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CASE REPORT

A large tumour of the left atrium – A 10-year follow-up

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Abstract Benign myxomas are the most common primary tumors in the left atrium of the heart, and they usually require urgent operative management. However, with the constant aging of the population and treatment of patients with severe concomitant diseases, an interesting question is the efficacy of the conservative approach. We present a case of a 75-year-old woman who was previously diagnosed with a left atrial myxoma, underwent an operation to resect it and then developed a recurrent tumor at the site of resection, which was found by control echocardiography 1.5 years later. She has been observed for over 10 years, as she repeatedly refused reoperation. In the setting of this example of a relatively favorable clinical course of left atrium myxoma, we discuss the current knowledge about the natural history of these tumors and the role of echocardiography in predicting their growth and complications.

1. Introduction

Among primary tumors found in the left atrium, benign myxomas constitute a vast majority (80-90%).[1] Over 20% of them are asymptomatic.[2] In other cases, the following three main presentation types are observed: embolic complications, obstruction to the intracardiac blood flow and systemic symptoms. Considering the risk of life-threatening complications, the recommended therapeutic method is tumor resection. However, in the literature, there are some infrequent situations that require a more conservative approach. This paper describes a case of a 75-year-old woman with recurrence of a left atrial myxoma, diagnosed over 10 years ago who refused to consent to undergo another surgery.

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2. Case study

A 75-year-old female patient with arterial hypertension and hypothyroidism treated with supplementation and with a history of anxiety neurosis and insomnia was first referred to transthoracic echocardiography 13 years ago because of paroxysmal palpitations and a slight reduction in her exercise tolerance. The examination revealed a tumor of the left atrium that was associated with the interatrial septum; it had a size of 33 × 57 mm in the four-chamber (4C) view, and no additional abnormalities were found in the heart. The tumor was surgically excised. Pathological examination confirmed the diagnosis of myxoma. The patient had no significant clinical improvement following the procedure, which was attributed to her symptoms being mostly psychogenic in character. Control echography was performed after the surgery. Another investigation, performed 1.5 years after the procedure, revealed a well-circumscribed lesion that demonstrated characteristic signs of a myxoma. It was located at the resection site and was most likely a recurrence with a size of 22 × 37 mm. It lacked a visible pedicle or calcification, but it had a smooth surface and did not disturb the mitral flow. Since that time, the patient has remained under the supervision of the Cardiology Clinic (over 10 years now). She has been repeatedly informed about the need for surgical removal and has refused to give informed consent to undergo a repeat cardiologic surgery. Multiple echocardiographic investigations revealed a progression in the tumor size, which was initially rapid (up to over 10 mm/year in both dimensions) and then slowed significantly. In the 10 years of follow-up, the tumor has reached a size of 42 × 62 mm in 4C projection, and the patient has simultaneous echocardiographic features of pulmonary hypertension (diastolic movement of the interventricular septum towards the left ventricle and a transvalvular retrogradient through the tricuspid valve of approx. 70 mmHg compared with the retrogradient first defined of 35 mmHg 8 years ago). She has also developed moderate/severe tricuspid and pulmonary regurgitation. During that time, slowly increasing signs of heart failure with preserved ejection fraction were observed, which were mainly in form of paroxysmal nocturnal dyspnea. However, those symptoms caused only a slight impairment of the patient's everyday function. The patient required hospitalization once because of exacerbation of circulatory failure, presenting as pulmonary edema, which was probably associated with intensive initial tumor growth. Despite the signs of right ventricular insufficiency demonstrated by echocardiography, a clear clinical manifestation has been absent. The patient has not experienced any registered episodes of atrial fibrillation, and her electrocardiography (ECG) appeared normal. Only an increased erythrocyte sedimentation rate (ESR) was remarkable in the additional laboratory tests. In course of the 10-year follow-up, the patient has irregularly been using drugs, beta-blockers and diuretics at low doses, and she has also been non-compliant in terms of antithrombotic treatment. For the majority of the time, she has failed to achieve therapeutic values of the international normalized ratio (INR).

