Left atrial myxoma attached to the anterior mitral leaflet with symptoms suggestive of infective endocarditis

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The case of a 55-year-old woman is presented, whose clinical signs were initially suggestive of infective endocarditis. Transthoracic echocardiography (TTE) provided the diagnosis of a large left atrial myxoma attached to the anterior mitral leaflet. Perioperative transesophageal echocardiography (TEE) confirmed preoperative findings and assisted the surgical team in the assessment of tumour size, area of attachment, and mobility. Following tumour resection, TEE demonstrated residual moderate mitral valve regurgitation, which resulted in a change of surgical strategy. This report reinforces the importance of intraoperative TEE to facilitate and optimize surgical and anaesthesiological management of patients presenting with non-specific cardiorespiratory symptoms.

Keywords
Echocardiography • Cardiac tumour • Myxoma • Mitral valve

Case report

A 55-year-old woman without significant previous illnesses presented to her family practitioner with a temperature of 37.5°C, recurrent cough, progressive exertional dyspnoea, and declining functional capacity corresponding to NYHA Class II. A new diastolic heart murmur was noted with maximum over the cardiac apex. On the basis of these symptoms, which were associated with an increased white cell count and elevated C-reactive protein levels, a possible diagnosis of infective endocarditis was considered. The patient underwent transthoracic echocardiography (TTE), but no vegetations were detected. In contrast, a mobile solid left atrial (LA) mass was found, which was attached to the anterior mitral leaflet (AML). Few days later cardiac surgery was performed. After induction of anaesthesia, transesophageal echocardiography (TEE) confirmed and delineated the findings more precisely. A large, spherical, hyperechogenic, pedunculated tumour with central echolucenty was found attached to the base of the AML between segments A1 and A2 (Supplementary data online, Video S1 and Video S2). During diastole, the mass prolapsed into the left ventricle, obstructing a major portion of left ventricular inflow (Figure 1). In addition, a moderate mitral stenosis was present (functional valve orifice area 1.05 cm²; mean transvalvular gradient 10 mmHg). LA and left ventricular dimensions were found normal, as well as ejection fraction (60%). Surgery was performed through median sternotomy, using bicaudal cannulation, moderate hypothermic cardiopulmonary bypass (CPB), and cardioplegic arrest. A transapical approach to the LA revealed a pedunculated tumour sized 37 x 30 x 27 mm with smooth surface (Figure 2), which was attached to the AML and the roof of the left atrium. Post-operative histological analysis revealed a mesenchymal tumour with myxoid matrix, but without atypical cells or necrosis.

Complete resection of the tumour left a defect in the anterior part of the mitral leaflet (A1). This necessitated reconstruction of the leaflet with a patch of xenopericardium, and of the mitral annulus, which was performed using a Physio Annuloplasty Ring (size 26 mm, Edwards Life Sciences®). After deairing, closure of the heart and resumption of partial CPB with pulsatile systolic arterial pressure at 90 mmHg, intraoperative TEE showed a still moderate-degree mitral regurgitation. On the basis of this information, the surgeon reverted to mitral valve replacement with a mechanical prosthesis (size 27 mm, ATS Medical Inc.®).

Thereafter, weaning from CPB was uneventful. TEE re-evaluation showed no residual masses, correct function of the prosthetic valve without paravalvular leak, no regional wall motion abnormality and complete resolution of pulmonary hypertension.

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Discussion
Clinical distinction between endocarditis and cardiac myxoma is not always straightforward. Because of the variety, non-specificity, and partial concordance of symptoms, it is usually not possible to make the differential diagnosis based on medical history and physical examination alone. Also, laboratory chemistry and microbiological results may sometimes be misleading. For example, blood cultures, which would establish endocarditis, are not always positive in all patients with infective endocarditis.1 On the contrary, myxomas can clinically present with signs of systemic inflammation (raised erythrocyte sedimentation rates and C-reactive protein levels) directing the diagnosis away from cardiac origin.2

TTE and TEE are simple, efficient, generally available, and evidence-based methods to resolve this differential. Typical echocardiographic findings constitute one major criterion to confirm the diagnosis of infective endocarditis, according to the Duke Criteria for Infective Endocarditis.3 Therefore, echocardiography represents Class I indications with evidence level B or C for the detection of endocarditic valvular vegetations.4 Likewise, TTE represents the most reliable diagnostic tool for cardiac myxoma.5 The technique, however, has limitations: in addition to a suboptimal transthoracic window, small tumours and tumours far removed from the transducer are poorly visualized. TEE has been demonstrated to be superior to TTE for diagnosis and characterization of cardiac masses, particularly within the LA.6

Although a precise diagnosis is possible only by means of histological assessment, echocardiography significantly narrows down the differential diagnosis. It also helps to devise management strategies, particularly since tumour types differ in their echogenic appearances. Most of myxomas are pedunculated and show a central echolucent area, corresponding to either haemorrhage or tumour cell necrosis. These findings distinguish myxomas from large vegetations, thrombus, or non-myxomatous tumours like papillary fibroelastoma or rhabdomyosarcoma.7

In addition to the information derived from TTE, the significance of perioperative TEE for detailed diagnosis and therapeutic guidance is underscored by (i) confirmation of preoperative findings with precise localization of the myxomas attachment site, which helped in surgical planning; (ii) documentation of complete tumour removal; (iii) evaluation of mitral valve repair; and (iv) diagnosis and grading of post-repair mitral valve regurgitation, which led to an intraoperative change in surgical strategy, i.e. prosthetic mitral valve replacement. Furthermore, as is routine in mitral valve surgery, TEE provided to the perioperative team the functional assessment of the prosthetic mitral valve, rule-out of a para-valvular leak prior to the termination of CPB, post-operative detection of new regional wall motion abnormalities, presence of blood flow signal in the circumflex artery, assessment of left ventricular function and guidance for appropriate fluid and inotrope management.

In summary, cardiac myxomas may present with non-specific symptoms, which are also associated with other cardiac and inflammatory disease including infective endocarditis. Cardiac myxoma should therefore be considered in the differential diagnosis in patients with progressive non-specific cardiorespiratory symptoms. Echocardiography is the most important tool to reach the definitive diagnosis rapidly and non-invasively. In addition, perioperative TEE is indispensable in planning the surgical procedure, in real-time assessment of its results and for medical management in the early post-operative period.

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

Conflict of interest: none declared.

References

