SURGICAL TREATMENT OF LEFT ATRIAL MYXOMA IN CARNEY'S COMPLEX

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Altough rare, cardiac myxomas are the most frequently encountered primary neoplasms of the heart. Cardiac myxomas are typically sporadic, benign, non recurrent tumours, and they are usually seen in the left atrium.
Carney's complex is a clinical setting associated with cardiac myxomas, spotty pigmented skin lesions, overactive endocrine neoplasms, and noncardiac myxomatous tumors. A 21 year old woman with Carney's complex was admitted to the hospital. The patient had previously gone under open heart surgery, when she was two years old because of Tetralogy of Fallot. The surgical history also included a bilateral adrenalectomy two years ago for primary pigmented nodular adrenocortical tumor that resulted in Cushing's disease. The left atrial myxoma originated from the interatrial septum just above the anterior mitral leaflet. The entire mass and its pedincule was excised with the underlying endocardium. The right atrium and ventricle was explored through a right atriotomy, and no evidence of a recurrent myxoma was noted. The patient was discharged from the hospital seventh postoperative day.
Complex cardiac myxomas have distinctive clinical features that are characterised with unusual biologic behaviors including early development of myxomas at atypical locations, and a high risk of recurance. For these patients all of the heart chambers should be searched for multiple tumors at operation. A close long term postoperative follow-up and screening should be performed for both the patient and family members.

Key words: Complex cardiac myxomas, Carney's complex, Cushing's syndrome

Cardiac myxomas are the most common benign neoplasms of the heart and are found primarily within the left atrium[1,2]. They are isolated, benign, nonrecurring tumors of the left atrium without associated pathological conditions[3] , and they are usually attached to the atrial septum[4]. However, multiple, recurrent, complex myxomatous lesions of the heart has described. In

addition, a subset of patients with cardiac myxomas are recognized and associated with pigmented skin lesions, benign noncardiac myxomatous tumours, and endocrine tumours with overactivity [5]. This association has been called "Carney's Complex" by Carney and his colleques [5-7]. These patients have distinctive features that separate them from the larger group of patients with the more common sporadic myxomas [4].

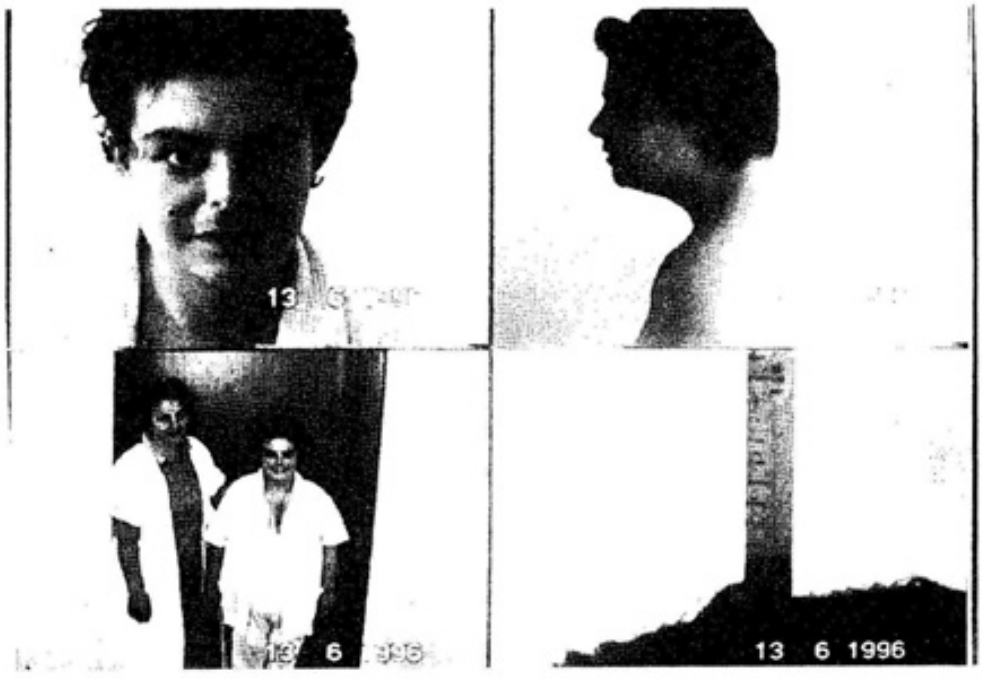
MATERIALS AND METHODS

A 21 year old woman was admitted to the hospital because of a left atrial myxoma. The patient had previously undergone open heart surgery when she was two years old, and total correction of Fallot of Tetrology was performed. The surgical history also included a bilateral adrenalectomy two years ago because of primary pigmented nodular adrenocortical tumour that resulted in Cushing's disease.

There was no family history of Cushing's syndrome or cardiac myxomas. The left atrial myxoma was discovered accidentally on an echocardiographic examination when she had the bilateral adrenalectomy operation. The patient had been treated with corticosteroids following bilateral adrenolectomy.

