CASE REPORT

A rare case of acquired aortopulmonary fistula with bicuspid aortic valve: report of successful surgical repair

Rajendra Kumar Premchand,[1] Bolleneni Bhaskar Rao,[2] Kaustubh Partani[1]

[1]Department of Cardiology, Krishna Institute of Medical Sciences (KIMS), Secunderabad, India
[2]Department of Cardiothoracic and Cardiovascular Surgery, Krishna Institute of Medical Sciences (KIMS), Secunderabad, India

Correspondence to
Dr Rajendra Kumar Premchand, kumarpre@hotmail.com

Accepted 28 October 2014

SUMMARY

An acquired aortopulmonary fistula is a rare and usually fatal phenomenon. Rarer still are reports of successful surgical repair of aortopulmonary fistulae. We present the case of a 48-year-old hypertensive man who presented with congestive cardiac failure. Examination revealed a bicuspid aortic valve and a large aneurysm of the arch of the aorta, which was communicating with the main pulmonary artery. The diagnosis of acquired aortopulmonary fistula was made using transthoracic echocardiography findings and confirmed by CT. The patient was successfully managed by surgery, with an uneventful postoperative recovery, with control of congestive cardiac failure. At 1-year follow-up, the patient had Class I symptoms.

BACKGROUND

Aortopulmonary fistulae, reported since 1812, are an unusual complication of aortic aneurysms rupturing into the pulmonary artery.[1 2] Very few patients survive rupture of an aortic aneurysm as it may lead to cardiac tamponade or haemorrhagic shock and ultimately death.[2] In addition, the risk of severe pulmonary hypertension and congestive heart failure remains high in patients with aortopulmonary fistulae.[2] The diagnosis of a fistula between the aortic arch aneurysm and the pulmonary artery is usually obtained during postmortem and rarely in live patients. As a result, very few cases of successful surgical management are reported.[3] According to a study, only 108 live cases of acquired aortopulmonary fistulae have been reported worldwide up to 1992, of which surgical correction was attempted in only 13 cases while successful outcomes were reported in only four cases.[4] In spite of being a condition associated with high morbidity and mortality,[2] early and accurate diagnosis of aortopulmonary fistulae can lead to successful surgical outcomes, as demonstrated in this case.

CASE PRESENTATION

We report a case of a 48-year-old hypertensive man who presented to our hospital with sudden onset of congestive cardiac failure and shortness of breath. Physical examination revealed elevated jugular venous pressure and normal blood pressure. Cardiovascular examination revealed a hyperdynamic left ventricular apex at the fifth intercostal space in the mid-clavicular line. The pulmonary component of the second heart sound was loud. Further, a continuous murmur was heard all over the precordium on auscultation. Basal crepitations were audible near the lung fields. The patient's laboratory investigations were within normal ranges.

INVESTIGATIONS

The patient's ECG was normal. Chest radiograph revealed mediastinal widening with cardiomegaly (figure 1). Further, an aneurysm of the aorta was causing tracheal deviation to the right. Pulmonary oedema and pulmonary venous congestion were present (figure 1). Transthoracic echocardiogram revealed a bicuspid aortic valve with an aneurysm of 7 cm at the arch of the aorta (figure 2). Two-dimensional echocardiogram showed a defect measuring 7 mm in the common wall formed by the aneurysm at the arch of the aorta and pulmonary artery. This resulted in a large left-to-right aortopulmonary shunt, that is, aortopulmonary fistula. A colour Doppler imaging study demonstrated a shunt across the defect entering the main pulmonary artery near its bifurcation, the direction of flow being perpendicular to the long axis of the main pulmonary artery (figure 2). Left ventricular systolic function was normal. Right ventricle and right atrium were mildly dilated with right ventricular dysfunction. There was moderate tricuspid and mild mitral regurgitation. Estimated pulmonary systolic pressure was 88 mm Hg.

CT verified a huge saccular aneurysm of the ascending aorta (maximum diameter 8 cm) starting above the level of the infundibulum and extending up to the right brachiocephalic artery (figure 3). Communication measuring 1 cm was observed between the aneurysm and the main pulmonary

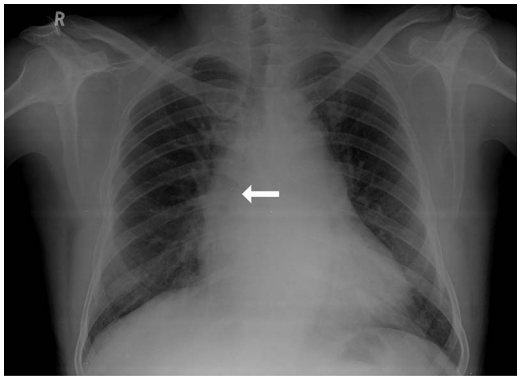

Figure 1 Chest X-ray revealing cardiomegaly with bilateral lung congestion/pulmonary oedema; aortic aneurysm of the arch of aorta is seen, causing a tracheal shift towards the right, represented by an arrow.

