthopnoea limited detailed evaluation.

In view of the elevated D-dimer and the chest X-ray and echocardiographic findings, CT pulmonary angiography (Figure 2 (a-d) was performed to rule out pulmonary thromboembolism.

CT showed ectatic ascending aorta, measuring 4.7cm in diameter with intimal calcification involving ascending aorta and aortic arch. Large saccular aneurysm was seen arising from the inferior wall of the distal aortic arch (9.8 x 9.6 x 9.2 cm) with a wide neck of 6.8 cm and a peripheral intraluminal thrombus. Communication was seen between the aortic aneurysm and main pulmonary artery over a length of 8 mm representing aorto-pulmonary artery fistula. The aortic aneurysm was compressing the left pulmonary artery with dilatation of the main and right pulmonary artery. No evidence of pulmonary thrombo-embolism was seen. Cardiomegaly was noted with dilated right atrium, right ventricle and dilated IVC. Moderate right sided pleural effusion was seen. On the lung reformatted images, patchy alveolar air space disease was present in the right lung, suggestive of right-sided pulmonary edema. Left lung parenchyma was normal. Another partly-thrombosed saccular aneurysm of the right innominate artery (9 x 6.8 x 6 cm) was seen compressing the superior vena cava with multiple venous collaterals along the right anterolateral chest wall.

Digital subtraction angiography (Figure 3 (a-b) performed subsequently, confirmed the aortic aneurysm and aorto-pulmonary

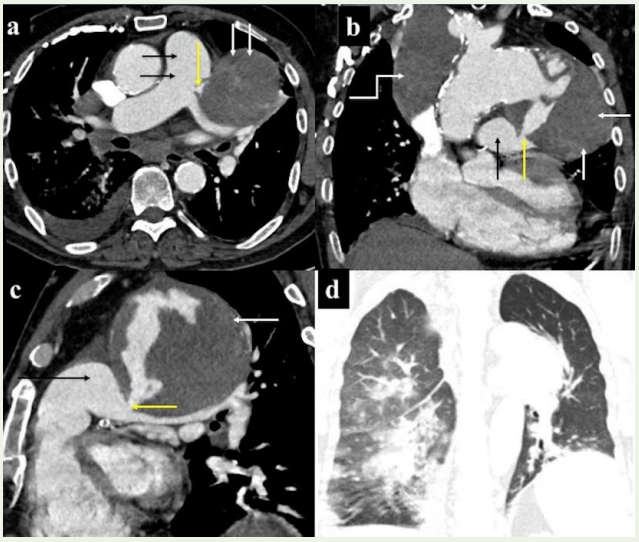

Figure 2: (a-d): Contrast enhanced computed tomography (CECT) pulmonary angiogram images (a-c) Axial, oblique coronal and oblique sagittal images showing showing partially thrombosed aortic arch aneurysm (white arrows) communicating with the main pulmonary artery (black arrows) resulting in the aorto-pulmonary artery fistula (yellow arrows). Note is also made of fusiform aneurysm of the brachiocephalic trunk (curved arrow) and right sided mild pleural effusion. (d) Coronal lung window CT image showing patchy alveolar air space disease in the right lung, suggestive of unilateral right-sided pulmonary edema.

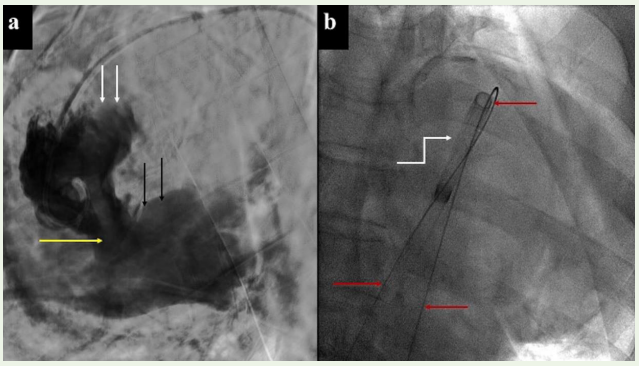

Figure 3: (a). Digital subtraction angiography (DSA) left anterior oblique (LAO) lateral projection image showing large fistulous communication (yellow arrows) (~22 mm) between the partially thrombosed aortic arch aneurysm (white arrows) and main pulmonary artery (MPA) (black arrows). The pigtail catheter is in the ascending aorta. (b). DSA antero-posterior projection image showing placement sheath of closure device (curved white arrow) in the aorto-venous loop (red arrows) across the aorto-pulmonary arterial fistula.

artery fistula and delineated the precise location and size of the fistulous communication (22mm) using multiple projections. Ostial stenosis was noted of LCA. Surgical option was given but declined by the relative in view of high risk. Urgent interventional percutaneous placement of closure device was thus planned. Pigtail catheter was placed in the ascending aorta and contrast injection demonstrated the fistula. The fistulous communication was engaged from the aortic aspect. The terumo guide wire was introduced and advanced to form aorto-venous loop by snaring from the venous access

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through main pulmonary artery. The placement sheath of Flex II ASD occlusive device (33mm waist) was placed over the guide wire. However unfortunately during the intervention sudden clinical and hemodynamic deterioration resulted in cardiac arrest and the patient succumbed, despite all attempts to revive him.

Discussion

Aneurysms of the thoracic aorta may present with chest pain, thrombo-embolism resulting in stroke, compressive symptoms like recurrent laryngeal nerve compression resulting in hoarseness of voice, dysphagia due to esophageal compression etc. or may be asymptomatic [1]. They can be complicated by aortic dissection, rupture, congestive heart failure, and fistulous communication with adjoining structures including aorto-pulmonary, aorto-bronchial and aorto-esophageal fistula.

