A large atrial myxoma in a young woman

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The incidence of primary cardiac tumours in autopsy series range from 0.02 to 2.8%—much more rare than metastatic cardiac tumours [1]. They are predominantly benign (75%). Myxomas are the most common type of primary cardiac tumour [2], comprising 30–50% of most pathological series [2] and increasingly gaining significance among the elderly [3]. Clinical presentations and symptoms are comparable with those arising from other cardiovascular and systemic conditions, and include variable cardiac murmur, atrial fibrillation, uneasiness, fatigability, dyspnoea, atypical chest pain, dizziness, systemic and pulmonary embolism, heart failure and sudden death [4, 5].

We present this case, as a reminder of the fact that atrial myxomas can appear in young patients too, and that despite their low incidence should nonetheless be considered for diagnosis by emergency departments.

Case report

A 32-year-old healthy woman repeatedly attended an emergency department, with a 1 month history of increasing dyspnoea and fatigability. She had previously been labelled as suffering “panic attacks”. The patient’s previous medical history was not relevant. Physical examination revealed a blood pressure of 90/60, heart rate of 120 per minute, respiratory-rate of 26 per minute, and a temperature of 36.5°C. During chest auscultation, we noticed a soft diastolic murmur in the mitral area. There were no signs of respiratory insufficiency in arterial blood gases. The white cell count was 19.6 per microliter, no other cause of leucocytosis besides myxoma was found. Chest X-ray was normal and ECG revealed sinus tachycardia without ischemia, with a rate of 130 beats/min and criteria of right atrial enlargement (P wave amplitude >2.5 mm in lead II and >1.5 mm in lead VI). Chest computed tomography was requested to exclude pulmonary embolism. The images showed a solid mass of about 5 cm in the right atrium (Fig. 1a). As a result of the chest computed tomography, transthoracic echocardiograms were requested to verify the diagnosis of myxoma. The images confirmed a large mass attached to the posterior wall of the right atrium (Fig. 1b). Colour Doppler revealed a patent foramen ovale with a turbulent speed from the right atrium to the left atrium and a moderate tricuspid regurgitation. The patient was diagnosed with right atrial myxoma and transferred to the cardiac surgery department, where a surgical resection of the mass was performed and the patent foramen ovale closed. The postoperative course was unremarkable and she was discharged on postoperative day 5. At her last follow-up visit she was still well. Histological examination of the mass showed abundant hyaline and myxoid stroma containing small groups of tumour cells, compatible with the diagnosis of atrial myxoma.

Discussion

Atrial myxomas are the most common primary heart tumours. Due to non-specific symptoms, early diagnosis may be a challenge [6]. Fifty percent of all primary cardiac tumours are myxomas. Myxomas occur most often in
patients aged between 30 and 70, in women, and in families with a tendency of developing myxomas [7].

About 85% of myxomas occur in the left atrium, 10% in the right atrium, and 5% in the ventricles [8]. Five percent of patients present more than one myxoma or a polycentric myxoma. A common site of attachment for atrial myxomas is the fossa ovalis region of the septum; another is the posterior atrial wall [9].

Depending on the size of the mass and the degree of obstruction, symptoms range from non-specific and constitutional to sudden cardiac death. In about 20% of cases, myxoma may be asymptomatic [6]. More frequently, vague constitutional symptoms such as fever and weight loss are present [10]. There is evidence that the tumours secrete interleukin-6, messenger ribonucleic acid and proteins [11]. Tumour production of various cytokines and growth factors may contribute to clinical and laboratory abnormalities, such as leucocytosis [12]. Anaemia has also been reported, presumably from wall-valve movement of the mass into the valve orifice with consequent erythrocyte destruction [10]. Syncope is an extremely rare finding, and is likely due to intermittent complete occlusion of the tricuspid valve [10]. Sudden death may occur in 15% of the patients with atrial myxoma. Death is typically caused by coronary or systemic embolization or by obstruction of blood flow at the mitral or tricuspid valve [6]. Laboratory investigation result may therefore sometimes mimic those for rheumatic or autoimmune disease [13].

Without doubt, transthoracic and transesophageal echocardiography are the methods of choice [14] in the detection of atrial myxomas. More recently, however magnetic resonance imaging has become a valuable option. These techniques depict size, localization, point of attachment, and cardiac myxoma characteristics even more clearly than computed tomography [15]. In addition to systematic echocardiography, this method may offer a helpful tool for surgical planning.

In general, resection of the tumour is sufficient to ensure patient recovery; however, if there is total or partial occlusion of the coronary artery, coronary angioplasty or coronary artery bypass surgery may be necessary [7].

Although atrial myxomas are very rare, their presence should be considered, particularly in young patients with no cardiac risk factors. Our case demonstrates the need for thorough clinical examinations and appropriate investigation selection, and that atrial myxomas are strongly considered for patients with recurrent symptoms.

References