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STUDIUM PRZYPADKU / *CLINICAL VIGNETTE*

Cardiac malignant peripheral nerve sheath tumours arising from atrial neurofibroma as an unusual complication of neurofibromatosis

Złośliwy nowotwór wywodzący się z osłonek nerwów obwodowych rozwijający się z nerwiakowłókniaka jako powikłanie neurofibromatozy

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Being relatively rare, primary malignant cardiac neoplasms occur less commonly than metastatic disease of the heart. The most common cardiac tumour group comprises mesenchymal tumours with unarguable dominance of myxomas amid benign lesions and various sarcomas as representatives of malignancy. Malignant peripheral nerve sheath tumours (MPNST) are sarcomas that originate from peripheral nerves from cells associated with the nerve sheath, such as Schwann cells or perineural cells like fibroblasts or myofibroblasts. Currently, we report the case of a 60-year-old female patient, untreated cardiologically, admitted to the Clinic of Cardiology due to dyspnoea, weakness, and significant reduction of exercise tolerance. Echocardiography revealed the presence of a tumour in the left atrium, growing into the interatrial septum and penetrating into the right atrium. The tumour obstructed blood flow to the left ventricle and penetrated into the left ventricle during diastole (Fig. 1A, B). Magnetic resonance imaging (MRI) confirmed the presence of the tumour (Fig. 1C, D). Due to worsening heart failure an urgent cardiac surgery was performed. Intraoperative examination revealed a large tumour measuring $8 \times 6 \times 4.5$ cm in the left atrium and $3.5 \times 2 \times 1.5$ cm in the right atrium, which was blocking the mouths of pulmonary veins and the mitral valve and was invading the surrounding coronary sinus. The majority of tumour mass was excised with slender or doubtful surgical margins for the obvious reason of its location. Histopathological examination revealed a low-grade myxoid malignant peripheral nerve sheath tumour, which developed from pre-existing neurofibroma (Fig. 2). Since surgery, heart failure symptoms have been disappearing gradually. The patient has been under cardiological observation by standard MRI and positron emission tomography. The disease-free period was restricted to two years, then local recurrence was stated. Secondary surgery has been performed and the pathology report confirmed MPNST recurrence. Until the present day no distant metastases have been revealed.

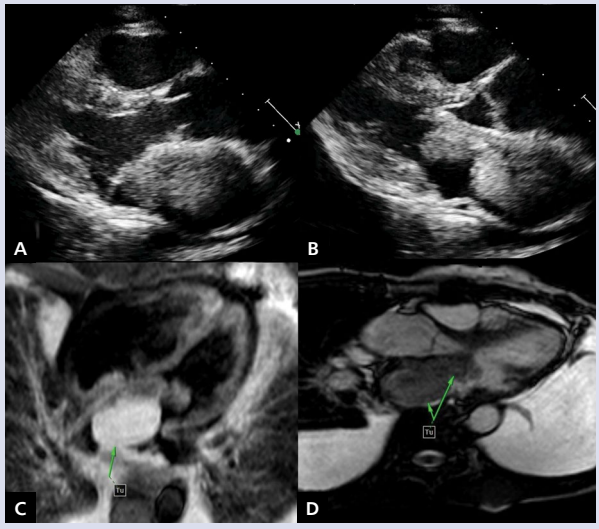

Figure 1. **A**. Echocardiography, projection of parasternal long axis, large tumour in the left atrium (LA); **B**. Echocardiography, left atrial tumour penetrates the left ventricle (LV) during diastole and obstructs blood flow to the LV; **C**. Magnetic resonance imaging (MRI) of the heart shows the tumour in the LA (arrow), which invades the septum and protrudes into the LV; **D**. MRI of the tumour in the LA (arrow), image after administration of contrast

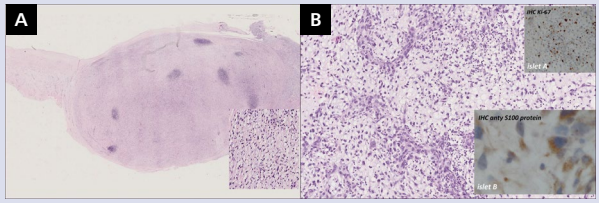

Figure 2. Panel **A** (left side) presents well-circumscribed, endocardium-covered tumour with typical histopathological pattern for neurofibroma (pasted islet with high magnification). Panel **B** (right side) depicts malignant transformation with abundant cellular atypia, significant proliferative index at 30% (islet 1), and positive reactivity for S-100 protein (islet 2)

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