Large patent ductus arteriosus in an adult complicated by pulmonary endarteritis and embolic lung abscess

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Abstract

Patent ductus arteriosus in the adult is an extremely rare clinical phenomenon. We report the case of a 34-year old man who developed pulmonary endarteritis and subsequent embolic lung abscess secondary to a large patent ductus arteriosus. This brief report also provides an overview of the natural history, potential complications, optimal therapy, and diagnostic dilemmas associated with this persistent congenital cardiac defect in adults.

Introduction

Patent ductus arteriosus (PDA) is failure of the duct between the aorta and the pulmonary artery to close, comprising approximately 5-10% of congenital heart defects.[1] First described by Galen in the 3rd century, Gross carried out the initial successful surgical repair in 1939.[2] Early childhood detection and safe operative correction mean that adulthood PDA is now rarely encountered in the Western world.[3] However, its association with a significantly reduced life expectancy has been highlighted by several studies in the past.[4-6]

Length, shape, elasticity and narrowest diameter determine PDA resistance, and together with the transductal pressure gradient, both factors predominantly control shunt magnitude, and hence the resulting detrimental physiological impact of the PDA.[7] The consensus view advocates the closure of large PDA for the prevention of the deleterious sequelae of chronic pulmonary overcirculation and endarteritis.[8] Infective endarteritis associated with untreated PDA was historically a fatal complication and the leading cause of mortality.[3] However, a dramatically declining incidence over the past 30 years allied with a greatly improved prognosis prompts the suggestion that routine *prophylactic* closure as a means of eradicating the risk of infection is unjustified.[9]

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Case Report

A 34-year old builder presented to his family doctor in an acute condition with shortness of breath. He described a 2-month history of malaise, lethargy, rigors and progressive exercise limitation. There was no past medical history of any risk factors for cardiac disease or endocarditis. Physical examination revealed him to be febrile at 38.3°C; he weighed 95 kg and was 193 cm tall. Blood pressure was 120/60, with 100% oxygen saturations and a regular pulse at 72 beats/min. Auscultation of his chest revealed a harsh, grade 3/6 continuous murmur which was loudest at the second left intercostal space; there were no signs suggestive of volume overload, ventricular failure, or raised pulmonary pressures. Chest X-ray demonstrated mild cardiomegaly with normal pulmonary vasculature, and electrocardiogram revealed normal sinus rhythm with no features of ventricular strain or atrial enlargement. Blood tests showed elevated inflammatory markers with a C-reactive protein of 295 and white blood cell count of 18.3 with normal renal and liver biomarkers.

Initial transthoracic echocardiography (TTE) preformed at the referring center showed the presence of a PDA (Figure 1); color flow doppler demonstrated high velocity retrograde flow from the descending aorta towards the main pulmonary artery bifurcation in the parasternal short axis view. Left ventricular dimensions were mildly elevated, with systolic and diastolic diameters of 42 mm and 61 mm, respectively. Biventricular function was good, pulmonary artery pressures were not raised, and no other valvular abnormality was noted. Following transfer to our center, the diagnosis was confirmed using 3-D computed tomographic (CT) reconstruction imaging (Figure 2A and B), and subsequent transoesophageal echocardiography (TOE) importantly revealed a large PDA of maximal diameter 16 mm and length 25 mm, appearing to be intimately related to a 25 mm × 15 mm vegetation in the main pulmonary artery (MPA) (Figure 3).

Empirical intravenous antibiotic therapy resulted in complete resolution of fever and normalization of inflammatory markers within four days. Blood cultures grew a fully sensitive Viridians *Streptococcus* organism, and the patient was discharged home on day 7 with the intention of continuing long-term intravenous therapy and regular clinical follow up.

Seven days post discharge the patient represented with acute onset shortness of breath and left sided pleuritic chest pain. CT imaging of the thorax showed a 20 mm × 12 mm cavitating lesion in the left lower lobe suggestive of a lung abscess (Figure 4). Repeat TOE generated support for a diagnosis of septic embolic lung abscess, showing an obvious reduction in size of the previously identified vegetation. A referral to our surgical team was then made for urgent surgical ligation of PDA plus removal of MPA vegetation.