3. Discussion

The case presented above is another example of a natural course of cardiac myxoma. In rare cases, it appears to be relatively benign and demonstrates no tendency for clinically overt complications (including embolic events).[3]

As is generally known, the diagnosis of left atrial myxoma constitutes an urgent indication for resection. The periprocedural mortality rate is below 3%,[4] and surgically treated myxomas are associated with a favorable prognosis, in both the short- and long-term. Rarely is myxoma excision surgery associated with necessary implantation of an artificial valve or pacemaker.[2] Recurrence following excision of sporadic tumors is observed in some percentage of patients; it most commonly occurs during the first 4 years after the initial surgery. During that time, annual echocardiographic monitoring is recommended to detect possible tumor re-growth.[2,5]

A diagnosis of heart myxoma is an indication for coronarography before considering a cardiologic procedure. In case of a co-existing advanced coronary disease, the decision concerning the surgical procedure is additionally justified by the possibility of simultaneous coronary-aortal bypass grafting.[6,7] In the case of small, asymptomatic tumors and the absence of other indications for surgery, the decision may be less obvious than expected.[6]

As the majority of myxomas are promptly excised after making the diagnosis, little is known about the natural development of the condition. There are scarce data regarding growth rate, which are often contradictory, that indicate the growth rate ranges from an absolute absence of growth to several millimeters per month.[8] Recurrent myxomas are considered to have tendency for a faster growth rate compared to primary ones.[3] As indicated by this case, the myxoma growth rate is also variable in a single patient. Initially, a faster rate was observed (up to over 10 millimeters/year), which was followed by a later period during which the growth was several times slower. The calculated mean myxoma growth increased 2 years after the diagnosis of recurrence (and for the subsequent 8 years), and it was only approx. 0.15 millimeters/month. The myxoma size ranges from several millimeters to as much as 15 cm in diameter. Large tumors (diameter of over 5 cm) are considered to be more commonly associated with cardiologic symptoms. In the presented case, an initially faster tumor growth was correlated with the development of symptoms related to circulatory system decompensation (although the tumor was a relatively small size). Therefore, both the size of myxoma[3] and current growth rate play an important role in the development of clinical manifestations in response to a tumor in the left atrium.

The question of whether it is possible to predict the behavior of a myxoma of the left atrium based on its features in follow-up echocardiographic investigations is an important one. In their study, Pinede, Duhaut and Loire noted that when a myxoma is round and has a smooth surface, the incidence of embolic events is over twice less frequent than when a tumor is described as villous, polypous and fragile.[1,9] Similarly, for calcified tumors, the risk of a cardiogenic embolism may be lower. Kay and Chow described a case of an 85-year-old patient with a left atrial

myxoma who refused surgical excision of the tumor, as with our patient. In their study, the patient was followed for another 15 years with a stable tumor.[10]

A pedicle and the tumor mobility play important roles in the clinical manifestations. The effect of a so called "wrecking ball" is known, which involves injury of the mitral valve resulting from repeated contact of the tumor with cusps.[1] Moreover, tumor prolapse through the mitral ostium hinders blood flow into the left ventricle, causing clinical signs of a mitral defect (stenosis or incompetence). It is believed that mobility of the myxoma may, in those cases, lead to the development of paroxysmal symptoms, which sometimes depend on posture.[1] Conversely, atrial fibrillation is not often observed in patients with atrial myxoma because atrial enlargement is not common in those patients. Atrial enlargement is described in approx. 30% of patients,[7,9] and atrial fibrillation in 20%.[7] In the discussed patient, the left atrium enlarged from 36 to 41 millimeters during the period of 10 years and was almost completely filled by tumor (Figs. 1—4).

As it is generally known, many tumors occur more frequently in aged organisms, but myxomas are believed to be rather rare in the elderly.[11] Although senescent cells are considered more prone to oncogenesis, the senescent immune system may contribute to the phenomenon of slower growth of some tumors and smaller aptitude for metastasis of malignant cells in that population. However, tumor seeding is much more complicated, depending on the tumor type as well as host humoral and local factors.[12,13] Whether any of these observations can also be accurate for the growth of myxomas as benign neoplasms in aged patients remains a field to explore.

In the context of common applications of surgical treatment, there are no currently detailed guidelines for conservative therapy. Peripheral embolism with tumor fragments or myxoma cell-containing clots is a complication that occurs in half of patients with a left atrial myxoma. The complication is mainly associated with the central nervous system and limbs, but myxoma sometimes also causes embolism in the coronary circulation, abdominal organs, or skin. It is evident that with constant secretion of proinflammatory factors, those tumors induce a chronic inflammatory response, favoring coagulation and the formation of clots on the tumor surface. Chronic antithrombotic therapy is a treatment option that can reduce the risk of embolism. However, difficulties associated with determination of an appropriate dose of a Coumadin derivative, and with monitoring of the therapy in elderly and sometimes non-cooperative patients, often make proper management of this therapeutic approach impossible. The emergence of novel oral anticoagulants is

