Case report

Congenital absence of the left pericardium

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Accepted 7 July 1987.

Congenital complete absence of the left pericardium is a rare cause of clinical and radiological cardiovascular abnormalities. We describe a patient in whom this diagnosis was made coincidentally when she presented with another problem.

CASE HISTORY

A 20-year-old female sought advice for diarrhoea and vomiting which followed a holiday in Majorca. She had no other symptoms. Apart from investigation of a cardiac murmur when she was six, past medical history was unremarkable. On examination she appeared healthy, with no fever or dehydration. Her abdomen was normal. Pulse was 100/min, regular, and blood pressure 110/60. Her apex beat was not displaced and of normal character. On auscultation there was wide but variable splitting of the second sound with a loud pulmonary component, and a grade 3/6 mid-systolic murmur heard in the pulmonary area only. Her first heart sound was normal and no other murmurs or bruits were heard.

Electrocardiograph (Fig 1) showed right axis deviation and clockwise rotation. Chest X-ray (Fig 2) showed an abnormal cardiac outline, consistent with leftward rotation. The trachea was central, and lung fields and pulmonary vascularity

Fig 1. Electrocardiograph showing right axis deviation and clockwise rotation of the heart.

Fig 2. Chest X-ray consistent with leftward rotation of the heart.
were normal. There were no abnormalities on echocardiography or 48-hour ambulatory ECG monitoring.

Following referral at the age of six with a systolic murmur heard at the left sternal edge and similar chest X-ray appearance, right and left cardiac catheterisation had been performed. Pressures, oxygen saturations and ventricular contraction had all been normal: in particular there had been no evidence of pulmonary hypertension, pulmonary valve disease, or shunts. No diagnosis had been made.

It was concluded after literature review that her signs and ECG and chest X-ray appearances were consistent with complete absence of the left pericardium. Investigation of her presenting symptoms was negative and they settled without treatment. Six months after presentation she remains well.

DISCUSSION

Congenital pericardial defects are rare. The total number of reported cases is less than 200, and one case was found in a series of 14,000 autopsies. The clinical findings have been described by Nasser.

Complete absence of the left pericardium is thought to result from premature atrophy of the left duct of Cuvier, causing impaired circulation to the pleuropericardial membrane from which it is derived.

ECG changes include right axis deviation, incomplete right bundle branch block, clockwise rotation and tall peaked P waves in the right chest leads. Radiological appearances are characteristic. The heart is shifted to the left, causing apparent cardiomegaly, in the presence of a midline trachea. Prominence of the main pulmonary artery, and interposition of lung between diaphragm and heart and between aorta and pulmonary artery on anterior oblique views may also be seen. Echocardiography is often normal, but may show right ventricular dilatation and paradoxical movement of the interventricular septum. Echocardiography is, however, essential to exclude other cardiac abnormalities which co-exist in one third of reported cases.

The demonstration of a pneumopericardium, after the artificial induction of pneumothorax, had been considered the procedure of choice in the diagnosis of complete left pericardial absence. However, the X-ray appearances are sufficiently characteristic for this potentially hazardous procedure not to be necessary, particularly since most cases are asymptomatic.

Rarely, chest pain and syncope may occur, attributed to lack of cardiac support and subsequent torsion and strain of the great vessels. Abnormal cardiac mobility is also thought to cause the characteristic systolic murmurs heard at the left sternal edge, and the echocardiographic findings.

While complete defects of the left pericardium appear not to have significant morbidity or mortality and require no treatment, sudden death may occur with partial defects. Coronary arteries may be compressed at the margin of a partial defect, and herniation and incarceration of the cardiac apex may occur. With partial defects the chest X-ray is normal, except where cardiac incarceration produces a local bulge in the left cardiac contour.

We thank Dr J G Murtagh and Dr G C Patterson for permission to report the echocardiograph and cardiac catheterisation findings respectively.

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BOOK REVIEW

Davidson's Principles and practice of medicine. 15th ed. Edited by John Macleod, Christopher Edwards and Ian Bouchier. (pp 841. £17.50). Edinburgh: Churchill Livingstone, 1987.

I first read Davidson 30 years ago: at that time it was already in its 5th edition. A brief comparison shows that it then contained about 440,000 words in 1,100 pages: the current edition has compressed 625,000 words into 800 pages. I think I bought it as a student because it was cheap: but also because it seemed to be comprehensive and straightforward, and I have used it as a basis for my medical knowledge ever since.

So I miss some of the old pictures of the acute skin rashes such as scarlet fever: in fact infectious disease has been transferred to the back and genetic factors take the first chapters. But careful linguistic comparison will still uncover the old phrases which some of us know by heart — there are minor changes such as 'alarming reactions to intravenous iron are uncommon, but have occasionally been noted', which becomes 'alarming systemic anaphylactic reactions can occur'.

Sir Stanley Davidson made 'no attempt to describe every rare disease or syndrome, but devoted most of the space available to those disorders most commonly encountered in practice'. I have grown up with successive editions, and have gradually come to appreciate the problems of the authors in the compression of knowledge. Having got to know many of them personally as real people rather than as names I can still recommend the book. It is the essential starting point for the study of internal medicine and for many doctors will remain their base reference work. The present editors have kept up-to-date and been prepared to prune the dead wood. There are many competitors in the market, and the publishers must take care with layout and illustration, although Davidson is still the best value for money.

I will continue to recommend it to my clinical students: they will need to read it and know it to pass final MB. Postgraduates will need to remember the facts, but also to be able to place them in a broader perspective. The older consultant will still happily read it, and to get to know the authors themselves is really to complete your medical education. Dr John Macleod and his team have successfully kept alive the primary objective 'to provide a rational and easily comprehensible basis for the practice of medicine'. The book is economical in price and compact in size, but still contains the essential truths for the practice of good medicine.

DRH

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