Surgical access was established via a median sternotomy. Under controlled hypotensive anesthesia careful dissection revealed a 25 mm long, non-calcified PDA, with a maximal

diameter of 15 mm; aortic and pulmonary ends of the PDA measured 25 mm and 5 mm, respectively. Following isolation of the recurrent laryngeal nerve, cardiopulmonary bypass (CPB) was instituted via aorto-bicaval cannulation. Cardiac arrest was achieved via antegrade cold blood cardioplegia with pre-emptive systemic cooling to 24°, in anticipation of the potential need for circulatory arrest.

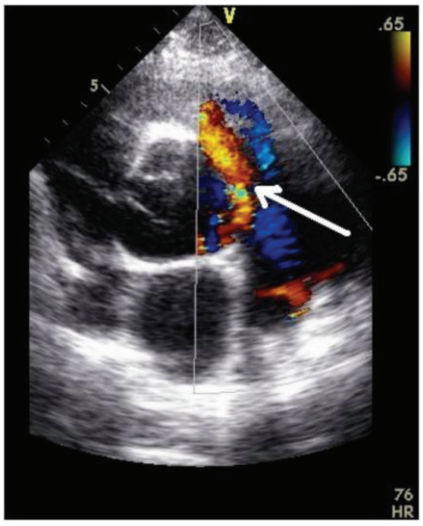

Figure 1. Transthoracic echocardiography image showing the patent ductus arteriosus (arrow).

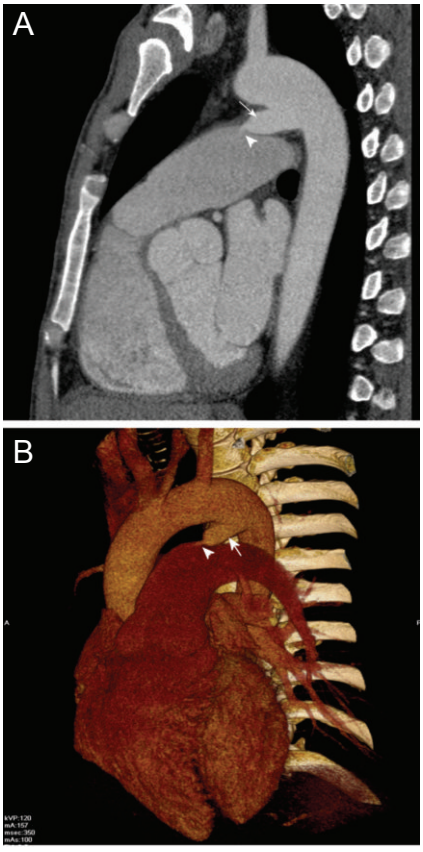

Figure 2. ECG-gated CT scan: sagittal oblique multiplanar reconstruction (A) and 3-D reconstruction (B) showing the PDA (arrow) with the narrow end towards the main pulmonary artery.

In contrast to ligation of *simple* non-infected PDA, the surgeon's initial priority was to safely evacuate vegetation from the MPA, thus preventing further systemic or pulmonary embolization. This was achieved via incision of the MPA and inspection of the lumen, which revealed no obvious vegetation but a hypertrophic tunica intima, consistent with endarteritis. The pulmonary orifice of the PDA was then easily oversewn from within the lumen, with maintenance of a clear bloodless field via placement of a vascular clamp towards the aortic end of the PDA. The presence of a large aortic PDA orifice determined our decision to perform the remaining division and repair under hypothermic circulatory arrest.

The patient made a completely uneventful recovery, being discharged to his referring hospital on day 6 with normal blood indices for continuing intravenous antibiotic therapy. Two years following surgery he presents no cardiac, respiratory or systemic symptoms. TTE shows no residual shunt with good ventricular function, reduction in ventricular dimensions, and chest X-ray shows complete resolution of the previously noted changes.

