Cardiol Young 2008; 18: 525–527
© Cambridge University Press
ISSN 1047-9511
doi:10.1017/S1047951108002692
First published online 18 August 2008

Brief Report

Familial atrial myxoma

Pin Sun, Zhi-Bin Wang

Department of Cardiac Ultrasound, Affiliated Hospital of Medical College, Qingdao University. Qingdao City, Peoples Republic of China

Abstract The familial variant of cardiac myxoma is known to be an autosomally dominant disease. Early diagnosis, and removal of the tumours, are of great importance. In this regard, echocardiography is considered the simplest and most reliable diagnostic method. We report our experience with echocardiographic diagnosis of a family with atrial myxomas.

Keywords: Hereditary; atrial occupying lesion; echocardiography

Cardiac myxoma is the most common primary tumour of the heart,[1] accounting for about half of all cardiac tumours,[2] and being more frequent in females. Most tumours originate from the region of the oval fossa,[3,4] with about four-fifths occupying the left atrium, and most of the remainder found in the right atrium. Ventricular myxomas are rare.[4,5] The tumours are also known to exist in a familial variant, showing multiple and genetic heterogeneity.[3,6,7] Echocardiography is now well established as the most reliable diagnostic technique, having sensitivity which can reach to 95%.[8] We describe here our experience with echocardiographic diagnosis of 2 familial cases of cardiac myxoma.

Case reports

Our initial patient was a 10-year-old female, hospitalized for chronic coughing of nearly one year, which had become aggravated over the previous 3 days with chest pain and distress. On physical examination, she was found to have a regular cardiac rhythm, and a systolic murmur, graded at 2 from 6, was audible in the tricuspid area. The electrocardiogram revealed sinus tachycardia, while the chest radiographic showed an enlarged heart. Computed tomographic scanning of the lungs suggested pulmonary embolism. Echocardiographic examination revealed a smooth mass, initially thought to lack a pedicle, and measuring 7.2 centimetres by 5 centimetres. The mass filled the right atrium, obstructing the right atrioventricular orifice in diastole. Its upper pole was partially blocking the orifice of the superior caval vein, while inferiorly it encroached on the orifice of the inferior caval vein (Fig. 1). The maximal pulmonary arterial systolic pressure was calculated at 49 mmHg, so that the final echocardiographic diagnosis was a right atrial occupying lesion and moderate pulmonary hypertension. At surgery, the entirety of the right atrium was confirmed to be occupied by the tumour, which measured 8 by 5 centimetres. Surgical exploration revealed that the tumour did possess a pedicle, of 2 centimetres, and it was adherent not only to the base of the atrial septum, but extended into the right ventricle. Despite its soft and fragile texture, the tumour was successfully removed. The pathological findings confirmed the diagnosis of right atrial myxoma, and the patient recovered well.

Our second patient was a 34-year-old female, the mother of our initial patient. She was hospitalized urgently for paroxysmal limitation over 1 day of movement of her right arm and leg. At physical

Correspondence to: Pin Sun, Department of Cardiac Ultrasound, Affiliated Hospital of Medical College, Qingdao University. 16 Jiangsu Road, Qingdao City 266003, P. R. China. Tel: 86 532 8584 8885; Fax: 86 532 8291 1999; E-mail: sunnysunpin@yahoo.com.cn

Accepted for publication 22 June 2008

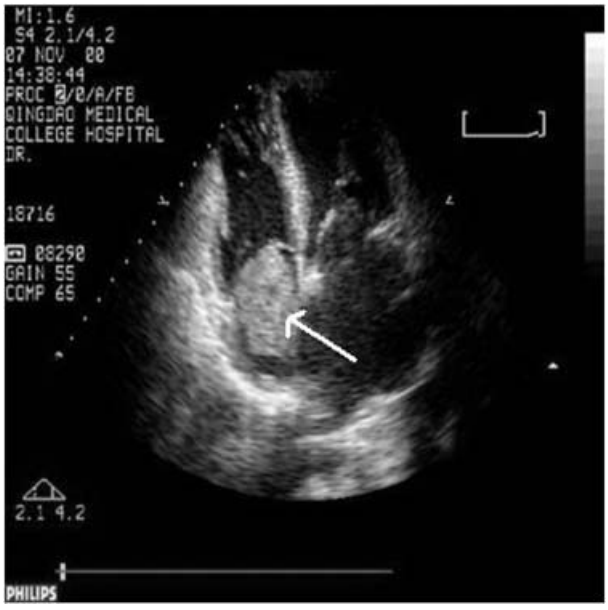

Figure 1.
The right atrial myxoma (arrow) as indetified in our first patient.