To cite: Premchand RK, Bhaskar Rao B, Partani K. *BMJ Case Rep* Published online: [*please include* Day Month Year] doi:10.1136/bcr-2014-207374

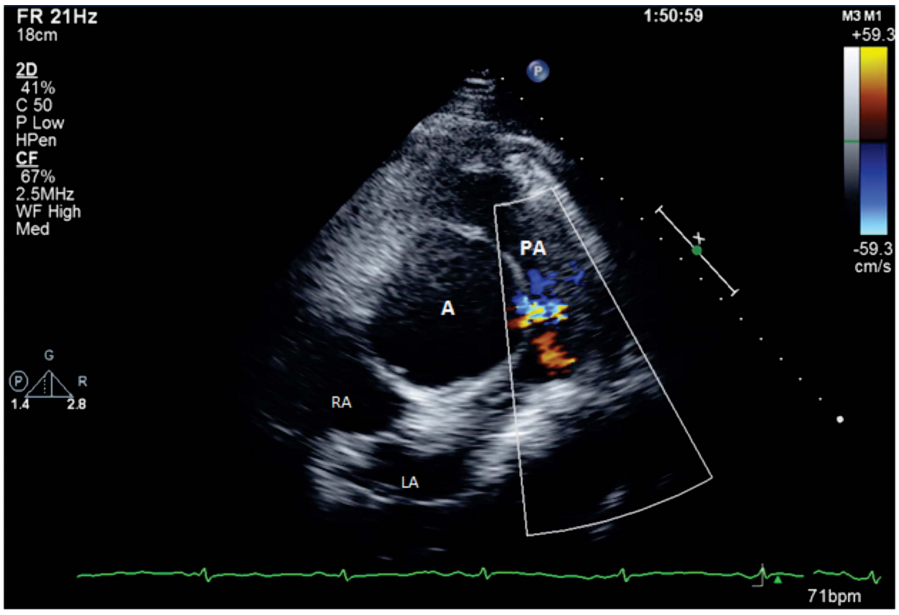

Figure 2 Transthoracic echocardiogram* showing a giant aneurysm at the arch of the aorta; colour Doppler study displaying the direction of blood flow from the aneurysm to the main pulmonary artery (*basal short axis view; A, aneurysm; LA, left atrium; PA, pulmonary artery; RA, right atrium).

artery (figure 3). Coronary arteries were normal. The patient was haemodynamically stable with class IV heart failure.

DIAGNOSIS AND TREATMENT

The patient was diagnosed with acquired aortopulmonary fistula associated with aortic aneurysm. Surgical management within 24 h was advised to repair the aneurysm and aortopulmonary fistula. During the procedure, a median sternotomy was performed by making an incision on the chest. The pericardium of the heart was opened and the patient was put on cardiopulmonary bypass. When the core temperature reached 25°C, total circulatory arrest was instituted by cross clamping the aortic arch. The aneurysm sac was then opened longitudinally. The aortic valve was found to be bicuspid without any evidence of incompetence or stenosis. A defect measuring 1 cm was seen between the aorta and the pulmonary artery (figure 4). Through the defect, it was possible to probe the pulmonary trunk.

The closure of the defect was performed with a Dacron patch using 5–0 polypropylene sutures. A Gore-Tex tube graft of size 22 (Gore Medical Devices, USA) was anastomosed to the aorta by continuous-suture technique at proximal and distal ends using 4–0 polypropylene sutures. The redundant aneurysm aortic wall was closed over the graft to minimise the dead space between the aorta and the main pulmonary artery. Subsequently, the cross clamp was removed and cardiopulmonary bypass was discontinued. The heart resumed spontaneously to normal sinus rhythm.

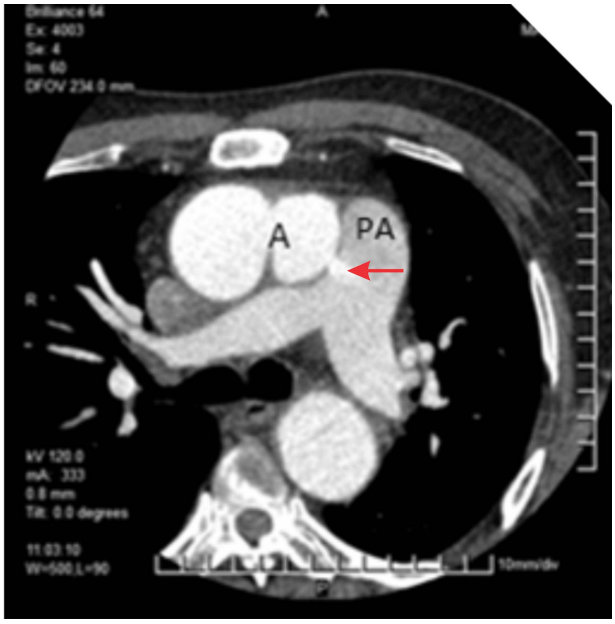

Figure 3 CT scan displaying the contrast axial view of the aneurysm with connection to the main pulmonary artery; the solid arrow points to the connection and depicts the flow from the aneurysm to the pulmonary artery (A, aneurysm; PA, pulmonary artery).