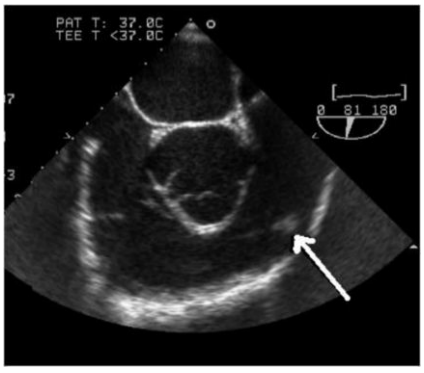

Figure 3. Transesophageal echocardiography image showing vegetation in main pulmonary artery (arrow).

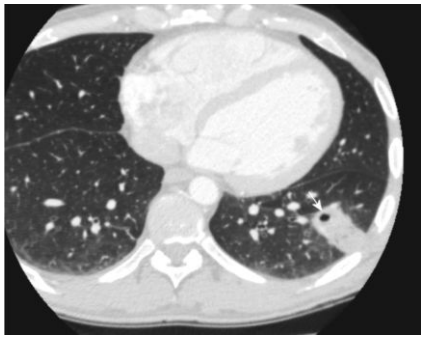

Figure 4. CT scan of the chest demonstrating the area of air space shadowing with a cavity (arrow) in the apical segment of left lower lobe.

Discussion

Endocarditis associated with PDA is rare.[10] There are less than 30 reported cases of pulmonary endarteritis associated with PDA in the adult, and overall incidence has dramatically decreased over the last 30 years.[7,9] Large shunts with diameters greater than 4.5 mm seem to be most susceptible, and infection of small clinically silent PDA is extremely rare and virtually non-existent.[11,12] Improving echocardiographic technology has increased detection of these small, hemodynamically insignificant *silent* PDA, and their optimal management, particularly in adults, remains controversial. Some groups recommend immediate closure as prophylaxis against future infection,[8] whereas others maintain that the incidence of endarteritis and secondary mortality to this is too low to justify intervention.[9] Detection of PDA via TTE is easy in children but can prove difficult and sometimes impossible in adults due to interference of the lung and the increased size of the thoracic cage.[13] Diagnosis rests upon showing the presence of an abnormal turbulent *mosaic* flow pattern within the pulmonary artery that must be proven to be originating from the descending aorta; this is because several other disorders, such as aortopulmonary septal defect, rupture of the sinus of Valsalva into the pulmonary artery, and pulmonary stenosis are also known to cause this flow pattern.[14] TOE has been shown to be significantly more sensitive in detection of adult PDA than TTE, with no obvious difference in specificity between the two modalities.[14]

In this case, TTE suggested the diagnosis which was then confirmed by both CT and TOE. However, there was inconsistency within and between modalities with regards to the presence of vegetation within the MPA, a diagnostic dilemma that has been previously reported.[15] Such anomalous findings are thought to arise from inter-observer variation in technique and the multiplanar nature of echocardiographic imaging. These findings emphasize the importance of a multimodal imaging approach in assessing adult PDA, as previously advocated in other reports,[14] particularly when infection is suspected.[13]

Open surgical division or ligation has traditionally been an extremely successful and safe method of PDA closure and there are large studies demonstrating excellent closure rates with low mortality (<2%).[16] Comparable results are now achievable using transcatheter devices and they are now the first-line closure

method in many centers; however, their use is contraindicated by the presence of infection. Historically the majority of surgical repairs have been via a postero-lateral thoracotomy. However, newer techniques including video assisted thoracoscopic and muscle sparing approaches are emerging. The need to explore the MPA and evacuate vegetation determined our decision to perform a median sternotomy, a manoeuvre that would have been impossible to carry out safely via thoracotomy.

A wide variety of surgical repair techniques, including ligation, patch closure[17] and balloon occlusion method[18] have been utilized in treating adult PDA, influenced mainly by the size, shape, calcification and friability of the PDA encountered. The use of CPB is not always necessary[19] but makes repair less hazardous. Successful repair is also feasible without cardioplegic arrest of the heart, or during electrical ventricular fibrillation.[20] After over-sewing the pulmonary orifice, we employed hypothermic circulatory arrest as we felt it best facilitated the safe, secure repair of a large aortic defect.

