Right atrial myxoma associated with an atrial septal defect by real-time three-dimensional echocardiography

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A 40-year-old woman presented with a 1 month history of shortness of breath and easy fatigability. Two-dimensional echocardiography and real-time three-dimensional echocardiography (RT3DE) showed a large right atrial mass protruding into the right ventricle and an atrial septal defect (ASD). Successful excision of the mass, which proved to be a myxoma, and closure of the ASD completely relieved her symptoms. To our knowledge, this is the first report of RT3DE in a patient with right atrial myxoma associated with an ASD.

KEYWORDS
Right atrial myxoma; Atrial septal defect; Real-time three-dimensional echocardiography

Introduction
Cardiac myxoma is the most frequent primary tumour of the heart. It accounts for 50% of the benign tumours of the heart. They may occur in any chamber, but usually originate in the left atrium (75%), the next most frequent site being the right atrium (10–20%). Myxomas are rarely associated with congenital cardiac anomalies. We report a rare combination of a right atrial myxoma with an ostium secundum atrial septal defect (ASD), diagnosed by two-dimensional echocardiography (2DE) and real-time three-dimensional echocardiography (RT3DE). To our knowledge, this is the first report of RT3DE in a patient with a right atrial myxoma associated with an ASD.

Case report
A 40-year-old female patient presented with a 1 month history of exertional dyspnoea (NYHA class II) accompanied by cough and palpitations with no orthopnoea or chest pain. Clinical examination revealed a blood pressure of 114/75 mmHg, a regular pulse of 70 bpm, a respiratory rate of 24 bpm, and she was afebrile. The jugular venous pressure was normal, with a systolic murmur (grade 2/6) at the left lower sternal border and her chest was clear. A complete blood count and serum biochemistry profile were normal. D-dimer was negative.

Arterial blood gases on room air showed severe hypoxia with a PO2 of 48 mmHg, PCO2 of 26 mmHg, pH of 7.4, and an oxygen saturation of 80%. Chest X-ray revealed mild cardiomegaly mainly on the right side. An ECG showed sinus tachycardia. Transthoracic 2DE showed dilated right ventricle (43 mm) and right atrium (52 mm) with trivial tricuspid regurgitation and right ventricular systolic pressure: 25 mmHg. A large mobile lobulated right atrial mass measuring 7.0 × 5.0 cm was seen protruding into the right ventricle and partially obstructing the tricuspid valve (mean diastolic gradient: 3 mmHg). It also showed an interatrial septal aneurysm with a tiny colour flow from right to left (Qp/Qs: 1.2) suspecting an ASD (Figure 1). Left-sided chambers were normal in size and function (left ventricular ejection fraction: 60%).

Real-time three-dimensional echocardiography was performed by Philips Sonos 7500 ultrasound machine using an X4 matrix transducer. The 3D data were stored digitally and analysed by the Q-lab software (see Supplementary data online, Video 1). Real-time three-dimensional echocardiography demonstrated a large mobile multilobular mass measuring 7.0 × 3.7 cm. Enface view from left atrial side showed a large ostium secundum ASD measuring 2.3 × 4.0 cm with a slight protrusion of the mass inside the defect. The mass stalk was seen attached to the atrial septum at the proximal edge of the ASD (Figure 2). The myxoma volume (76.9 ml) was calculated using the advanced quantification software for left ventricular volume.

Urgent surgery was undertaken on the following day. Intra-operative transoesophageal 2DE (TEE) confirmed the
findings of the transthoracic 2DE and revealed a partial protrusion of the mass into left atrium through the ASD (Figure 3; see Supplementary data online, Video 2). At surgery, a large lobulated tumour almost filling the entire right atrium measuring 7.2 × 4.1 cm was encountered. The stalk appeared to originate from the proximal atrial edge of the ASD which measured 2.2 × 3.8 cm. The left atrium was inspected through the defect, and there was no evidence of other masses. The tumour with its stalk was carefully excised (Figure 4) and the ASD was closed with a bovine pericardial patch. Postoperative TEE showed no residual masses and no leakage across the atrial septum. Histopathologic examination revealed myxomatous tissue with acid-mucopolysaccharide matrix and polygonal cells. Repeated transthoracic 2DE before discharge showed reduction of the gradient across the tricuspid valve to 1 mmHg and normalization of right ventricular size with no residual masses. The patient had an uneventful postoperative course and her arterial blood gases showed normal oxygenation. She was discharged home on the forth postoperative day.

**Discussion**

Right-sided myxomas are rare and usually present with signs and symptoms of right heart failure and pulmonary embolism. They are typically broad based on their attachment and are more likely to be calcified compared with left-sided lesions. Ha et al. identified two distinct types of myxoma by echocardiography: (i) round type characterized by a solid spherical shape with non-mobile surface (52%) and (ii) polypoid type characterized by soft and irregular shape with mobile surface (48%). Our patient was of the latter type (Figure 4). Surgical resection is the only effective therapeutic option for patients with cardiac myxoma and should not be delayed since death from obstruction to flow or embolization may occur in 8% of patients awaiting operation. The co-association of a myxoma with an ASD resulting in symptomatic right to left shunting in adults is extremely rare. Only a few cases were reported in the literature. To the best of our knowledge, this is the first case report utilized RT3DE for the diagnosis of such combination.

Our patient was hypoxic on presentation and we believe that the partial obstruction of the tricuspid valve due to the right atrial tumour and subsequent elevation of the right atrial pressure had resulted in right-to-left shunt through the ASD. Recurrent pulmonary embolism secondary to right atrial myxoma has been described previously. In our patient, there was no evidence of pulmonary embolism pre-operatively.
Echocardiography is the most useful test for diagnosing myxomas. Transoesophageal 2DE is extremely important and valuable in providing information about the myxoma and its anatomical relationship.\(^5\) Real-time three-dimensional echocardiography proved to be helpful in identifying the site of attachment of the tumour and clearly defined the margins of the ASD. It provides valuable information on the extent of the intracardiac tumours, accurate measurements, their influence on valvular function, and the coexistence of other congenital anomalies.\(^6,7\) By RT3DE, the ability to electronically dissect cardiac structures simulating surgical view will encourage the use of RT3DE pre-operatively to facilitate surgical planning.

Supplementary data

Supplementary data are available at *European Journal of Echocardiography* online.

References