

CLINICAL MANIFESTATIONS IN ATRIAL MYXOMA

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ABSTRACT

In Nepal, we started regular open-heart surgery since February 1997. Primary tumors of the heart are rare and the commonest is myxoma. Left atrium is the commonest site of occurrence. There is diagnostic difficulty in this most frequent benign tumor of the heart.

We present 5 cases of left atrial myxomas who presented with different clinical features. Among them 4 cases were diagnosed preoperatively by transthoracic echocardiography.

The youngest was 32 years whereas the oldest was 58 years (mean 49.8 years) and 4 of them were female. Among them, one presented with right unilateral pulmonary venous hypertension; another mimicking mitral stenosis with large left atrial clot; with embolization to brain with motor the third aphasia and the remaining two had only constitutional symptoms. All of them were discharged with successful removal of tumor and are surviving at present with no features of recurrence.

There are no pathognomonic features of atrial myxoma. All suspected cases must be subjected for echocardiography. With the advent of open-heart surgery, extirpation of tumors with good outcome has been a regular service in our hospital.

Key Words: Atrial myxoma, Transthoracic Echocardiography, Embolization, Recurrence.

INTRODUCTION

Primary tumors of the heart are rare, but more than half of them are myxoma.¹ There is diagnostic difficulty in this most frequent benign tumor of the heart.² Myxoma may resemble many cardiovascular or systemic diseases, and can arise in any of the cardiac chambers, 75% of them occur in the left atrium.³

Before the introduction of angiocardiology in 1951, cardiac myxoma diagnosis was made only at autopsy.⁴ The introduction of echocardiography has provided an important non-invasive means of diagnosis for cardiac myxomas.⁵ Feasibility of open heart surgery made us possible to extirpate all the cases of atrial myxoma. We report five cases of left atrial myxomas

who presented to us with different clinical manifestations.

MATERIALS AND METHODS

We operated five cases of left atrial myxomas since the introduction of open heart surgery in our hospital. Since February 1997, open heart surgery has become a regular service. Among them four cases were diagnosed as left atrial myxomas by transthoracic echocardiography (fig. 1). One was diagnosed as left atrial thrombus with critical mitral stenosis. However, once diagnosis was made, all of them were prepared for surgery and operated at the same during the same admission. Among them 3 myxomas were arising from atrial septum and remaining two from atrial wall.

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Fig. 1

Fig. 1 : Left atrial myxoma (Echocardiography).

Fig. 2

Fig. 2 : Villous and friable myxoma occupying almost entire left atrium (the largest tumor).

RESULTS

In six years 5 cases of left atrial myxomas were admitted in our hospital. Four of them were female. The youngest patient was 32 years old and oldest of 58 years. Clinical manifestations varied in our patients. The first case presented in the form of chest infection with chest pain, fever and cough. Chest X-ray revealed congestion of right lower lobe (unilobar pulmonary venous hypertension). This suspected pulmonary venous hypertension led us to evaluate by echocardiography, which picked up large left atrial myxoma. At operation a huge left atrial myxoma was found to be occupying almost entire atrial cavity. In addition there was a tongue of myxoma extending into right inferior pulmonary vein resulting in pulmonary venous hypertension.

The second case was investigated for rheumatic heart disease with mitral stenosis. Echocardiography reported as critical mitral stenosis (0.8 cm²) with left atrial thrombus. Contradicting to echocardiography reporting, huge left atrial myxoma was excised during surgery. Mitral valve was normal.

The third and the last case had only constitutional symptoms. Both of them had symptoms of palpitation, shortness of breath and fever. Echocardiography was helpful in making the diagnosis of left atrial myxoma. One of them was suspected as a case of bacterial endocarditis.

The fourth case presented with features of embolization. He had right-sided hemiparesis with right upper motor neuron (VII nerve) palsy and motor aphasia. His computerized tomography scan of head revealed temporo-parietal infarction. Echocardiography detected left atrial myxoma. The large myxoma surface was villous and friable (fig. 2), which obviously had potential complications of embolization. Post operatively the patient substantially recovered from motor aphasia. There was wound infection in incisional area, which healed with antibiotics and regular dressing.

