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NONSYNDROME FAMILIAL ATRIAL MYXOMA IN TWO GENERATIONS

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Since the first report of a family with cardiac myxomas in 1971 by Krause and associates,¹ 17 such families have been reported worldwide.² Among these there are only two cases in which four members in a single family had cardiac myxomas.^{2,3} A high percentage of families also have noncardiac cutaneous and endocrine manifestations, which are termed *Carney syndrome*.⁴ We report a case in which four members in a family are involved without any of the extracardiac features described in the syndrome.

A 17-year-old girl (B-F6) was brought to our emergency department with signs suggestive of pulmonary edema in November 1991. On a review of her records we found that she was being treated with decongestive therapy in our cardiology clinic for symptoms of dyspnea and palpitation with a clinical diagnosis of mitral stenosis of rheumatic origin. An echocardiogram performed immediately showed a large myxoma in the left atrium prolapsing in and out of the mitral valve. After her condition was clinically stabilized, an electrocardiogram-gated magnetic resonance imaging study with contrast medium was performed. This confirmed the diagnosis of left atrial myxoma arising from the interatrial septum. She was immediately taken to the operating room. With the aid of cardiopulmonary bypass and cardioplegic arrest, a right atriotomy was performed and the interatrial septum from which the stalk originated, along with the tumor, was excised with a 1 cm margin. The defect was closed with a Dacron patch. A thorough search in all chambers of the heart did not reveal multiple tumors. The tumor was 10 by 8 cm and did not deviate from the usual sporadic myxoma pattern by either gross or histopathologic examination. The excised tumor was placed in a glass container filled with saline and was subjected to magnetic resonance imaging with tissue spectroscopy. These images also did not show any variation when compared with those of a

sporadic myxoma. The patient made an uneventful post-operative recovery.

On investigating the family history we learned that her elder brother (B-M2) underwent a similar excision of left atrial myxoma at our institute in 1979 at the age of 14 years when he presented with left hemiparesis and seizures after a cerebral embolism. He is currently doing well without any evidence of recurrence and taking only antiepileptic medication. On further inquiry we learned that the eldest of the siblings (B-M1) had died suddenly while running when he was a 12-year-old boy in 1968. He had a history of dyspnea on exertion for a short period before his death and proper medical attention was not sought. No autopsy was performed.

At this point we decided to investigate the whole family. When we did an echocardiogram on the mother (A-F1) and father (A-M1) of those patients, the 55-year-old mother was found to have a biatrial myxoma with the stalks growing from either side of the interatrial septum. She was completely free of symptoms. The father (A-M1) had a normal study. In the second generation of the three living elder sisters of the operated patients two, (B-F3 and B-F4) had a normal study. The third sister (B-F5) refused to undergo any kind of medical evaluation for social reasons. In the third generation the two children of (B-F3) had a normal study. The two third-generation children of (B-F5) could not be evaluated along with their mother. None of the other second-generation subjects have children. A thorough physical examination was performed in all members of the family with emphasis on any skin or mucocutaneous pigmentation, cutaneous or breast tumors, and any clinical signs of endocrine hyperactivity. Serum levels of various adrenocortical, pituitary, and thyroid hormones were done. We could not find any evidence of Carney syndrome in any member of the family (Fig. 1).

The mother (A-F1) was subjected to contrast magnetic resonance imaging with tissue spectroscopy (Fig. 2), which confirmed the diagnosis of biatrial myxoma. She was taken to the operating room. Cardiopulmonary bypass was established with bicaval cannulation. The right atrium was opened, a 3 by 3 cm myxoma arising from the posterior wall of the right atrium was excised

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Fig. 1. Family members of the first and second generations, marked as 1 and 2, with one female member in the second generation missing. Note the absence of any kind of skin pigmentary or nodular changes.

Fig. 2. Magnetic resonance image of (A-F1), showing a left atrial myxoma arising from the interatrial septum. The right atrial myxoma cannot be well appreciated in this view.

with a margin of tissue, and the defect was closed directly. The left atrial myxoma, 4 by 3 cm, along with its stalk arising from the fossa ovalis region of the interatrial septum, was excised with a margin of the tissue, and the defect was closed with a Dacron patch. A thorough search did not reveal any tumors in other chambers of the heart. The pathologic and the magnetic resonance images of this tumor did not deviate from any other sporadic myxoma. The patient made an uneventful postoperative recovery. Currently the family

is being followed up with annual echocardiographic and endocrine evaluations.

van Gelder and associates,² in a review of world literature, found 15 families with myxoma of the heart, to which they added two more. Typically, the patients were young (mean age 27 years), with multicentric lesions in 22% of cases. The majority of these tumors occurred in the left atrium (61%), with a recurrence rate of 10% after removal. The mode of inheritance in these families appears to be autosomally dominant.

Thirty-five percent of these families had extracardiac manifestations in the form of skin nevi, lentigines, cutaneous myxomas, myxoid mammary fibroadenomas, Cushing's syndrome caused by nodular adrenocortical hyperplasia, testicular tumors (Sertoli cell tumors), and thyroid and uterine lesions, in addition to hemolytic anemia and thrombocytopenia. These features are in deference to an isolated classic cardiac myxoma that typically occurs in elderly women and tends to be the only lesion in 95% of the cases. The majority (76 to 88%) of the lesions arise in the left atrium.

In our case we believe that the 12 year old boy (B-M1) in the second generation who died suddenly after having cardiac symptoms had cardiac myxoma. The mean age at diagnosis in this family is 24 years, with a multicentric lesion in one case, although the left atrial location was a feature in all cases. So far there is no evidence of recurrence. The mode of inheritance in this family appears to be autosomal dominance. Complete absence of any of the syndrome features is notable in this family. The occurrence of a biatrial myxoma in our family is also significant, with only nine cases of suc-

cessful removal noted in the literature.⁵ A close watch needs to be kept for the development of cardiac tumors in other members of this family, especially in the third-generation siblings.

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