

MULTIPLE ARTERIAL EMBOLIZATION FROM LEFT ATRIAL MYXOMA IN A MALTESE BOY

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Keywords: Heart, Neoplasm, Myxoma, Embolic, Arterial.

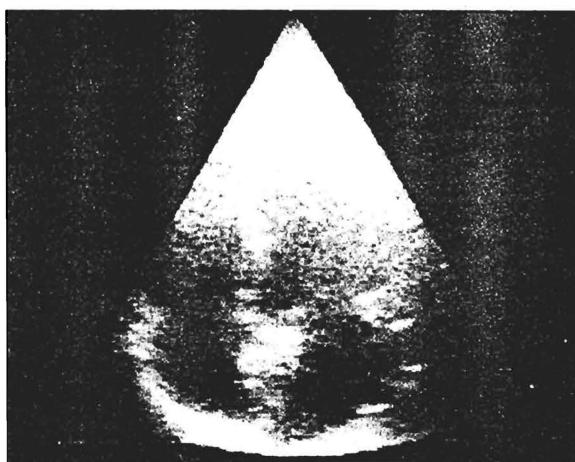
INTRODUCTION

Atrial myxoma is rare in adults and even rarer in children. The presentation may be with embolic, obstructive or constitutional symptoms, which may be dramatic and life-threatening. With present day diagnostic measures, especially echocardiography, diagnosis is not difficult and treatment is curative and rewarding. A case of a left atrial myxoma with multiple simultaneous arterial embolism in upper and lower limbs in a 12 year old Maltese boy is here described.

CASE REPORT

A.C., a 12 year old boy, presented in Paediatric Casualty at St. Luke's Hospital, Malta, with a half-hour history of excruciating pain of sudden onset in the left upper limb and the right lower limb. There was no history of trauma. On examination, he was conscious and co-operative. Peripheral pulses were absent in the affected limbs, which were also pale and cool to touch, BP 170/100 mmHg in left lower limb. No other abnormality was detected on routine systemic examination. The heart sounds were normal. Initially, it was thought that the problems were vascular. A DSA examination showed the right femoral artery occluded by thrombus and the patient was heparinized. An urgent embolectomy was performed. The right lower limb became markedly swollen and painful, and while waiting for fasciotomy to be performed, an echocardiogram showed a large left atrial tumour attached to and moving with the anterior mitral valve leaflet, almost prolapsing into the left ventricle during diastole (see figure). He was therefore transferred urgently to the Hospitals For Sick Children, Great Ormond Street, London, where he had removal of a large left atrial myxoma on cardio-pulmonary bypass. He also had a secundum atrial septal defect which was repaired. He subsequently had left brachial embolectomy (twice) and left femoral embolectomy. He had a stormy post-operative period, complicated by renal failure necessitating peritoneal dialysis which was itself complicated by Klebsiella peritonitis. The right lower limb

became grossly oedematous and split-skin grafts were needed to bridge the fasciotomy scars. Recovery was slow, over several months, with intensive physiotherapy and splinting below the knee.



DISCUSSION

Cardiac tumours are rare in adults and even rarer in children. In a series of 20,305 consecutive echocardiographic studies over a four-year period, only 30 cases of cardiac tumours (0.15%) were detected; 21 of these were primary tumours. 18 of these were benign cardiac myxomas, 90% of them occurring in the left atrium. None of these cases were in the paediatric age group¹.

In over 3,000 echocardiographic studies performed over a five year period in children aged 0-14 years at St. Luke's Hospital, Malta,

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there was only one case of a cardiac tumour (left atrial myxoma reported here).

Cardiac myxomas have been reported at all ages, from a 35 day old infant² to an 83 year old woman. They may be symptomless for an unknown period of time, though the rate of growth, at least in some cases, may be judged from a report of a patient who was found to have a left atrial myxoma, measuring 6 cm x 4 cm, 8 months after a coronary bypass³. The occurrence in a 35 day old infant means that myxomas may be congenital. They are considered to be benign but recurrence has been recorded on more than one occasion, a third recurrence being recorded. They are more common in the left than in the right atrium, and occasionally they may arise in the right or left ventricle.

The tumour may produce a wide spectrum of symptoms and/or signs, either as a result of the local effects of the growth, or by embolization or metastasis. Multiple myxomas, may occur, on both the right and the left sides of the heart. Children apparently tend to have fever and systemic emboli more commonly than adults and the differential diagnosis will then include subacute bacterial endocarditis. If pedunculated, the myxoma may protrude into the atrioventricular orifice during ventricular diastole, thus simulating mitral valve disease in the classical presentation in adults. The surface of the myxoma may be friable and tumour emboli, either pulmonary or systemic, may result. The clinical picture which will be produced may vary greatly according to the site affected. For example, acute lower limb ischaemia may result from arterial embolization in a previously well child⁴. In the case presented here, the sudden onset of severe simultaneous pain in left upper

and right lower limbs was the result of a central cardiac source of multiple, divergent emboli each of sufficient size to occlude the lumen of a large vessel and an echocardiogram clinched the diagnosis. Sometimes, micro-embolization may occur, and their presence may only become manifest months after the atrial myxoma has been excised.

Atrial myxoma is a feature of several syndromes: such as the NAME Syndrome (naevi, atrial myxoma, mucinosis of the skin and endocrine overactivity); syndrome of right atrial myxoma, spotty skin pigmentation and acromegaly⁵; Carney's complex : cardiac myxoma, changes in skin pigmentation and abnormal endocrine function. Moreover, these clinical features may be separated by a period of several years, so that for example a child with Cushing's disease may be found to have an atrial myxoma several years later.

The diagnosis of myxoma is not difficult with present-day non-invasive methods of cardiac investigation. Transthoracic echocardiography is simple, rapid and non-invasive, with a high degree of diagnostic reliability⁶; a transoesophageal probe is useful⁷ if there is an inadequate echo window, but this is rare.

The treatment is by surgical excision on cardiopulmonary bypass as soon as the diagnosis is made. The tumour should be routinely examined histologically, especially to distinguish it from the more aggressive malignant fibrous histiocytoma of the heart⁸. The latter is indistinguishable from myxoma on clinical or echocardiographic grounds, but has a greater tendency for local recurrence and infiltration. Follow up every 6 months by echocardiography is recommended.

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