In three cases tumors originated from left atrial septum, and in remaining two had left atrial wall origin. Total excision was possible in all cases. The largest tumor size was 10cm x 6cm x 6cm (fig. 2) and the smallest was of 4.5cm x 2.8cm x 2.8cm in dimensions. The smooth myxoma (fig 3) has less potential for embolization. Postoperative ICU (intensive care unit) stay ranged from 2 days to 5 days (mean of 3 days). Total postoperative hospital stay ranged from 5 days to 28 days (mean of 12.6 day). One of them had prolonged stay because of wound infection and another stayed in the hospital for a long period as she had arrhythmia problem (uncontrolled bigeminy). They got better with medicines.

Fig. 3

Fig. 3 : Smooth left atrial myxoma.

Histopathologically all of them were suggestive of benign myxoma with variation in myxoma cell population. In the short-term follow-up no one has evidence of recurrence in repeated echocardiography. Long-term follow-up is needed and we have been following patients from time to time.

DISCUSSION

Atrial myxomas are reported with a frequency between 0.5 and 1 per million of population per year.⁶ Myxoma occurs 75% in the left atrium, 25% in right atrium and rarely in

ventricles.⁷ Recurrence is estimated to be about 5%, but reaches up to 25% in some literature reports.³ All of our cases were of left atrial myxomas. They occur predominantly in women (70%), although currently there is no explanation for this female predominance which could be due to hormonal influence.⁸ Eighty per cent of our cases were females.

The clinical features of these tumors are determined by their location, size and mobility; there are no pathognomonic signs and symptoms that suggest the presence of a myxoma.⁹ Left atrial myxomas become symptomatic when they obstruct the mitral valve, embolize peripherally, or cause systemic effects.¹⁰

In our study one case had mitral valve obstructive feature that was preoperatively diagnosed as mitral valve stenosis with large atrial thrombus. Systemic emboli are the second arm of the classic triad, occurring in 10-45% of myxoma patients.⁹ The myxoma surface was found to be friable or villous in 34% and smooth in 66% in one of the largest series published.⁸ One of our patients had cerebral embolization with neurological deficit who had villous myxoma excised. Another case with villous myxoma did not have features of embolization. Constitutional signs are the third arm of classic triad.¹¹ These signs and symptoms include myalgia, muscle weakness, arthralgia, fever, weight loss, and fatigue. Our two patients were suffering from these non-specific symptoms. One of our cases presented as pulmonary venous hypertension, which has not been mentioned in the studies of atrial myxoma. It is simply due to extension of tumor inside a division of pulmonary vein.

Echocardiography is the diagnostic method of choice.¹² It is accurate, reliable, non-invasive, and it does not entail any risk of tumor fragmentation and embolization (unlike angiography and cardiac catheterization).⁸ Transthoracic echocardiography is less invasive than transesophageal approach with an excellent sensitivity upto to 95%.¹³ We had all the cases diagnosed by transthoracic echocardiography except one in which myxoma was considered as atrial clot. Nevertheless, a high index of suspicion remains the most important element in diagnosing a myxoma.

The myxomas are benign neoplasm of endocardial origin. It can be removed safely by surgery, which remains the treatment of choice.¹⁴ Once the diagnosis is made, urgent excision of the tumor is essential. Embolic complications or sudden death are always possible, even in asymptomatic patients.¹⁵

The reported recurrence rate ranges from 5% to 14% in the literature.¹⁶ This is the reason needing long term follow up of all operated cases. However, the long-term prognosis is excellent.¹⁷

CONCLUSION

These short numbers of cases illustrate that myxoma may cause

a wide range of clinical symptoms. Diagnosis is usually not made only on clinical findings, as there are no specific physical signs or symptoms. Transthoracic echocardiography remains an effective tool and once diagnosis is made surgical extirpation of tumor is not difficult in a center which has facilities for open-heart surgery.

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