Rhabdomyosarcoma of the heart

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Summary

A case of rhabdomyosarcoma of the heart is described, which is one of the rare primary tumours of the heart documented in the literature. The presenting post-mortem appearances were noted.

Introduction

Although a rhabdomyosarcoma (Anderson, 1971) is among the common malignant tumours of the soft parts of the body, it is a rare primary tumour of the heart.

Case report

An 81-year-old male was brought to the Casualty Department, Kingston Public Hospital, Jamaica, and was pronounced dead on arrival. According to the relative, the main complaint of the deceased had been, for some time, recurrent abdominal pain and loss of weight.

External examination revealed the deceased to be moderately built and emaciated. There was no clubbing of the fingers or evidence of external injury or violence. There was marked cyanosis.

Internal examination showed the lungs to be congested and oedematous and there were signs of chronic bronchitis with emphysema. The pericardium and mediastinal glands were normal.

The heart weighed 300 g and the valves were healthy. The coronary arteries were patent but showed arteriosclerotic changes. Marked arteriosclerotic changes were also seen in all the major blood-vessels. There was a bulging mass, yellowish white in colour, in the upper end of the anterior wall of the left ventricle (nearer to the mitral valve). It was a non-polypoid mass measuring $2 \times 1.5 \times 1$ cm and growing from the myocardium and partially filling the left ventricular cavity. The base of the tumour involved the anterior wall of the left ventricle. Microscopically, HE-stained sections showed a fairly cellular tumour extensively infiltrating the heart muscle. The cells were arranged diffusely in some areas and in others were separated by connective tissue septa giving an alveolar pattern. The tumour cells irregularly lined the 'alveoli' with some cells lying free in the lumina (Fig. 1). The cells were round with abundant eosinophilic cytoplasm and

FIG. 1. HE-stained section of the tumour, showing extensive infiltration of the heart muscle.

FIG. 2. HE-stained section showing multinucleated cells and some strap cells.

FIG. 3. PTAH-stained section showing some striated cells.

some cells had vacuolated cytoplasm. Multinucleated giant cells could be seen. Strap cells were seen in some areas (Fig. 2). There were also a few mitotic areas. PTAH-stained sections revealed some striated cells (Fig. 3).

Discussion and conclusion

Prichard (1951) was of the opinion that the status of this tumour had not yet been established. Straus and Merliss (1945) reviewed the world literature on primary tumours of the heart and

concluded that the incidence of this condition was 0.19%. Gould (1968), mentioning the variation in the incidence of primary cardiac tumours, listed various types of which only seven cases were primary tumours of the heart and only one case of rhabdomyosarcoma. The most recent review, which is the one for the years 1951–1973 by Hardin *et al.* (1974) was of seventeen patients with primary cardiac tumours, of which one was a case of rhabdomyosarcoma.

As a rule, primary tumours of the heart are very

rare and this is especially so in the case of rhabdomyosarcoma of the heart. Clinical diagnosis of this condition has not been recorded, and diagnosis before death would depend on the condition causing symptoms brought about by interference with cardiac mechanism. The tumour described in this report was a silent one.

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Aplasia of the right lung and calcifying epithelioma in association with Goldenhar's syndrome

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Summary

A case of Goldenhar's syndrome (oculoauriculovertebral dysplasia) with the rare association of aplasia of one lung is presented with a discussion of the clinical findings and the aetiology. The major abnormalities, pulmonary, renal and the undergrowth of the jaw were all right-sided, confirming previous reports. This child developed a calcifying epithelioma and this is cited as support for the theory of the origin of these lesions from epidermoid cysts.

Introduction

Goldenhar's syndrome (1952) or oculoauriculovertebral dysplasia is a complex syndrome with ocular, auricular, oral and musculoskeletal features (Magalini, 1971). Cardiac anomalies occur occasionally but aplasia of the lung has been reported only once before (Gorlin *et al.*, 1963). Although epibulbar dermoid cysts are frequent, this is the first reported association with a calcifying epithelioma of Malherbe.

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Case report

M.M. presented to the Hospital for Sick Children, Great Ormond Street, London, at the age of 3 months with dyspnoea, recurrent cough and poor feeding, and failure to thrive. Her weight was only 3.4 kg (below 3rd centile). The heart sounds were best heard in the right chest, with a pansystolic murmur at the right sternal margin and a loud pulmonary second sound. The right hemithorax was under-developed and immobile. There were pedunculated accessory auricles along the line from the tragus to the angle of the mouth on both sides. Congenital heart disease with dextro-cardia due to right pulmonary atelectasis was diagnosed.

A chest radiograph showed displacement of the mediastinum to the right and no aeration of the right lung (Fig. 1). Abdominal radiographs were normal. An ECG showed left ventricular hypertrophy with marked axis rotation. Cardiac catheterization, via the inferior vena cava, revealed pulmonary hypertension and a patent ductus arteriosus with bi-directional shunting. Angiocardiography demonstrated a single left pulmonary artery continuous with the parent trunk. The right pulmonary artery was atretic (Fig. 2). A small sequestered right