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B-cell lymphoma in a 70 year-old male patient with a cardiac tumour

Chłoniak B-komórkowy u 70-letniego mężczyzny z guzem serca

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Abstract

We present a case of a 70 year-old male with B-cell lymphoma of which the first clinical presentation was cardiac infiltration. The patient underwent full chemotherapy with complete tumour regression.

Key words: B-cell lymphoma, cardiac infiltration

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CASE REPORT

A 70 year-old patient was admitted to our department with a three month history of exercise intolerance and dyspnoea (NYHA III), primarily associated with combined aortic valve disease. The patient had never been hospitalised and was fully physically active before these complaints occurred. Family history was negative. At admission: good general condition, except for jugular veins distension and bradycardia, no other pathological signs. The ECG revealed a third degree atrioventricular (III° A-V) block with an escape junctional rhythm (HR = 50/min). Transthoracic (TTE) and transoesophageal (TEE) echocardiography confirmed a moderate aortic valve stenosis with massive valvular calcifications, hyperkinetic, non-enlarged left ventricle and excessive pericardial fluid (30 mm layer). Pericardiocentesis was performed and 790 mL of dark-yellow exudative fluid was drained. Coronary angiography revealed the critical lesion within the second segment of right coronary artery — primary coronary angioplasty with a bare metal stent implantation was performed (TIMI 3). However, a III° A-V block persisted, which prompted us to implant a DDDR pacemaker. Pericardial fluid cytology, received on the third day after pericardiocentesis, revealed Burkitt-like cells. Laboratory blood tests showed high erythrocyte sedimentation rate (ESR) (53/100 mm/h) and low lipid parameters.

A 64-multislice computer tomography (MSCT) of the chest revealed a left atrium posterior wall (thickness up to 2 cm) and inter-atrial septum (IAS) (thickness up to 3 cm) neoplasm infiltration with a density of 70 HU (soft tissue) — no fluid in pleura or pericardium (Fig. 1). Further imaging tests showed no other primary or secondary neoplastic infiltrations. The recent pacemaker implantation and coronary angioplasty procedure motivated us to withdraw from the tumour biopsy. Afterwards, the patient was discharged in a good and stable general condition with a recommendation of hospitalisation within four weeks.

At second admission, the patient was complaining of significant fatigue, apathy, and weight loss (5 kg in eight weeks). There were no clinical signs of circulatory failure. Physical examination revealed supraclavicular and axillar lymphadenopathy (3 cm). Control TTE demonstrated blurrily sepa-

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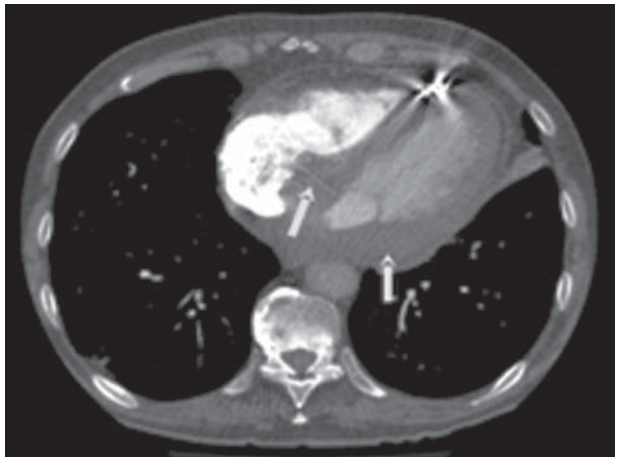

Figure 1. Scan of 64-multislice computer tomography; infiltration of the left atrial posterior wall and inter-atrial septum (arrow)

