Mediastinal yolk sac tumour mimicking pericardial effusion

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In this report we present a case of primary yolk sac tumour of the anterior mediastinum in which two dimensional echocardiography showed an extracardiac mass simulating a large pericardial effusion. Mediastinal yolk sac tumour is a rare germ cell neoplasm affecting mainly young men.¹ The most common presenting symptoms are dyspnoea, cough, and chest pain.² The prognosis is usually poor, though in this case complete resolution of the tumour has been achieved.

Case report

A 26 year old man was referred with a six week history of cough, dyspnoea, and weight loss (12 kg). On examination he was very ill and dyspnoeic at rest and he had tachycardia. His blood pressure averaged 110/70 mm Hg with pulsus paradoxus of 20 mm Hg. The jugular venous pressure was raised 7 cm. The apex beat was impalpable, heart sounds were very faint and there were signs of a left pleural effusion. The edge of the liver was palpable 4 cm below the right costal margin. His testes were not enlarged. Electrocardiography showed a sinus tachycardia and the plain chest radiograph showed a massive, homogeneous soft tissue shadowing in the anterior mediastinum obliterating the cardiac silhouette and displacing the trachea posteriorly (fig 1). There was a small left pleural effusion.

Two dimensional echocardiography was performed with a Hewlett-Packard Model 77020 real time phased array scanner. It showed a large space completely surrounding the heart with what was initially thought to be the fibrinous strands floating within the space (fig 2a). The heart chambers were not dilated and the heart did not show excessive cardiac motion (swinging), as is often seen in malignant or tuberculous pericardial effusion. The appearances were those of a pericardial effusion but pericardial aspiration yielded no fluid.

A right mediastinotomy showed a large necrotic mass surrounding the heart, containing green gelatinous material. The pericardium was intact and no pericardial effusion was found. The histological appearance of biopsied material showed the tumour to be of germ cell type with features suggesting yolk sac tumour (endodermal sinus tumour). The tissue was strongly positive for α fetoprotein, confirming the diagnosis of yolk sac tumour. The serum α fetoprotein was substantially raised at 23 600 U/ml (normal < 10 U/ml).

Six courses of a combination of vinblastine, etoposide, and cisplatin were given, with partial resolution of the tumour

Fig 1 Chest radiograph showing a massive homogeneous shadowing obliterating the cardiac silhouette.

and a fall of the serum α fetoprotein concentration to 1000 U/ml. Six months later surgical resection of the residual tumour was performed but it was incomplete and was therefore followed by radiotherapy. Since then serum tumour markers have been undetectable (α fetoprotein < 10 U/ml). The patient is well two years after presentation.

Discussion

Yolk sac tumour (endodermal sinus tumour) is a subtype of germ cell tumour and a highly malignant neoplasm. In addition to presenting in ovaries and testes, the tumour has been detected at several extragonadal sites, including the presacral area, the anterior mediastinum, and the pineal gland.³ Primary yolk sac tumour of the anterior mediastinum is rare and carries a grave prognosis.^{3,4} Patients often present with advanced, bulky tumours that are unresectable.²

The differential diagnosis of mediastinal masses remains an important clinical problem and patients with a primary mediastinal tumour may present with a clinical and radiographic picture suggesting a primary cardiovascular abnormality.^{5,6} The standard radiological examinations may be inadequate to differentiate between cardiomegaly, pericardial effusion, and adjacent extracardiac mass. The patient

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(a)

(b)

Fig 2 (a) Apical four chamber, two dimensional echocardiogram showing the yolk sac tumour surrounding the heart. (b) Apical four chamber view one year later showing resolution of the tumour. CW—chest wall structures; SP—tumour space masquerading as pericardial effusion; TS—tumour strands; RV—right ventricle; RA—right atrium; LV—left ventricle; LA—left atrium. Arrows indicate the pericardium.

described had a clinical picture of cardiac tamponade and an abnormal cardiac silhouette. Two dimensional echocardiography showed what appeared to be a large pericardial effusion with fibrinous strands and raised the possibility of malignant pericardial effusion. The lesion was found at

surgery to be a large necrotic tumour containing gelatinous material and surrounding the heart.

Various extracardiac tumours have been detected by echocardiography, including mediastinal and pericardial tumours, intrathoracic neoplasms, and pericardial cysts. Pericardial angiosarcoma simulating pericardial effusion on M mode echocardiography has been reported.⁸ Coplan *et al* described the value of two dimensional echocardiography in the diagnosis of a pericardial mesothelioma masquerading as a benign pericardial effusion. An intrapericardial teratoma resembled a pericardial effusion on echocardiography,¹⁰ with a dense layer of echoes at the level of the aortic root. In this case the differentiation between an intrapericardial and extrapericardial lesion was difficult initially. In retrospect two features favoured an extrapericardial mass rather than a pericardial effusion: firstly, absence of the excessive cardiac motion (swinging of the heart) that may be seen in malignant pericardial effusion⁷ and, secondly, the absence of bright echoes surrounding the outer edge of the clear space.

We believe that this is the first report in which the echocardiographic features of an extrapericardial yolk sac tumour surrounding the heart have been described.

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