

# Multicentric Familial Cardiac Myxoma

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## Abstract

Familial cardiac myxoma is a rare syndrome which constitutes approximately 10% or less of all myxomas. We describe a rare case of a simultaneous left atrial and left ventricular mass in a 35-year-old female who presented to our hospital for the evaluation of recurrent cardiac myxoma. Echocardiography revealed a concurrent left atrial and left ventricular mass. Histological findings after surgery confirmed the diagnosis of myxoma.

**Key words:** myxoma ■ cardiac ■ echocardiography ■ Carney's syndrome

### Case report

A 35-year-old woman was referred to our echo lab for the evaluation of dyspnea. She had a previous history of two cardiac surgical operations due to confirmed left atrial (LA) myxoma. She had suffered from dyspnea on exertion for one month and transient paresis of the left hand 3 days before admission. She explained some transient tender palmar and plantar maculas with spontaneous resolution. She had a history of CVA and right hemiplegia with flexure contracture of the distal part of the right hand 18 years before her first surgery. A second cardiac surgical operation had been performed 2 years previously upon the occurrence of transient cutaneous symptoms and dyspnea as a result of recurrent myxoma. The family history reads very interesting inasmuch as both her 57-year-old mother and 33-year-old brother had a history of cardiac surgery on account of confirmed LA myxoma, the former undergoing cardiac surgery 4 years before and the latter twice - first 9 years and then one year previously in another center. The patient's physical examination on presentation revealed BP 130/80, HR 78 bpm, RR 14/min and normal jugular venous pressure. There was facial freckling

and a dark macula (2x3mm) on the right buccal mucosa: two skin tags were seen on her body. On cardiac auscultation, S1 and S2 were normal with an S4 gallop. Lung and abdominal exams were normal. Right hand paralysis with flexure contracture was seen as a sequel following her first presentation. While there was right foot paresis, other extremities were normal. ECG showed NSR, normal axis and no significant pathologic change. Lab tests revealed mild hypochromic anemia with increased ESR; Hgb 9.5g/dL; Hct 35%; RBC=4,700,000; MCH=20; MCV=75; MCHC=27; ESR=36; IRON=23 (N); TIBC=88 (increased) and FERRITIN=28 (N). Other lab tests were normal. chest X-ray was within normal limits.

### Echocardiographic findings

While the findings demonstrated normal left ventricular (LV) size and function, there was a large semi-mobile mass (1.9 x 1.4cm) in the LV cavity, which was attached to the anterolateral papillary muscle. Right ventricular size was mildly enlarged, but the function was normal. The mitral valve had mild regurgitation with normal orifice area. There were two clusters of mobile mass in the LA.

The larger one (3 x 2cm) was highly mobile and was attached by a narrow stalk to the fossa ovalis. The smaller one (1.8 x 1cm) was partially mobile and was attached to the anterior mitral leaflet's base. Neither of them was prolapsing to the LV. Other cardiac structures were normal (Figs. 1, 2).



**Fig. 1:** Transesophageal long axis view shows mass attached to anterior mitral leaflet's base.



**Fig. 2.** Transgastric short axis view shows mass attached to anterolateral papillary muscle.

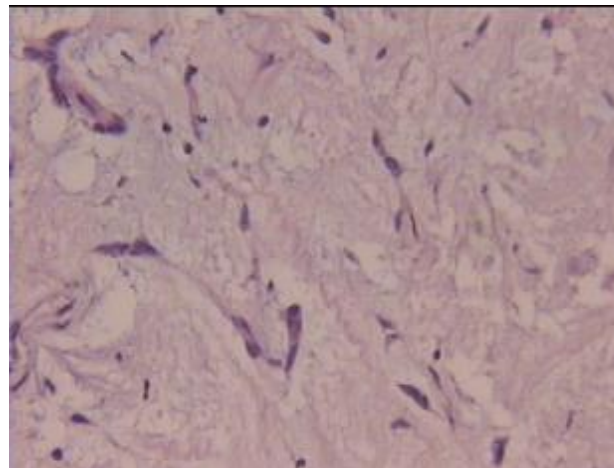
**Operative findings**

There was a jelly-like LA mass with attachment to the intraatrial septum by a pedicle and a cluster of mass attached without a pedicle to the anterior mitral leaflet. LV evaluation revealed two jelly-like masses with attachment to the anterior papillary muscle without pedicles.

**Pathologic findings**

**Gross:** The specimen consisted of multiple irregular fragments of creamy-brown, soft to firm and gelatinous tissue (3x3x1cm) in aggregate. Grossly foci of hemorrhage are seen.

**Microscopy:** Hypocellular tumor tissue is seen with a myxoid background. There were some isolated spindle and stellate cells which formed vessel-like structures and groups of cells in some areas. No mitosis or pleomorphism was observed (Fig. 3).



**Fig. 3.** Microscopy of mass showed myxoma.

**Discussion**

Familial cardiac myxomas appear to have an autosomal dominant transmission. Syndrome myxoma or Carney's syndrome consists of: 1-myxomas in other locations (breast or skin) 2-spotty pigmentation (lentigines, pigmented nevi or both) and 3-endocrine over-activity (pituitary adenoma, primary pigmented nodular adrenocortical disease, or testicular tumors involving the endocrine components). Patients with Carney's syndrome tend to be younger (mean age 20) and are more likely to have myxomas in locations other than the left

atrium. They sometimes have bilateral tumors and are more likely to develop recurrences. Although the cause of the syndrome myxoma is unknown, it has been suggested that it results from a widespread abnormality leading to an excessive proliferation of certain mesenchymal cells, and excessive glycosaminoglycan production by them, possibly analogous to the neural masses in Von Recklinghausen's neurofibromatosis. Patients may have two or more components of this complex, and the first component is generally diagnosed at a relatively young age (mean age 18 years). Some patients have been said to have the NAME syndrome (nevi, atrial myxoma, myxoid neurofibroma, ephelides) or the LAMB syndrome (lentigines, atrial myxoma and blue nevi). In patients who have a familial history or other components of the previously described syndrome and who are undergoing resection, not only is a careful preoperative search for several cardiac myxomas strongly recommended but also a close postoperative observation for the possible development of other tumors seems advisable.

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