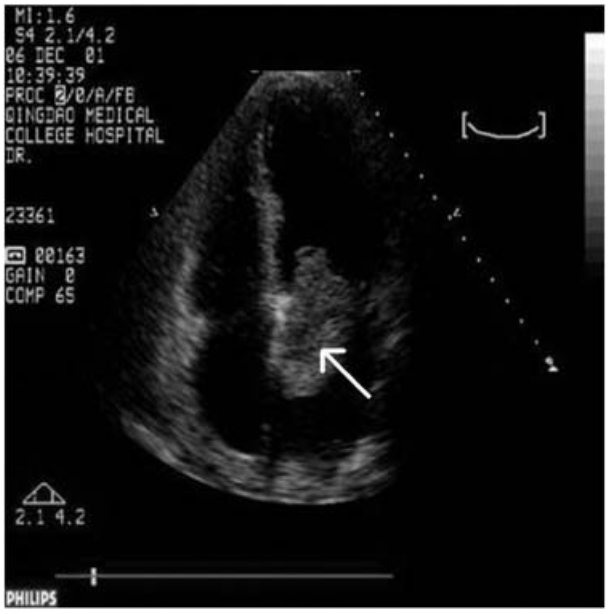

Figure 2.
The left atrial myxoma (arrow) in our second patient, the mother of the index case.

examination, the cardiac rhythm was regular, and a diastolic murmur was heard at the cardiac apex, albeit without any palpable thrill. The muscle power and tone of the right limbs was degraded. The activity of the left limbs was normal, and the bilateral Babinski sign was positive. Inversion of the T waves was seen in leads II, III, and aVF. Injection of contrast showed embolization of the left middle cerebral artery. Echocardiography showed enlargement of the left atrium, which contained a pedicled mass of 2.7 by 5.2 centimetres, adherent to the base of the atrial septum (Fig. 2). With cardiac activity, part of the tumor intruded through the mitral orifice in diastole, returning to the left atrium in systole, and hence produced mechanical obstruction of the mitral valvar orifice. The ultrasonic diagnosis was a left atrial occupying lesion, and the patient was referred for surgical removal. At surgery, a tumour measuring 5 by 4 by 3 centimetres was found in the left atrium, attached by a pedicle of 1 centimetre to the base of the oval foramen. Pathological studies showed it to be a myxoma. The patient recovered well, and the power of the right limbs was restored at the time of discharge.

Discussion

All familial cardiac myxomas have been shown to be associated with chromosomal abnormalities, the haploid cells having abnormal contents of deoxyribonucleic acid, with some showing heightened proliferative activity.[9] Chromosomes 2 and 12 have been involved, and the tumour has been shown to be inherited in autosomal dominant fashion.[10] Significantly, the familial form of cardiac myxoma is now considered potentially malignant.

Echocardiography is well recognized as being capable of showing the size of such tumours, their different positions during the cardiac cycle, and the degree of valvar stenosis and counter-flow that they can produce. Hence, echocardiography is the simplest, as well as the most reliable, means of early detection. Such early detection is important, since fragments of the tumours can embolise. Cerebral embolism is particularly dangerous. Once diagnosed, surgery is needed as soon as possible. Not only should the myxoma be thoroughly excised, but also the normal endocardium and cardiac muscle at the point of attachment of its pedicle. The familial myxoma is also known frequently to be multiple, and recurrences are well recognized. Thus, periodic follow-up is necessary. Should recurrences occur, the new tumours should again rapidly be removed.

References

1. Gavrielatos G, Letsas KP, Pappas LK, Dedeilias P, Sioras E, Kardaras F. Large left atrial myxoma presented as fever of unknown origin: a challenging diagnosis and a review of the literature. Cardiovasc Pathol 2007; 16: 365–367.
2. Meng Q, Lai H, Lima J, Tong W, Quian Y, Lai S. Echocardiographic and pathologic characteristics of primary cardiac tumors: a study of 149 cases. Int J Cardiol 2002; 84: 69–75.

https://doi.org/10.1017/S1047951108002692 Published online by Cambridge University Press

3. Rogov KA, Sheremet'eva GF, Nechaenko MA. Family myxoma of the heart with metachronic multiple growth. Arkh Patol 2004; 66: 41–44.
4. Alter P, Grimm W, Rominger MB, et al. Right ventricular cardiac myxoma. Diagnostic usefulness of cardiac magnetic resonance imaging. Herz 2005; 30: 663–667.
5. Grebenc ML, Rosado-de-Christenson ML, Green CE, Burke AP, Galvin JR. Cardiac myxoma: imaging features in 83 patients. Radiographics 2002; 22: 673–689.
6. Casey M, Mah C, Merliss AD, et al. Identification of a novel genetic locus for familial cardiac myxomas and Carney complex. Circulation 1998; 98: 2560–2566.
7. Basson CT, MacRae CA, Korf B, Merliss A. Genetic heterogeneity of familial atrial myxoma syndromes (Carney complex). Am J Cardiol 1997; 79: 994–995.
8. Percell RL, Henning RJ, Siddique Patel M. Atrial myxoma: case report and a review of the literature. Heart Dis 2003; 5: 224–230.
9. Kirklin JW. The future of cardiac surgery. Heart Dis Stroke 1993; 2: 361–364.
10. Stratakis CA. Mutations of the gene encoding the protein kinase A type I-alpha regulatory subunit (PRKAR1A) in patients with the "complex of spotty skin pigmentation, myxomas, endocrine overactivity, and schwannomas" (Carney complex). Ann N Y Acad Sci 2002; 968: 3–21.