OUTCOME AND FOLLOW-UP

In our patient, closure of the fistula and replacement of the ascending aorta with a prosthetic tubular graft were successfully carried out. The patient reported an uneventful postoperative recovery and was able to return to work. At 1-year follow-up, the patient had Class I symptoms.

DISCUSSION

Among the normal population, bicuspid aortic valve is prevalent in 1–2% of individuals.[5] Approximately 33% of these patients

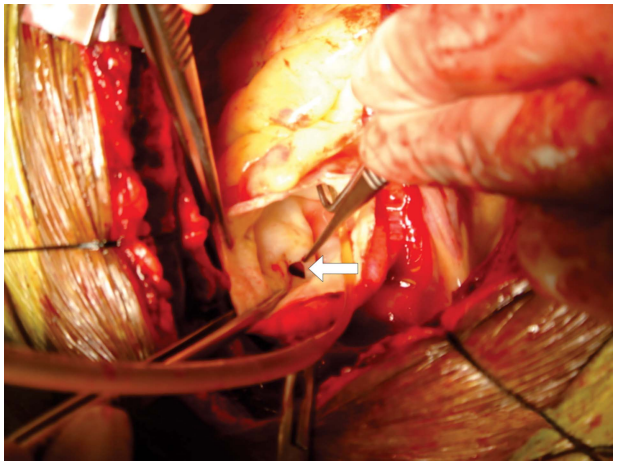

Figure 4 Intraoperative image displaying a 1 cm fistula between the aorta and the pulmonary artery, represented by an arrow.

develop serious complications. An aortic aneurysm is one such complication.[5] In rare instances, aortic aneurysm may lead to further complications of fistula formation between the aorta and the pulmonary artery.[1 6] In 1924, Boyd reported that the occurrence of aortopulmonary fistula was nearly 4% among the 4000 autopsy cases with thoracic aortic aneurysm investigated in the study.[7] The other rare causes of acquired aortopulmonary fistula include Marfan's syndrome, trauma, aortic dissection, infective endocarditis, inflammatory aortitis and syphilitic aortitis.[8] The most common cause, however, is atherosclerotic aneurysm, as described in our patient.

Although the majority of patients with aortopulmonary fistula remain asymptomatic, when they do occur, symptoms may include chest pain, haemoptysis and dyspnoea.[1 6] Further, an aortic aneurysm of more than 6 cm diameter is common in patients with aortopulmonary fistulae and is frequently associated with the development of acute left-to-right shunt, new murmur, systemic arterial hypertension, signs of cardiac insufficiency and severe pulmonary hypertension.[1 2] In the presenting case, we found an aortic aneurysm communicating with the pulmonary artery leading to an acute left-to-right shunt, with volume overload of the left ventricle causing acute congestive heart failure. The signs and symptoms of dyspnoea and pulmonary hypertension, and a continuous murmur heard over the precordium, indicated a suspicion of fistula between the great vessels, which required further investigations. Subsequently, multiple diagnostic tests were used for the definitive diagnosis.

Chest X-ray revealed mediastinal widening and cardiomegaly, while the two-dimensional and Doppler echocardiography detected abnormal aortopulmonary shunt flow. The diagnosis was confirmed using CT. We opine that early and accurate diagnosis is essential for appropriate planning and management of an aortopulmonary fistula.

Immediate surgical management of an acquired aortopulmonary fistula is always mandatory because of potentially lethal complications.[2] Surgical repair involves closure of the fistula followed by tubular graft replacement of the ascending aorta. Cerebral protection using profound hypothermia (≤25°C) and management of pulmonary complications by total circulatory arrest are vital to the surgical procedure.[3] Although very few cases of successful surgical repair are reported in the literature, recent advances in cardiopulmonary bypass, myocardial preservation, anaesthesia and surgical graft techniques have played a significant role in improving survival rates.[9] In 1960, Giacobine and Cooley[10] reported the first case of successful surgical repair for an aortopulmonary fistula. Since then, various reports of successful surgical repair of acquired aortopulmonary fistulae secondary to bacterial endocarditis,[11] Marfan's syndrome,[12] syphilis,[13] aortic valve replacement[14] and severe aortic stenosis[15] have been described. In India, a few cases of acquired aortopulmonary fistulae[8] with successful surgical repair[9] have been reported. However, to the best of our knowledge, this is the first report with a successful surgical outcome of a patient in India with an acquired aortopulmonary fistula and bicuspid aortic valve. Since the patient in our case was haemodynamically stable, prompt surgical intervention was possible. Accordingly, closure of the fistula and replacement of the ascending aorta with a prosthetic tubular graft were successfully carried out. Percutaneous intervention was ruled out in our patient since the ascending aorta was aneurysmal. The outcomes of surgical repair were excellent and the patient was able to return to work.

