Asymptomatic murmurs

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A 57-year-old man presented for a routine medical examination for his non-insulin-dependent diabetes. At the time of examination he was asymptomatic. His diabetes in the past had been well controlled and he had not developed any sequelae. The only physical sign found was a systolic murmur. As a result, he had a transthoracic echocardiogram which revealed mild aortic stenosis with a peak gradient of 10 mmHg and a 'bizarre' right atrial echo, which was thought to be as a result of a hiatus hernia and a barium swallow was arranged. The posterior-anterior and lateral X-rays (figures 1 and 2) showed no evidence of a hiatus hernia but instead a large calcified mass.

Figure 1 Posterior-anterior chest X-ray

Figure 2 Left lateral chest X-ray

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Questions

- 1 What is the most probable diagnosis?
- 2 How does the condition usually present?
- 3 What further investigations and management may be advised?

Answers

QUESTION 1

The posterior-anterior and lateral chest X-rays show a large calcified mass in the region of the right atrium. This was in keeping with the results obtained from the transthoracic echocardiogram. The most probable diagnosis is a calcified aneurysm of the sinus of Valsalva.

QUESTION 2

This defect usually begins as a diverticulum which develops from one of the aortic sinuses. The natural history is for these diverticula to enlarge and rupture. The majority of these ruptures occur after puberty but usually before the age of 30 years. They are more common in males and have been diagnosed between the ages of 11 to 67 years.^{1,2} The physiological consequences depend on the speed at which the rupture develops, the size of the communication, and the chamber into which the communication is made.

In large perforations, death usually occurs within one year. Smaller perforations can initially go unnoticed but usually develop over a period of time. Chronic perforations have been shown to be susceptible to bacterial endocarditis. If the aneurysms are sufficiently large to slow blood flow, then thrombus can form which can affect the right coronary ostium or be a potential source of cerebrovascular embolism.

About 20% of sinus of Valsalva aneurysms are unperforated and present either at *post mortem* or at cardiac surgery.

QUESTION 3

Given the outcome, cardiac surgery is almost always advised. Cardiac catheterisation is, therefore, usually performed, especially in this patient's age group, not only to confirm the diagnosis but also to look for other cardiac pathology such as ischaemic heart disease or aortic root malformations. In our patient, cardiac catheterisation revealed a normal left ventricle, while immediately above the aortic valve lay a large aneurysm of the non-coronary sinus. This was intubated just below the right coronary artery and revealed a smooth appearance with large laminated thrombus extending to the back wall of the aneurysm (figure 3). The total size of the aneurysm was

Figure 3 Aortogram showing the right sinus of Valsalva and its aneurysm almost completely filled with thrombus. The right coronary artery is also visible

6 × 6 × 4 cm. The coronary anatomy was essentially normal.

Transoesophageal echocardiography can also be helpful in making the diagnosis.⁴ This allows both the left and right atriums to be visualised clearly as well as the aortic root, unlike transthoracic echo, and in this case should have been able to distinguish atrial myxoma from an aneurysm of the sinus of Valsalva.

Despite having been present for a number of years, the fact that it almost totally obliterated the right atrial cavity and contained thrombus with a high probability of distal embolisation, it was decided to resect the aneurysm surgically. Surgical mortality is low, especially if no pre-operative aortic regurgitation is present.

Final diagnosis

A large calcified unruptured aneurysm of the sinus of Valsalva (non-coronary) containing laminated thrombus.

Keywords: sinus of Valsalva, aneurysm, cardiac calcification, cardiac catheterisation

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