Rupture of a sinus of Valsalva aneurysm due to Aspergillus endocarditis

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We report a case of rupture of an acquired sinus of Valsalva aneurysm secondary to Aspergillus endocarditis in a patient with chronic lymphocytic leukaemia and idiopathic thrombocytopaenia purpura. Despite adequate repair and haemodynamic correction, with an apparent excellent initial result, the patient died of overwhelming sepsis at twelve days postoperatively. This case demonstrates an unusual cause of rupture of a sinus of Valsalva aneurysm and the morbidity associated with immunological dysfunction in the post-operative period.

Key words: Sinus of Valsalva - Aspergillus - Endocarditis.

Aneurysms of the sinus of Valsalva may be congenital, which are often associated with other cardiac anomalies, or acquired most often due to endocarditis. Uncomplicated aneurysms are invariably asymptomatic. Rupture is more common with the acquired variety with a poor outcome without operative intervention.

Case report

A 58 year old male was admitted in congestive cardiac failure. He gave a one month history of fatigue, progressive dyspnoea on exertion and orthopnoea. He had chronic lymphocytic leukaemia (CLL) diagnosed four years previously which had responded to chemotherapy. However this treatment was followed by the development of idiopathic thrombocytopaenia purpura (ITP). The ITP had initially responded to steroid and immunoglobulin treatment, but following a relapse one year prior to his current admission the patient had a splenectomy. Both of these conditions were in remission on this admission. He was in addition an insulin dependent diabetic.

However during these multiple admissions for his haematological disorders, he had never been noted to have any cardiac related problems. His medications at the time of presentation included, azathioprine 150 mg/day, prednisolone 15 mg/day, folic acid 5 mg/day, and penicillin 500 mg/day. He was afebrile and examination demonstrated a tachycardia, raised jugular venous pressure, a tender liver and respiratory crackles to the mid zones bilaterally. The patient had cardiomegaly with a prominent right parasternal heave and a grade 4/6 continuous cardiac murmur.

Chest radiograph demonstrated cardiomegaly and pulmonary oedema. Haematological investigations were normal. The cardiac failure and pulmonary oedema responded to diuretic therapy and the patient improved. Echocardiography demonstrated a ruptured sinus of Valsalva aneurysm with a fistula to the right ventricle and a competent aortic valve.

Left ventricular function was well preserved and no vegetations were seen. Cardiac catheterisation confirmed the diagnosis (Fig. 1) with a step up in oxygen saturations at the level of the right ventricle. Coronary angiography demonstrated a 40% stenosis in the proximal left anterior descending (LAD) coronary artery.

The patient was scheduled for repair of a ruptured sinus of Valsalva aneurysm. At operation the right sided cardiac chambers were dilated with a thrill over the right ventricular outflow tract. There was a ruptured sinus of Valsalva aneurysm extending from the right coronary sinus to the right ventricle. Necrotic and friable tissue was visible in the wall of the aneurysm and it looked infected. There was a solitary vegetation on the right cusp of the aortic valve. The aortic valve was otherwise normal. The right ventricular tract was opened longitudinally, the sinus unroofed and necrotic tissue debrided. The fistula was closed with a Dacron velours patch with interrupted butressed sutures. The vegetation was excised from the right cusp of the aortic valve. The LAD was a very small calibre vessel and was not grafted. Cultures diagnosed Aspergillus endocarditis and the patient was commenced on systemic amphotericin B. Haemodynamically the patient did well initially and was discharged from the intensive care unit on the third post-operative day. On day ten the patient became con-
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Fig. 1.—Angiography demonstrating a ruptured sinus of Valsalva aneurysm with communication to the right ventricle (arrow).

fused, sweaty and hypotensive. He was returned to the intensive care unit and despite inotropic support he continued to deteriorate becoming anuric and requiring dialysis. There was no evidence of infarction on his electrocardiogram. An echocardiograph demonstrated the repair to be intact and mild aortic incompetence but impaired overall myocardial contractility. Despite maximum supportive therapy the patient developed a bradyarrhythmia and died on the twelfth post operative day. Post mortem examination demonstrated extensive Aspergillus endocarditis.

Discussion

The syndrome of acute rupture of a sinus of Valsalva aneurysm was first reported in 1839.3 Lillehei reported the first series of successful repair of ruptured sinus of Valsalva aneurysms in 1957.4 In the majority of cases the aetiology of these aneurysms is a congenital defect in the muscular and elastic wall of the aorta.5 However acquired aneurysms secondary to medionecrosis, penetrating injury, atherosclerosis or as in this case endocarditis have been described. A congenital aetiology is often a diagnosis of exclusion, having failed to identify any other aetiology or an associated cardiac anomaly. In the acquired variety bacterial endocarditis is the commonest cause and without repair the natural history is death within days or weeks.6

Our patient presented with the pathognomonic findings of a harsh continuous murmur over the precordium and congestive cardiac failure. However up to 70% of patients with rupture of a sinus of Valsalva aneurysm experience a more insidious onset of symptoms.7

Echocardiography is an excellent means of diagnosis and identification of any associated ventricular septal defect. Angiography accurately delineates the relationship of the aneurysm to the adjacent cardiac structures.8 Our case presented the commonest origin of these aneurysms, which is the right sinus of Valsalva with rupture into the right atrium or right ventricle.

The objectives of surgical management are to repair the defect and to eradicate the associated left to right shunting. In this case inspection of both sides of the lesion was possible by opening the right ventricular outflow tract and this facilitated adequate inspection and debridement. This is particularly important in cases of endocarditis. A solitary vegetation was excised from the surface of the aortic valve which was judged not to be grossly incompetent and was left intact. This treatment of the aortic valve has been advocated by others, although subsequent replacement of the valve may be necessary.9 Direct closure of the fistula has been associated with a 20-30% reoperation rate for recurrent leakage,10 and therefore patch closure was used in this case.

Overall results are less favourable in the presence of endocarditis,11 of which bacterial endocarditis is the commonest form. Of significance in our patient was his history of CLL and ITP with splenectomy. These conditions are associated with an immunological dysfunctional state 12 and predisposed him to an atypical fungal endocarditis. Post operatively having done well initially with haemodynamic improvement he eventually succumbed to sepsis despite the appropriate antimicrobial therapy. This pattern of haemodynamic improvement but increased predisposition to postoperative sepsis in patients with CLL has been demonstrated following coronary artery surgery.13 Our case demonstrates a ruptured sinus of Valsalva aneurysm associated with an unusual fungal endocarditis. It emphasises the threat of postoperative sepsis despite early diagnosis and adept surgical treatment in the immunocompromised patient.

References

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