Case report

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A rare case of familial recurrent left atrial myxomas

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Summary

A 41-year-old female who had a left atrial myxoma excised was readmitted 21 months later with a recurrence which was also successfully excised. Suspecting a familial trait for atrial myxoma, close questioning of the patient regarding the health of her close relatives revealed that her 21 year daughter had recently become unwell with a cough and shortness of the breath on exertion. Investigations revealed that the daughter also had a left atrial myxoma which was successfully excised. When confronted with a rare recurring atrial myxoma it is essential to consider a familial trait and exclude this condition in close family members.

Case report

A 41-year-old female presented with a 3 month history of feeling unwell, tired, weight loss, persistent cough and shortness of breath on exertion. Blood tests revealed that she was anaemic with a haemoglobin of 9.5g/dl. A 2D ECHO cardiogram revealed the presence of a left atrial myxoma which was excised and a post-op 2D ECHO cardiogram confirmed that the excision of the myxoma was complete. Histological examination revealed that the myxoma was benign MV Grade 11-111. The patient was readmitted 21 months later with the recurrence of her original symptoms. 2D ECHO cardiogram revealed the recurrence of the left atrial myxoma which was excised. Histology confirmed MV Grade11-111 atrial myxoma and the post-op 2D ECHO cardiogram confirmed that the excision was complete. As the recurrence of the atrial myxoma pointed to a possible family trait the patient was closely questioned about her family medical history and although there was no known occurrence of atrial myxoma in the family her 21 year old daughter had been lately suffering from similar symptoms to hers, as of tiredness, weight loss, cough and shortness of breath on exertion. The daughter was admitted to the hospital and blood tests revealed that she had a normochromic anaemia with an Hb of 7.8g/dl and a 2D ECHO cardiogram revealed the presence of a left atrial myxoma. This was excised followed by tricuspid valve repair and histology confirmed benign atrial myxoma. Post-op recovery of both mother and daughter was uneventful.

Discussion

Myxomas are the commonest cardiac tumours and the prevalence at autopsy ranges from 0.001% to 0.3%. In 7%, it has a genetic origin and also arises as a component of a heritable disorder with other clinical manifestations[1].

As was the case with our two patients, in families, atrial myxoma is commonly seen in young women and is transmitted in autosomal dominant way[2]. Familial cardiac myxomas are characterized by a high recurrence rate and involvement in sites other than the left atrium[3]. Rarely it can be a component of Carney complex, characterised by myxomas, hyper pigmented skin, and extra cardiac tumours. As our case report demonstrates, a family history should always be sought in all patients with cardiac myxoma and in particular from those with a recurrence.

Both our patients in this case report presented with non specific symptoms of fatigue, weight loss, cough and shortness of breath on exertion which made the diagnosis difficult initially. It is known that some atrial myxomas produce no symptoms; others produce symptoms depending on their location in the heart and the size of the tumour whereas many only produce non-specific constitutional symptoms such as fever, fatigue, arthralgia, myalgia, and weight loss. Laboratory abnormalities (e.g., anaemia and elevations in the erythrocyte sedimentation rate, C-reactive protein, or globulin level) are present in 35 percent, usually those with systemic symptoms[4].

In both our patients definitive diagnosis was made by two dimensional echocardiography which is the main imaging modality for diagnosis that is simple, quick and non-invasive.

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Both our patients underwent successful excision of the atrial myxomas. An extended follow up of 54 patients who underwent excision of an intracardiac myxoma had revealed that excision of such tumours was curative and long term outcome was excellent[5]. In cases of familial atrial myxoma careful long term follow up of patients and their families is essential in order to deal promptly with recurrences and new occurrences of atrial myxoma.

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