Her complaints were mild shortness of breath and dyspnea on excertion.

On physical examination the patient had a blood pressure of 140/80 mmHg, pulse rate of 84 beats/min, and a temperature of 36.7°C. There was evidence of marked facial pigmentation, hirsutism, obesity, buffalo hump and stria of the skin (Picture 1). On cardiac examination all of the findings were normal; no heart murmur could be heard. The electrocardiogram demonstrated a sinus rythm with complete right bundle branch block, and biphasic P-waves in V_1 precordial derivation.

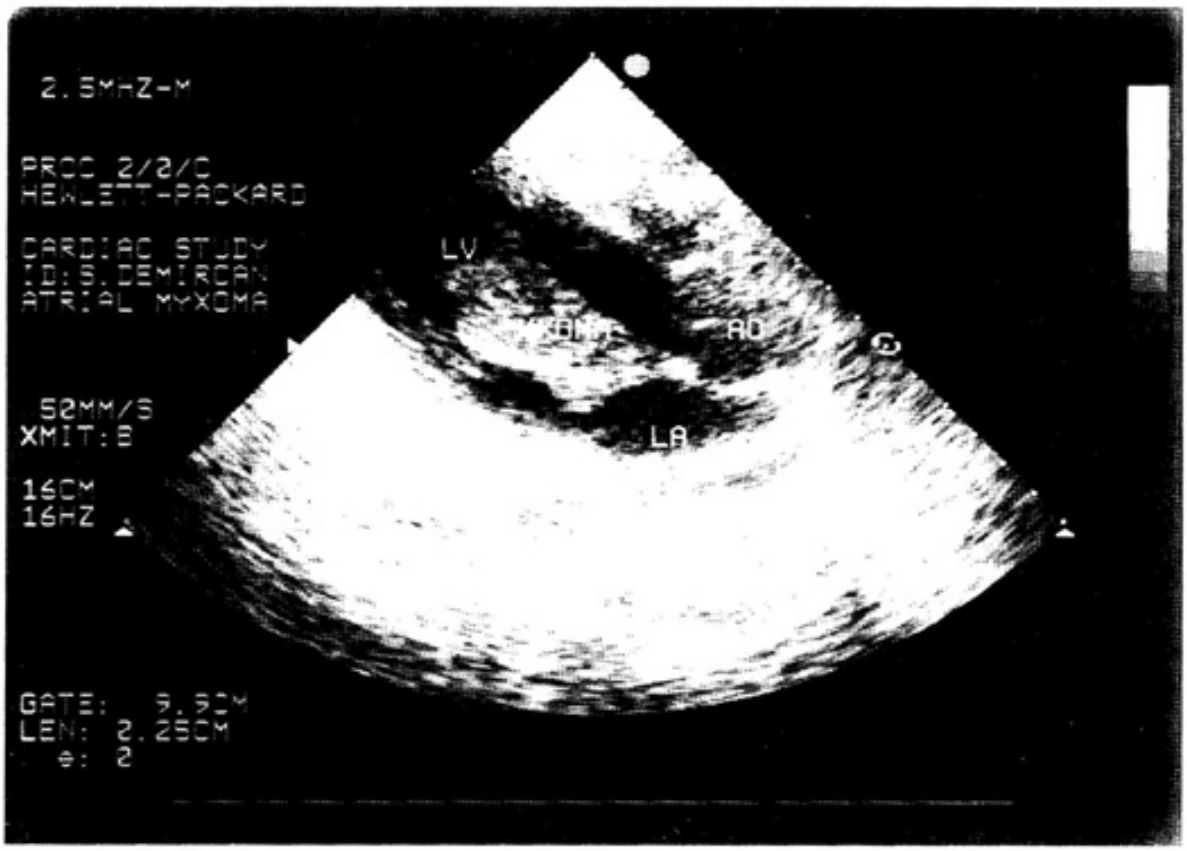


Picture 1: Marked facial pigmentation, hirsutism, buffalo hump, obesity were the main physical features of the patient.

The chest X-ray showed mild cardiomegaly and left atrial enlargement with pulmonary venous congestion. A two-dimensional echocardiography demonstrated a large mass, measuring 3.5 x 4.5 cm which nearly filled the left atrium at the base of the anterior mitral leaflet, and was prolapsing into the left ventricle during diastole (Picture 2).

Diurinal variation of blood cortisol levels remained constant throughout the day since the patient was receiving corticosteroids perorally. Her plasma cortisol levels were in normal ranges (12 and 16 μg/100 ml respectively). The examination of the other heart chambers revealed normal echocardiographic findings. The hematologic examination revealed thrombocytosis (467000/l) and slightly elevated levels of sedimantation (18/50 mm/h).

