Case reports

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Ventricular haemangioendothelioma diagnosed in life

D. S. THOMPSON, S. WESTABY, AND J. C. R. LINCOLN

From the Departments of Cardiology and Surgery, Brompton Hospital, Fulham Road, London

A case of right ventricular outflow obstruction is described in which the electrocardiogram and chest x-ray film suggested a cardiac tumour. Cardiac catheterisation confirmed outflow obstruction and angiography showed a tumour. At operation a haemangioendothelioma was found.

Case report

A 10-year-old Libyan girl was referred because of effort dyspnoea. Her parents complained that she was smaller and less active than her sibs.

The patient looked well and was acyanotic. Her height was 1.32 m (15th centile) and weight 26.8 kg (3rd centile). The pulse was in sinus rhythm and normal in character. Blood pressure was 100/70 mmHg. Praecordial examination indicated moderate cardiac enlargement and a systolic thrill at the left sternal edge. The second sound was widely split, moving normally with respiration and there was an ejection murmur loudest in the pulmonary area. There was no ejection click.

The electrocardiogram (Fig. 1a) did not show right ventricular hypertrophy. It showed an abnormal axis of the mean frontal QRS vector (-140°) and an abnormal direction of the mean frontal T vector. The abnormal T vector suggested a myocardial lesion.

The chest x-ray film (Fig. 1b) showed moderate cardiac enlargement with normal lung fields. The echocardiogram showed a greatly thickened ventricular septum.

The physical signs were those of right ventricular outflow obstruction. The electrocardiogram indicated a myocardial lesion and the heart size on the chest x-ray film was large for the degree of obstruction judged to be present. A clinical diagnosis was made of right ventricular outflow obstruction and cardiac tumour.

Cardiac catheterisation (Table) confirmed right ventricular outflow obstruction. Left heart pressures were normal apart from a left ventricular end-diastolic pressure of 15 mmHg. Right ventricular

angiography showed a filling defect in the outflow tract (Fig. 2a). Left ventricular angiography showed a filling defect in its left border (Fig. 2b). The left coronary artery was displaced upwards (Fig. 2c) and a tumour blush was seen beneath it.

The preoperative diagnosis was a tumour involving the right ventricular outflow, ventricular septum, and free wall of the left ventricle.

OPERATION

The heart was exposed through a median sternotomy. A tumour was found protruding from the anterior surface of the right ventricle extending to the lateral aspect of the left ventricle and around the origin of the pulmonary artery. The tumour was pink and nodular and had no capsule. It was firm and the cut surface bled freely. The surrounding myocardium was normal and the pericardium was not involved.

In view of the extent of the tumour, excision was not attempted. A biopsy was taken and the chest closed. The patient made an uneventful recovery.

HISTOLOGY

The tumour was highly cellular with a scanty fibrous tissue stroma (Fig. 3). Capillaries were visible in longitudinal section and their presence was emphasised by reticulin staining. The tumour cells were the same as those lining the capillaries. The appearances were those of an haemangioendothelioma.

Discussion

Haemangioendotheliomata are composed of small

vessels which show proliferation of the endothelial cells. This proliferation may obscure the vascular nature of the tumour. However, the basic structure can be seen by reticulin staining.

It is uncommon for this tumour to arise in the heart. Within the heart the right atrium is the most

frequent site, often with spread into the pericardium. Wider dissemination, especially to the lungs, may occur. Clinical presentation may be as tamponade resulting from obliteration of the pericardial cavity by tumour (Sauerteig, 1958; Crenshaw, Dowling, and Cresswell, 1959) or

Fig. 1 (a) Electrocardiogram, (b) posteroanterior chest x-ray film.

Fig. 2 (a) Right ventriculogram, (b) left ventriculogram showing filling defect, (c) left ventriculogram showing displacement of the left coronary artery.

Table Cardiac catheterisation

Site	Pressure (mmHg)
RV body	43/0
RV outflow	14/3
Pulmonary artery	14/10
Cardiac index	7.0 l/min per m ²

haemopericardium (Tacket, Jones, and Kyle, 1950; Ito, 1964; Allaire *et al.*, 1964). Diagnosis in such cases has been at thoracotomy (Crenshaw *et al.*, 1959; Hansson *et al.*, 1970) or by pericardial aspiration (Tacket *et al.*, 1950). A right atrial haemangioendothelioma has been shown angio-

graphically (Cheng and Sutton, 1955) with histological diagnosis at necropsy. Long-term survival has not been reported though temporary palliation of haemopericardium has been achieved with radiotherapy and cyclophosphamide (Allaire *et al.*, 1964).

The case described in this report is unusual; those previously reported have been in adults, and primary ventricular involvement is rare. At the time of writing, nine months after diagnosis, the patient remains well and the electrocardiogram and chest x-ray film are unchanged. Histological distinction between benign and malignant tumours of this type is difficult. Though the pericardium was not involved and there was no evidence of distant spread, the prognosis remains uncertain.

Fig. 3 Histology of biopsy specimen. Corresponding fields ($\times 120$). (a) (H. and E.), (b) reticulin staining.

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Requests for reprints to Dr. D. S. Thompson, Medical Unit, Queen Mary's Hospital, Roehampton Lane, London SW15 5PN.