In conclusion, we report the first case, in India, of successful surgical repair of an ascending aorta aneurysm associated with an aortopulmonary fistula in a 48-year-old hypertensive man with a recent onset of congestive heart failure with a bicuspid aortic valve. We opine that early diagnosis and prompt surgical intervention can be lifesaving in patients with acquired aortopulmonary fistulae.

Learning points

- The diagnosis of fistula between the aortic arch aneurysm and the pulmonary artery is usually obtained during postmortem and rarely in live patients. As a result, very few cases of successful surgical management are reported.
- We report an uncommon but successfully managed case of a fistula between the arch of the aorta and the main pulmonary artery as a complication of aortic arch aneurysm, and bicuspid aortic valve.
- Early and accurate diagnosis using transthoracic echocardiography and CT as well as prompt surgical intervention is vital for successful outcomes in patients with acquired aortopulmonary fistulae.

Acknowledgements The authors wish to thank Mr Nirlep Gajiwala and Dr Ashok Thakkar for providing valuable assistance in literature review and preparation of the manuscript.

Contributors RKP and BBR contributed to conception and design, acquisition of data, revising the manuscript critically for important intellectual content and final approval of the version to be published. KP contributed to conception and design, acquisition of data, preparation of the first draft of the manuscript and final approval of the version to be published.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

1 Kim SE, Kim HJ, Lee SH, *et al*. A case of aortopulmonary fistula caused by a huge thoracic aortic aneurysm. *Korean Circ J* 2009;39:209–12.
2 Sobral MLP, Santos LAS, Santos GGd, *et al*. Aneurisma do arco aórtico com fístula aorto-pulmonar: tratamento cirúrgico com sucesso. *Revista Brasileira de Cirurgia Cardiovascular* 2004;19:409–12.
3 Lahey SJ. Successful surgical management of an aortic arch aneurysm with acute aorto-pulmonary fistula. *Ann Thorac Surg* 1993;55:995–7.
4 Kakio T, Konishi T, Maruyama S, *et al*. A case of successful repair of aorto-pulmonary artery fistula secondary to aortic arch aneurysm. *Kokyu To Junkan* 1992;40:603–7.
5 Fedak PW, Verma S, David TE, *et al*. Clinical and pathophysiological implications of a bicuspid aortic valve. *Circulation* 2002;106:900–4.
6 Belgi A, Altekin E, Yalcinkaya S, *et al*. Acquired aorto-pulmonary fistula: a case of ruptured aneurysm of the thoracic aorta. *Anadolu Kardiyol Derg* 2003;3:275–8.
7 Boyd LJ. A study of four thousand reported cases of aneurysm of the thoracic aorta. *Am J Med Sci* 1924;168:654–67.
8 Shenoy AR, Naik GD, Varghese K, *et al*. Acquired aortopulmonary fistula. *Indian Heart J* 2003;55:268–71.
9 Dixit MD, Gan M, Narendra NG, *et al*. Aortopulmonary fistula: a rare complication of an aortic aneurysm. *Tex Heart Inst J* 2009;36:483–5.
10 Giacobine JW, Cooley DA. Surgical treatment of aortico-pulmonary fistula secondary to aortic arch aneurysm: report of successful case. *J Thorac Cardiovasc Surg* 1960;39:130–6.
11 Buxton BF, Cooley DA. Recurrent aortopulmonary artery fistula. *Thorax* 1973;28:521–5.
12 Massetti M, Babatasi G, Rossi A, *et al*. Aortopulmonary fistula: an uncommon complication in dystrophic aortic aneurysm. *Ann Thorac Surg* 1995;59:1563–4.
13 Panday S, Hishikar A, Karbhase J. Rupture of syphilitic aneurysm of ascending aorta into main pulmonary artery: successful emergency repair. *J Thorac Cardiovasc Surg* 1982;83:470–1.
14 Large SR, English TA. Aorto-pulmonary fistula: a rare acquired lesion. *J Cardiovasc Surg (Torino)* 1988;29:403–5.
15 Ferlan G, De Pasquale C, Testini M, *et al*. Acquired aortopulmonary fistula. A case report. *J Cardiovasc Surg (Torino)* 1998;39:821–3.