An open heart operation was performed on June 1996. All of the precautions of cardiac reoperations were foretaken. The femoral region was prepared to be explored just in case of femoral aortic and venous cannulation. The sternotomy was performed with a pneumatic saw. Cardiopulmonary bypass was instituted by aortic and bivenous cannulation. Following moderate hypothermia, the aorta was cross clamped, and cardiac arrest was induced using antegrade isothermic blood cardioplegia. The left atrium was opened with a standart incision. The myxomatous tumour prolapsing from the left atrium was seen. The mass measured 4 x 6.5 x 5.1 cm was arising from the interatrial septum just above the anterior mitral leaflet with a 1 cm stalk. The tumour was excised with the entire pedicle and the underlying endocardium (Picture 3). The mitral valve seemed to be uneffected from the tumor. Following the closure of left atrium, the right atrium was explored and was found to be normal. The post operative course of the patient went uneventfully. The patient was discharged from the hospital 13 days later. A 18 month follow up of the patient reveals no reccurence of the tumor.



Picture 2: Echocardiographic view of the patient.

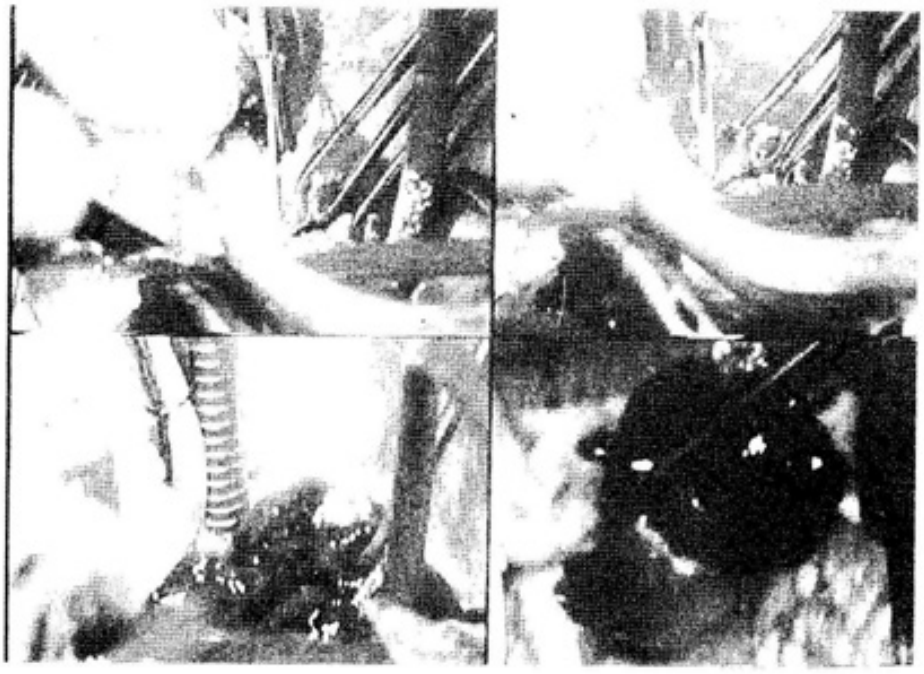
DISCUSSION

Cardiac myxomas are the most common form of primary cardiac tumors[8]. Most cardiac myxomas occur sporadically, and usually are seen in middle-aged women as an isolated tumor in the left atrium without accompanying pathological conditions [3]. A group of special findings in a subgroup of patients with cardiac myxomas has made the subject more interesting. Familial occurence may be a special feature in some patients. In this case familial occurence was not encountered. This complex was first recognized with lentigines and addition of cutaneous myxomas recognized as NAME (nevi, atrial myxoma, myxoid neurofibromma, ephelides) and LAMB (lentigines, atrial myxoma, blue nevi) syndromes. Later endocrine overactivity and spotty pigmentation was also recognized as a special feature to the myxoma complex which was first described by Carney and associates [6,11]. The complex includes seven rare conditions: (1) cardiac myxoma; (2) skin myxomas; (3) myxoid mammary fibroadenomas; (4) spotty skin pigmentation (lentigo and other types of nevi); (5) primary pigmented nodular adrenocortical disease, a cause of Cushing's syndrome; (6) pituitary adenoma; and (7) unusual testicular tumors, especially large cell calcifying Sertoli cell tumors.

Familial occurrence of the complex has been described. It now seems likely that the familial form is inherited as autosomal dominant trait[3,9] and another hypotesis of transmission involves viral infection with the Coxsackie B4 virus [10].

The cardiac myxomas of the Carney complex are different from the sporadic nonfamilial tumor: the patients are much more younger, myxomas develop in atypical regions, shows reccurence and have multicentric origin [11].

Any patient less than 40 years of age with a



Picture 3: Operative views from the operation. Excision of the entire pedicle and the underlying endocardium of the tumor, the stalk, and abortion of the myxomatous tumor from the left atrium.

cardiac myxoma at a site other then the left atrium should be suspected of having a syndrome form of myxoma. The integrity of the adrenal gland should be confirmed preoperatively, and routine intraoperative suspection of the other cardiac chambers for multiple myxomas should be conducted. Any patient with familial or nonfamial, recurrent or multicentric myxomas combined with cutaneous pigmentations or peripheral noncardiac myxomas should be closely followed up for recurrent disease.

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