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CASE REPORTS

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 LA and LV mass simultaneously in a 35-year-old female presenting to our hospital for evaluation of recurrent cardiac myxoma. Echocardiography revealed both LA and LV mass. Surgery was done and histological findings confirmed the diagnosis of myxoma.

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 surgery due to confirmed 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 distal part of the right hand 18 years ago (at the time of the first surgery). Second cardiac surgery was done 2 years ago when transient cutaneous symptoms and dyspnea occurred as a result of recurrent myxoma. The family history reads interesting in as much as her mother (57-year-old) had a history of cardiac surgery (4 years ago) because of confirmed LA myxoma. Her brother (33-year-old) also had a history of cardiac surgery twice for confirmed LA myxoma about 9 years and then 1 year before. The patient's physical examination on presentation revealed: BP = 130/80, HR = 78 bpm, RR = 14/min, normal jugular venous pressure. There were facial freckling and a dark macula (2 × 3 mm) on right buccal mucosa, two skin tags were seen on her body. S1 and S2 were normal with S4 gallope. 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 paresia, other extremities were normal. ECG showed NSR-normal axis and no significant pathologic change. Lab test showed mild hypochromic anemia with increased ESR in first test: Hb = 9.5 mg/dL, HCT = 35%, RBC = 4 700 000, MCH = 20, MCV = 75,

Abbreviations: LA, left atrium, LV, left ventricle, MV, mitral valve, IAS, interatrial septum, RV, right ventricle, NSR, normal sinus rhythm, MR, mitral regurgitation.

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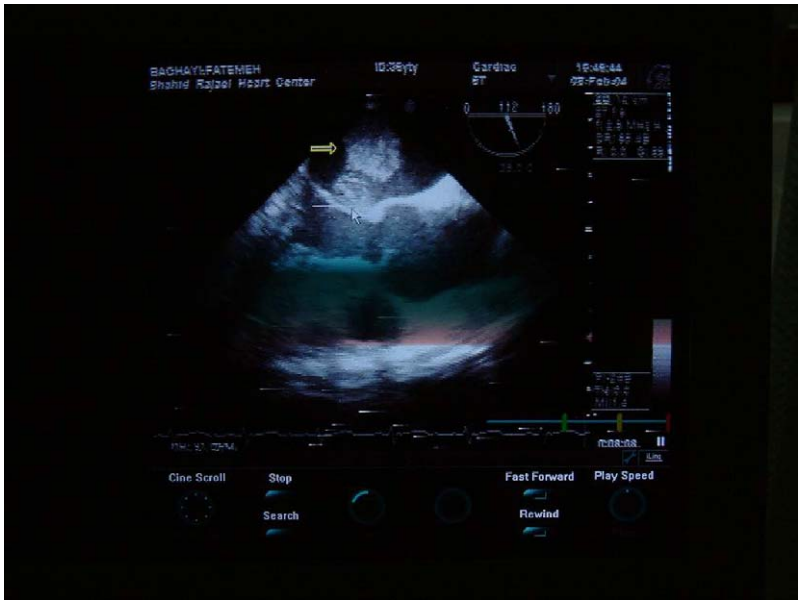

Figure 1 Transesophageal long axis view showing mass attachment to anterior mitral leaflet's base.

MCHC = 27, ESR = 36, IRON = 23(N), TIBC = 88 (increased), FERRITIN = 28(N), other lab tests were normal. Chest X-ray was within normal limits.

Echocardiographic findings

While the findings demonstrated normal LV size and function, there was a large semi mobile mass (1/9 × 1.4 cm) in LV cavity which was attached to anterolateral papillary muscle. RV size was mildly enlarged with normal function. Mitral valve had mild MR. There were two clusters of mobile masses in LA. The larger one (3 × 2 cm) was highly mobile and was attached to the fossa ovalis by a narrow stalk. The smaller one (1/8 × 1 cm) was partially mobile which was attached to the anterior MV leaflet's base. Neither of them was prolapsing to the LV. Other cardiac structures were normal (Figs. 1 and 2).

Operative findings

A jelly-like LA mass was seen with attachment to the interatrial septum by a pedicle and a cluster of mass attached without a pedicle to the anterior

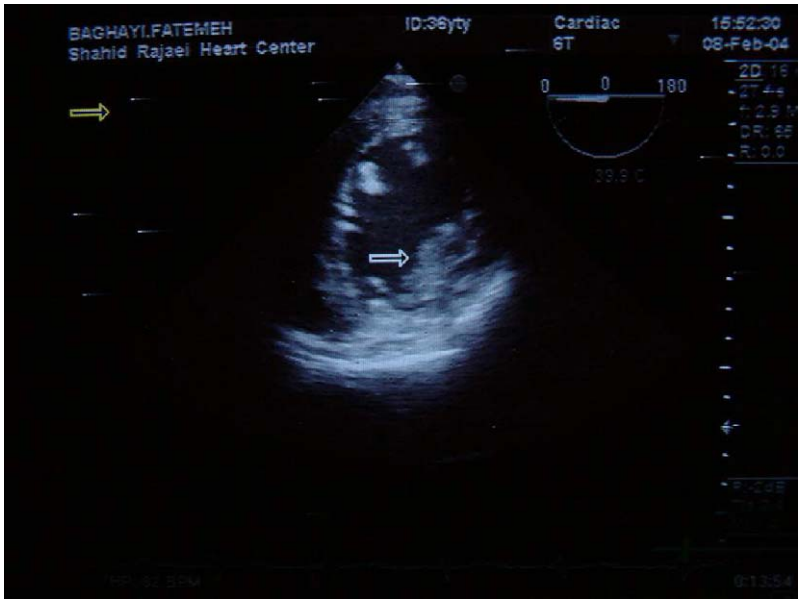

Figure 2 Transgastric short axis view showing mass attachment to anterolateral papillary muscle.