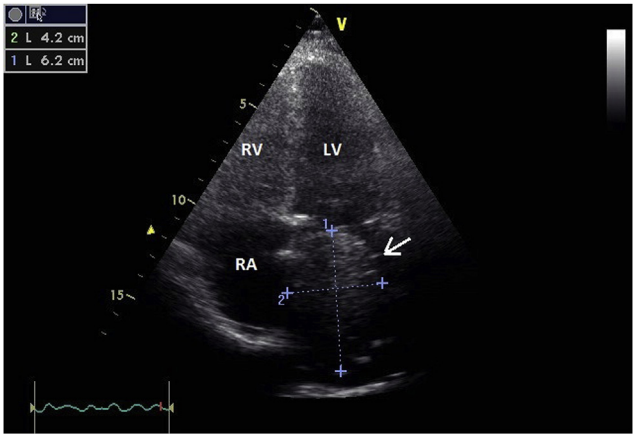

Fig. 1 TTE 2D. Normal 4-chamber apical view.

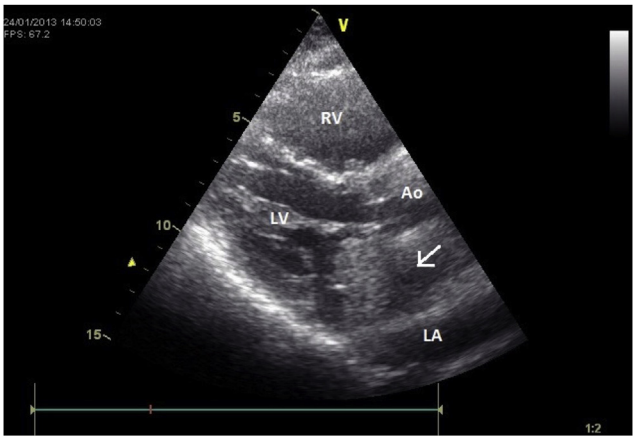

Fig. 2 Normal long axis view.

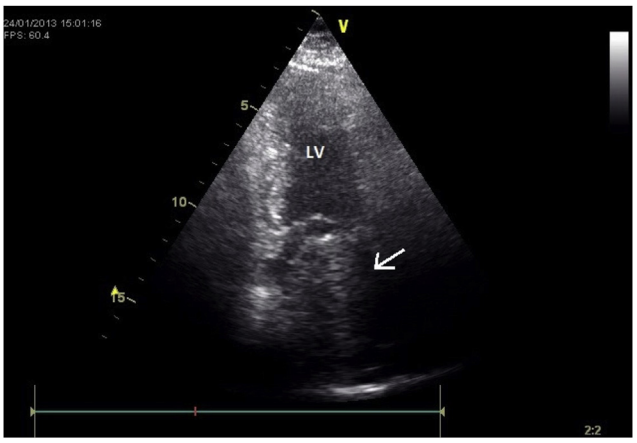

Fig. 3 Normal 2-chamber apical view.

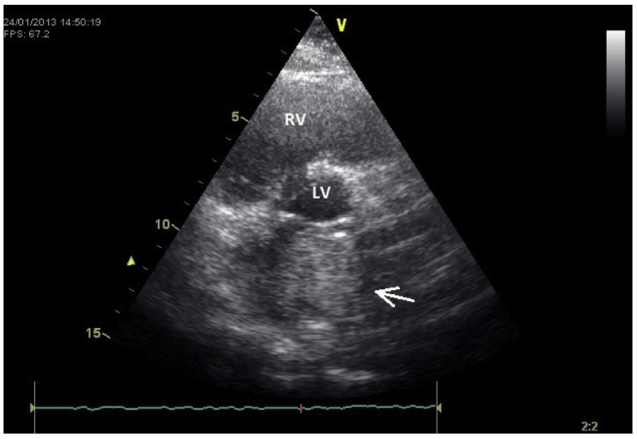

Fig. 4 Normal short axis view of the ventricles. Left atrial myxoma (arrow). LV-left ventricle, RV-right ventricle, LA-left atrium, RA-right atrium, and Ao-aorta.

an important alternative for these patients. However, the increased risk of serious bleeding events, including hemorrhages to the central nervous system, in this group of patients has to be considered.

In summary, there is no current evidence that would justify departure from the adopted rule of immediate surgical therapy for a left atrial myxoma, especially considering the unpredictability of its natural course and hazards associated with its complications. However, in some groups of patients, especially in elderly patients who have a high surgical risk and short life expectancy or who wish to avoid surgery, the non-surgical option is worth consideration.

Statements

We state that this manuscript has not been previously published and is not under consideration in the same or in a substantially similar form in any other peer-reviewed media.

All authors listed have contributed significantly to the final manuscript. To the best of our knowledge, no conflicts of interest, financial or otherwise, exist.

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