Asymptomatic giant prolapsing right atrial myxoma: comparison of transthoracic and transesophageal echocardiography in pre-operative evaluation

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Cardiac myxomas are uncommon with a reported incidence of 1 in 10,000 autopsies and there is still a controversy about their pathogenesis [1]. Right myxomas are three to four times less frequent than those located in the left atrium [2]. Formerly the diagnosis was only possible at autopsy or identically on angiography; with the application of transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) this has improved markedly and more than 90 % of atrial myxomas are adequately visualised [3]. Surgical removal is recommended due to their embolic tendency and haemodynamic consequences such as valvular obstruction or destruction [4]. Echocardiography plays an important role as a diagnostic aid not only in demonstrating the localisation, origin, calcification, peduncle attachment, mobility and dimensions of the tumour but also in recognising the degree of valve destruction and its haemodynamic consequences which affect the decision for a valve replacement [4].

Case report
We present and compare the TTE and TEE two-dimensional features of an asymptomatic patient with a prolapsing giant right atrial myxoma referred to our echocardiography laboratory.

Physical examination revealed a blood pressure of 130/70 mmHg, a regular and rhythmic heart rate of 72 beats per minute with normal first and second heart sounds. There was no precordial lift or thrill but a grade 2/6 holosystolic murmur increasing with inspiration, best heard at lower left parasternal border and a diastolic plop was recognised.

Two-dimensional TTE revealed dilatation predominantly of the right and left atrium. Left ventricular size and function were normal. A large (approximately 9.3 cm × 7.1 cm) lobulated and pedunculated mass prolapsing through the tricuspid valve in diastole and containing calcifications was noted; it almost totally occupied the right atrium.

However the attachment, the precise site of origin and the possibility of another tumour focus could not be evaluated accurately by TTE. The colour-flow and Doppler study showed mild to moderate tricuspid regurgitation.

TEE not only yielded a more measurement of the dimensions of the tumour (8.6 cm × 6.5 cm × 6.2 cm) as compared with postoperative pathological findings, it also demonstrated clearly existing calcifications, the attachment and precise site of tumour pedunculation: in this case it was the right atrium post-terior wall; there were no significant haemodynamic changes, the possibility of other tumour foci was eliminated. TEE gave correct anatomical and surgical data pre-operatively.

TTE and TEE were performed by a Hewlett-Packard Sonos 1500 ultrasound system (Hewlett-Packard Company, Andover, MA, USA) with a 2–2.5 MHz and omniplane 5.0/3.7 MHz transducer (Model 21364 A), respectively (Figures 1–3).

Because the mass was large and lobulated, urgent surgical intervention was performed. At surgery a large 8.5 cm × 6.5 cm × 6 cm calcified and pedunculated tumour, attached to the posterior wall of the right atrium by a small peduncle, was noted and the pathological findings confirmed the diagnosis as a myxoma.

Discussion
Myxomas are benign and the most frequent primary tumours, accounting for nearly 50 % of all primary masses of the heart. These tumours are located in the left atrium in 75 % and in the right atrium in 17 % to 25 % [1, 2]. Left and right ventricular myxomas are rare constituting 2.5 % to 4 % of all myxomas [5]. Right atrial myxomas show three types of clinical manifestations:

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Figure 1. Transthoracic parasternal short-axis view showing a giant atrial myxoma in the almost totally occupied right atrium; RA, right atrium; RV, right ventricle; LA, left atrium; AOV, aortic valve
1. Systemic reactions, due to the products of degeneration of the tumour leading to anaemia, fever, loss of weight, leucocytosis, elevated sedimentation rate and hyperglobulinaemia, especially in the early stages of the disease before the tumour is of sufficient size to cause mechanical obstruction [3, 6]. Also systemic involvement including the syndrome of right atrial myxoma with spotty skin pigmentation and acromegaly [7], right atrial myxomas with atypical features of the myxoma syndrome [8], lentiginosis and right atrial myxoma [9] and nephrotic-range proteinuria associated with right atrial myxoma [10] were described.

2. Embolic phenomena occur if fragmentation of the tumour appears or thrombi formed on the surface of the tumour may cause micro- or macroembolism [6]. Pulmonary emboli, as described by Heath et al. [11] may occur. Systemic embolisms are less common with right atrial myxomas and may be due to paradox emboli through a patent foramen ovale.

3. Haemodynamic changes due to mechanical obstruction, involvement and destruction of the valves can be observed [3]. It is believed that the growth is slow and may remain asymptomatic for a long time [12].

TEE is the standard instrument for diagnosing cardiac tumours [3, 13]. Recently TEE became a powerful diagnostic tool and its application rapidly increased. Advantages of TEE in the diagnosis and collection of surgical data were reported previously [13, 14].

Lyons et al. [15] reported the successful use and important role of TEE performed intra-operatively in the management of an asymptomatic giant right atrial myxoma.

Rey et al. [16] presented the first prolapsing right atrial myxoma evaluated by TEE and indicated the superiority of TEE in demonstrating the correct tumour size, morphologic details, prolapsing behaviour and the attachment. In this study, we emphasize the adequate visualisation of a right atrial myxoma by TEE and the importance of this method for current clinical practice in an echo laboratory.

We conclude that the pre-operative evaluation of right atrial myxoma by TEE is superior in clarifying the attachment site of the tumour, pedunculation and dimension and checking possible involvement of other chambers or structures that might modify the operative procedure. Furthermore, the better imaging quality of TEE compared with TTE, especially in patients with chest deformity or poor echocardiographic window, should enhance all these elements of preoperative assessment. We suggested that patients with cardiac tumours diagnosed by TTE must be evaluated further by TEE pre-operatively, or as previously reported intra-operatively [15] to obtain more information.

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