Aorto-pulmonary arterial fistula occurs when the wall of a degenerative or false aortic aneurysm rupture into the pulmonary arterial circulation due to pressure erosion caused by pulsatile friction [2,3]. Most of the fistulae originate from the ascending aorta; fistulous communication between the distal aortic arch or the descending aorta to the pulmonary artery is quite rare [4]. It can be seen in post-traumatic or post-operative setting and in infective conditions like pneumonia, lung abscess. The fistula causes volume overload in the pulmonary arterial circulation and is fatal if not treated timely [5]. The presentation is acute with chest pain, hemoptysis, orthopnea and paroxysmal nocturnal dyspnea.

In our case, there was neither previous history of surgery nor hemoptysis. Since there was no significant past history, the duration of the aortic aneurysm being asymptomatic remains unclear.

The left to right shunt caused by the fistulous communication between the major vessels caused a high output failure, resulting in pulmonary edema, pleural effusion and moderate pulmonary hypertension.

Chest X-ray shows mediastinal widening, which is contributed by the aortic aneurysm and/ or SVC dilatation. Pulmonary artery segment appears prominent and dilated in pulmonary arterial hypertension. Perihilar patchy opacities in characteristic "bat-wing" pattern with Kerley A and B lines may be present along with cephalisation, suggestive of pulmonary edema. "Meniscus" sign may be present with blunting of costophrenic angle signifying pleural effusion. Cardiomegaly with outward shift of the apex of the heart, if present, represents right ventricular dilatation in right heart failure.

2D echocardiography helps in visualising the cardiac anatomy i.e. cardiac chambers for size, valves, pericardial cavity, inferior and superior vena cava as well as physiology i.e. wall motion abnormality, valvular dysfunction and ejection fraction. Doppler-mode shows continuous flow in the pulmonary artery and increased pulmonary arterial pressure in aorto-pulmonary arterial fistula. The fistulous communication may also be visualised.

CT angiography is non-invasive modality for evaluation of unexplained high output cardiac failure owing to high spatial resolution, for the presence of aneurysm, fistulous communication of the aneurysm with adjoining bronchus, oesophagus, lung or pulmonary artery. Partial or complete thrombosis of the aneurysm, aortic dissection, leak and aortic rupture can also be well demonstrated. In addition, the mass effect on adjacent structures, status of the lung parenchyma and pleural cavity is confirmed on reformatted images.

In our case, there were evident features of pulmonary edema in the right lung parenchyma Left lung parenchyma appeared normal. The possible explanation of unilateral right sided pulmonary edema was secondary to compression of the left pulmonary artery. Thus, the consequence of left to right shunt and elevated pulmonary arterial pressures manifested as changes involving the right pulmonary artery and right lung [6].

Cardiac catheterization confirms the fistulous communication between the aorta and the pulmonary artery, outlining the precise location and dimension of the fistula. This modality is invasive but has diagnostic and therapeutic role in managing aorto-pulmonary arterial fistulas.

Conclusion

Aorto-pulmonary artery fistula is ideally treated surgically and percutaneous coil embolization has shown favourable outcomes [7]. Surgeries such as arch replacement and pulmonary arterial repair may be recommended too. Less invasive therapy including endovascular stent grafting, combined interventional radiological and surgical approaches are safer in patients at high risk for surgery [8-10] The occurrence of aorto-pulmonary fistulas remains rare, and those caused due to ruptured arch of aorta aneurysms are exceedingly rare at 4% incidence of all cases reported [11].

References

1. Halperin JL, Olin JW, Elefteriades JA, Ziganshin BA (2004) Disease of the aorta. In: Fuster V editor. Hurst's The Heart. 11th ed. New York: McGraw-Hil Pp: 2304.
2. Killen DA, Muehlebach GF, Wathanacharoen S (2000) Aortopulmonary fistula. South Med J 93: 195-198.
3. Kim TH, Moon CI, Choi JW, Choi MJ (2000) Congenital aortopulmonary fistula presenting as an exertional dyspnea. Korean Circ J 30: 1291-1294.
4. Hirose H, Svensson LG (2004) Chronic posttraumatic aneurysm of descending aorta with fistulous communication into pulmonary artery. J Vasc Surg 40: 564-566.
5. Vaideeswar P, Deshpande JR, Sivaraman A (2000) Aorto-pulmonary fistulization – an unusual complication of syphilitic aortic aneurysm. Int J Cardiol 75: 299-300.
6. Mukadam M, Barraclough J, Riley P, Bonser R (2005) Acquired aortopulmonary artery fistula following proximal aortic surgery. Interact Cardiovasc Thorac Surg 4: 388-390.
7. Koen JG, Wagenaar R, Janson JT (2020) A case report of an aorto-pulmonary-venacaval fistula after penetrating cardiac injury Eur Heart J Case 15: 1-6.
8. Kochi K, Okada K, Watari M, et al. (2002) Hybrid endovascular stent grafting for aortic arch aneurysm with aortopulmonary fistula. J Thorac Cardiovasc Surg 123: 363-364.
9. Gulati A, Kapoor H, Donuru A, Gala K, Parekh M (2000) Aortic Fistulas: Pathophysiologic Features, Imaging Findings, and Diagnostic Pitfalls 41: 1335-1351.
10. Koen JG, Wagenaar R, Janson JT (2020) A case report of an aorto-pulmonary-venacaval fistula after penetrating cardiac injury Eur Heart J Case 15: 1-6.
11. Dixit MD, Gan M, Narendra NG, Mohapatra RL, Halkatti PC, et al. (2009) Aortopulmonary fistula: a rare complication of an aortic aneurysm. Tex Heart Inst J. 36: 483-485.

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