Conclusions

Adult PDA can be complicated by pulmonary endarteritis and more rarely by septic embolus to the lung. Radiographic detection of vegetation within the PDA can prove difficult but has important implications with regards to operative strategy, and emphasizes the need for a multimodal imaging approach in PDA assessment. Prophylactic closure of large PDA is mandatory with surgery being favored. However, optimal management of small silent PDA amenable to safe transcatheter occlusion remains unclear, as its behavior characteristics in the modern era are not well defined.

References

1. Mitchell SC, Korones SB, Berendes HW. Congenital heart disease in 56,109 births. Incidence and natural history. Circulation 1971;43:323-32.
2. Gross RE. Surgical management of the patent ductus arteriosus: with summary of four surgically treated cases. Ann Surg 1939;110:321-56.
3. Rigby ML. Closure of a large patent ductus arteriosus in adults: first do no harm. Heart 2007;93:417-8.
4. Campbell M. Natural history of persistent ductus arteriosus. Br Heart J 1968;30:4-13.
5. Actis Dato GM, Cavaglia M, Aidala E, et al. Patent ductus arteriosus. Follow-up of 677 operated cases 40 years later. Minerva Cardioangiol 1999;47:245-54.
6. Marquis RM, Miller HC, McCormack RJ, et al. Persistence of ductus arteriosus with left to right shunt in the older patient. Br Heart J 1982;48:469-84.
7. Schneider DJ, Moore JW. Patent ductus arteriosus. Circulation 2006;114:1873-82.
8. Wiyono SA, Witsenburg M, de Jaegere PP, et al. Patent ductus arteriosus in adults: Case report and review illustrating the spectrum of the disease. Neth Heart J 2008;16:255-9.
9. Thilen U, strom-Olsson K. Does the risk of infective endarteritis justify routine patent ductus arteriosus closure? Eur Heart J 1997;18:503-6.
10. Johnson DH, Rosenthal A, Nadas AS. A forty-year review of bacterial endocarditis in infancy and childhood. Circulation 1975;51:581-8.
11. Parthenakis FI, Kanakaraki MK, Vardas PE. Images in cardiology: silent patent ductus arteriosus endarteritis. Heart 2000;84:619.
12. Balzer DT, Spray TL, McMullin D, et al. Endarteritis associated with a clinically silent patent ductus arteriosus. Am Heart J 1993;125:1192-3.
13. Andrade A, Vargas-Barron J, Rijlaarsdam M, et al. Utility of transesophageal echocardiography in the examination of adult patients with patent ductus arteriosus. Am Heart J 1995;130:543-6.
14. Takenaka K, Sakamoto T, Shiota T, et al. Diagnosis of patent ductus arteriosus in adults by biplane transesophageal color Doppler flow mapping. Am J Cardiol 1991;68:691-3.
15. Cerruto G, Mancuso L. Systemic and pulmonary embolization in a patient with patent ductus arteriosus. Eur J Echocardiogr 2005;6:376-8.
16. Ghani SA, Hashim R. Surgical management of patent ductus arteriosus. A review of 413 cases. J R Coll Surg Edinb 1989;34:33-6.
17. Taira A, Akita H. Patch closure of the ductus arteriosus: an improved method. Ann Thorac Surg 1976;21:454-5.
18. Matsumoto H, Morishita Y, Hashiguchi M, et al. Surgical treatment of the aged patient with patent ductus arteriosus. Kyobu Geka 1989;42:933-5.
19. Wernly JA, Ameriso JL. Intra-aortic closure of the calcified patent ductus: a new operative method not requiring cardiopulmonary bypass. J Thorac Cardiovasc Surg 1980;80:206-10.
20. Toda R, Moriyama Y, Yamashita M, et al. Operation for adult patent ductus arteriosus using cardiopulmonary bypass. Ann Thorac Surg 2000;70:1935-7.