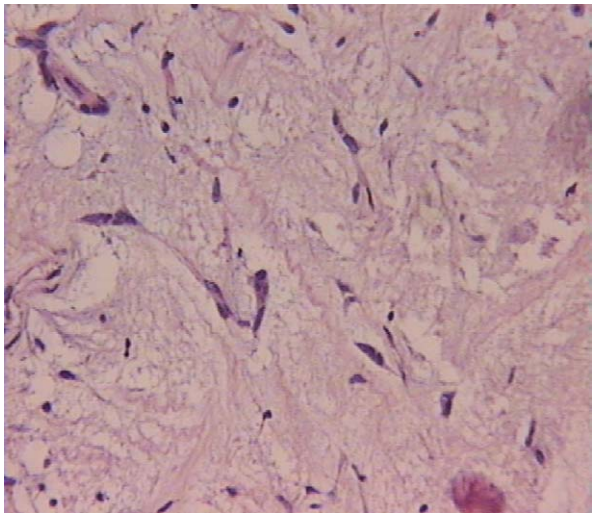

Figure 3 Microscopy of mass showing myxoma.

mitral leaflet. LV evaluation revealed there were two jelly-like masses with attachment to the anterior papillary muscle without pedicle.

Pathologic findings

Gross. The specimen consisted of multiple irregular fragments of creamy-brown, soft to firm and gelatinous tissue ($3 \times 3 \times 1$ cm) in aggregate. Gross foci of hemorrhage were detected.
Microscopy. There were both hypocellular tumor tissue with a myxoid background. There were some isolated spindle and satellite cells which formed vessel-like structures and groups of cells in some areas. No mitosis or pleomorphism was observed (Fig. 3).

Discussion

Familial cardiac myxomas appear to have an autosomal dominant transmission.[1,2] Syndrome myxoma or Carney's syndrome also consists of myxomas in other locations (breast or skin), spotty pigmentation (lentigines, pigmented nevi or both), and endocrine overactivity (pituitary adenoma primary pigmented nodular adrenocortical disease, or testicular tumors involving the endocrine components[3–6]). Patients with Carney's syndrome tend to be younger (mean age 20), 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 proposed to result from a widespread abnormality resulting in excessive proliferation of certain mesenchymal cells, and excessive glycosaminoglycans production by them, possibly analogous to the neural masses in Von Recklinghausen's neurofibromatosis.[7] Patients may have two or more components of this complex and the first component generally is 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)[2,8] or the LAMB syndrome (lentigines, atrial myxoma and blue nevi).[9,10] In patients who have a familial history or other components of the previously described syndrome and who are undergoing resection, a careful search should be made preoperatively for several cardiac myxomas. Postoperatively these patients should be observed closely for the development of other tumors.

References

1. Carney JA, Hruska LS, Beauchamp GD, Gordon H. Dominant inheritance of the complex of myxomas, spotty pigmentation and endocrine overactivity. *Mayo Clin Pro* 1986;**61**:165.
2. Koopman RG, Happle R. Autosomal dominant transmission of the NAME syndrome (nevi, atrial myxoma, mucinosis of the skin and endocrine overactivity). *Hum Genet* 1991;**86**: 300.
3. Carney JA, Gordon J, Carpenter PC, et al. The complex of myxoma, spotty pigmentation and endocrine overactivity. *Medicine* 1985;**64**:270.
4. Bennet WS, Skelton TN, Lehan PH. The complex of myxomas, pigmentation and endocrine overactivity. *Medicine* 1990;**65**:399.
5. Cohen C, Turner ML, Stratakis CA. Pigmented lesions of the conjunctiva in Carney complex. *J Am Acad Dermatol* 2000;**42**:145.
6. Watson JC, Stratakis CA, Bryant-Greenwood PK, et al. Neurosurgical implications of Carney complex. *Neurosurg* 2000;**92**:413–8.
7. Carney JA, Benas CT. Myxoid fibroadenoma and allied conditions (myxomatosis) of the breast. *Am J Surg Pathol* 1991;**15**:713.
8. Vidaillet Jr HJ, Seward JB, Fykle E, Tajik AJ. NAME syndrome (neviatrial myxoma, myxoid neurofibromatosis, epheliedes): a new and unrecognized sunset of patients with cardiac myxoma. *Minn Med* 1984;**67**:695.
9. Rhodes AR, Silverman RA, Harrist TJ, Perez-Atayde AR. Mucocutaneous lentiginies, cardiomucocutaneous myxoma, and multiple blue nevi: the LAMB syndrome. *J Am Acad Dermatol* 1984;**10**:72.
10. Braunwald. *Heart disease, textbook of cardiovascular medicine;* 2001. p. 1811–2.