

Repeated Recurrence of a Left Atrial Myxoma: a Case Report



Medical Science

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ABSTRACT

Cardiac myxomas are benign tumors of the heart that commonly occur in the left atrium. Myxomas can be either sporadic or as part of a familial syndrome. Surgical extirpation of myxomas is usually curative and recurrences are rare. Exceptions are noted with familial tumors. We present a case of a woman with a second recurrence of a left atrial myxoma. Her tumor was removed completely and histologic analysis confirmed the diagnosis of myxoma. Repeated recurrence suggests an invasive and aggressive nature of at least some myxomas. That is why all myxomas should be completely resected with a cuff of underlying endocardium or atrial septum.

Introduction.

Cardiac myxoma is a benign tumor of the heart with an average frequency 0.5 – 1 per million per year [1]. It has been suggested that it originates from a mural thrombus although most evidence favours a neoplastic origin. The most common location is the left atrium and the tumor is either sessile (about one third) or attached by a stalk to the interatrial septum (two thirds), typically arising high near or at the fossa ovalis [2,3,4]. At some cases the tumor may originate from the posterior left atrial wall. Other locations of attachment include valve leaflets, right atrial, right or left ventricular wall. 75% of myxomas originate in the left atrium, 15 to 20% in the right atrium, and only 3 to 4% in the ventricles [5]. Myxomas are usually soft, whitish, easily fragmentable tumors that do not infiltrate beyond the endothelial layer of the endocardium. Their growth rate is fairly rapid since they cause a high transatrial gradient without appreciable enlargement of the left atrium [6].

Nearly all solitary myxomas are sporadic or nonfamilial and recur in only about 1 to 3% of patients [7,8]. They have a normal DNA ploidy and are disorders primarily of middle-aged women. Seven percent of myxomas exhibit a familial pattern as a part of a complex syndrome known as Carney complex or Swiss syndrome. These patients may present with spotty pigmentation of the skin (lentiginos), atrial myxomas, cutaneous myxomas, blue nevi, and endocrine tumors (pituitary adenoma, adrenal adenomas, Sertoli cell tumors). Familial myxomas have a strong tendency to recur in 30 to 75% of patients [7]. Familial myxomas are aneuploid in all cases and are common in young men.

Since the first successful removal of an intracardiac myxoma in 1955, there have been numerous reports of the techniques and results of the surgical treatment of these tumors. Recurrence of a myxoma after successful extirpation is not frequently reported in the literature and surgical excision of nonfamilial myxomas is usually curative. However familial myxomas have a strong tendency to recur [8] even twenty years after excision [2]. Overall recurrence rate has been estimated at about 3% and repeated recurrence at about 1.3% [1]. Recurrence may be due to inadequate resection, intraoperative implantation (seeding at the time of removal), multicentric growth (growth from a new focus), or familial tumor type [9]. Complete excision of the tumor with its pedicle along with a wide cuff of atrial septum and copious irrigation of the cardiac chambers with saline to remove tumor debris helps to avoid recurrence [10, 11]. Careful handling of the myxoma is paramount in preventing implantation metastases

and peripheral embolization of tumor fragments [12].

Malignant transformation of intracardiac myxomas is still under discussion but when it occurs tumor recurrence is usually extensive and multifocal [13].

Follow up echocardiography is recommended in all myxoma cases to assess for recurrence.

Cardiac myxomas characteristically present with symptoms of intracardiac obstruction, hemodynamic alterations, cerebral or peripheral embolization, syncope or sudden death, and constitutional symptoms such as fever, weight loss, fatigue, loss of appetite or anemia.

Case report. We operated on a 59-year old woman who presented with a second recurrence of a left atrial myxoma. It was two years after the first recurrence and five years after the primary tumor was diagnosed which were both treated at another institution. Our patient's primary complaints were dyspnea on exertion and decreased physical capacity. Physical examination was unremarkable. Two-dimensional transthoracic echocardiography revealed a tumor mass in a mildly enlarged left atrium attached to the septum and measuring 4/2.4 cm.

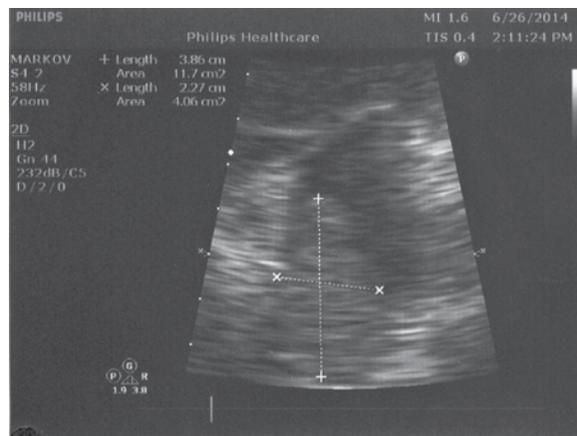
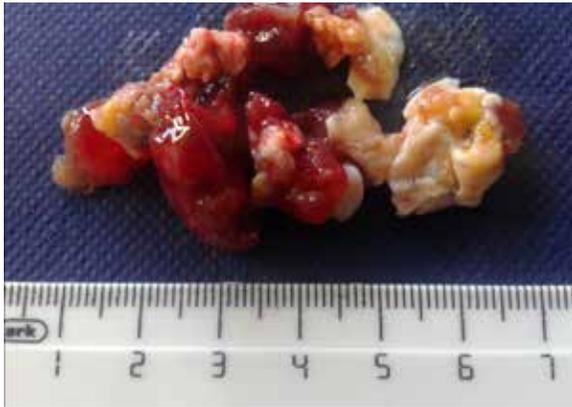


Fig.1 An echocardiographic image of a left atrial mass attached to the septum that proved to be a myxomatous tumor.

There was mild mitral regurgitation. At operation through a

right atrial transseptal approach a soft and friable, sessile, multicentric myxoma was discovered. It was attached to the septum and posterior left atrial wall. The tumor was removed fragmented but completely along with a wide cuff of the thickened septum.

Fig.2 The extirpated tumor in fragments. The reddish part is tumor tissue and the whitish parts represent the previously inserted pericardial patch; a suture can be seen.



Great care was taken not to manipulate heart before the aorta was cross-clamped and to remove any tumor tissue from the left atrial wall. The ventricles were explored and no concomitant myxomatous tissue was found. The atrial septal defect was closed with a preserved bovine pericardial patch.

Postoperative recovery was uneventful and echocardiography confirmed no residual tumor. Despite the wide excision of the atrial septum our patient had no conduction disturbances. However, she developed atrial flutter consistent with the atrial incisions made.

Histologic examination was conclusive of a typical myxoma with no signs malignancy or invasion of adjacent septal tissue.

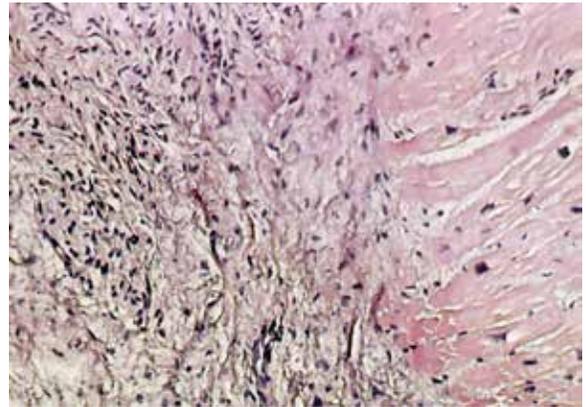


Fig 3. Borderline zone: myocardium - myxoma. Without mitotic activity. H&E stain, x250

Conclusion. Myxomas are benign tumors that rarely recur even if they show some signs of invasiveness. Repeated recurrence is even more rare. However, studies have shown that surgical techniques involving radical removal of tumor and its surrounding normal tissue ensure minimal recurrence rates. Our patient presented for a third-time cardiac surgery because of her myxoma. Since no family history was obtained and multicentric growth was evident we concluded that we dealt with a case of a tumor caused by a sporadic genetic mutation. We also favoured multicentricity and incomplete resection as the causes of tumor recurrence. No genetic testing was conducted. Thus we performed complete removal of the tumor along with a wide portion of healthy subendocardial tissue to minimize any possibility of future recurrence. Our case implies that at least some myxomas may have a stronger tendency to recur. That is why all myxomas should be completely resected with a cuff of underlying endocardium or atrial septum.

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