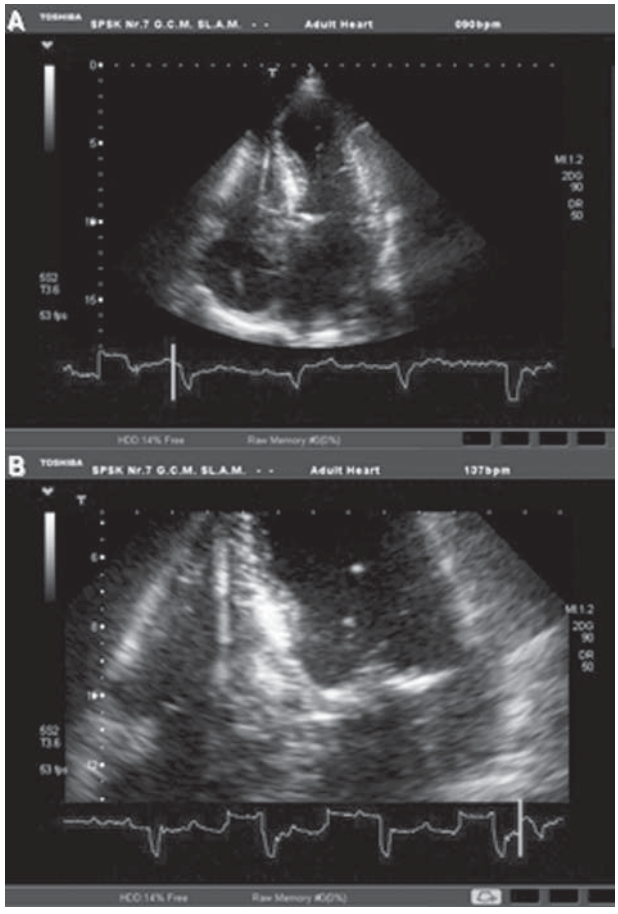

Figure 2. Transthoracic echocardiography — tumour localised within the right atrium and the right ventricle along inter-atrial septum; **A.** Four-chamber view; **B.** Zoom scan — tumour and pacemaker electrode

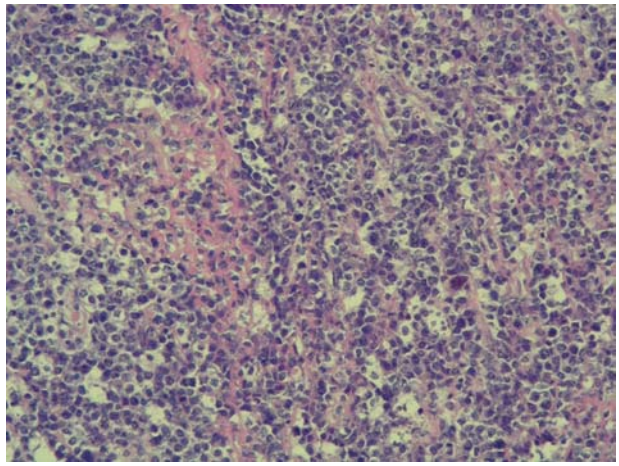

Figure 3. Histopathological examination of the supraclavicular lymph node: 1) effacement of correct lymph node structure; 2) extensive infiltration of mononuclear, middle size, *little bit* polymorphic, round shaped or angular with macrogranular chromatin and not quite visible nucleoli; cytoplasm moderately abundant, basophilic, partially with plasmocyte-like pattern; numerous division figures, intercellular nucleus debris, apoptotic bodies, microgranular necrosis; 3) diffused macrophages with bright cytoplasm giving the 'starry sky' pattern (magnification × 350, and reproduced here at 70%)

rated tumour, localised within right atrium, right ventricle, along IAS and inter-ventricular septum (IVS) approximately 57 × 22 mm in dimensions (Fig. 2) and unaffected haemodynamic cardiac function. The 64-MSCT and ultrasonography showed several lymph nodes (up to 20 mm) in mediastinum and only a single lymph node (17 mm) of extra-thoracic localisation, at the pancreas head area. Laboratory blood tests revealed increased ESR (80 mm/h), CRP (86.2 mg/L) and LDH (2614 U/L). The supraclavicular lymph node was taken for histopathological examination (Fig. 3): the primary diagnosis was lymphoma malignum. The immunohistochemical examination results were: CD20(+), CD3(+), CD43(+), CD79alpha(+), CD68(+), Bcl-2(+), Ki67>95%, TdT(–). The molecular studies revealed MYC translocation.

Finally, a diagnosis of B-cell lymphoma unclassifiable with features intermediate between diffuse large B-cell lymphoma (DLBCL) and Burkitt lymphoma (BL) was established. No pathology was found in the dermatomuscular biopsy. Afterwards, chemotherapy was administered (CHOP: cyclophosphamide, doxorubicin, vincristine, prednisone). The TTE examination performed just after the first cycle of treatment revealed an impressive regression of neoplastic heart infiltration limited only to the part of IAS (Fig. 4A). The patient was in a good general clinical condition with only a single axillary lymph node palpable. Further observation revealed complete regression of the cardiac tumour (Fig. 4B),

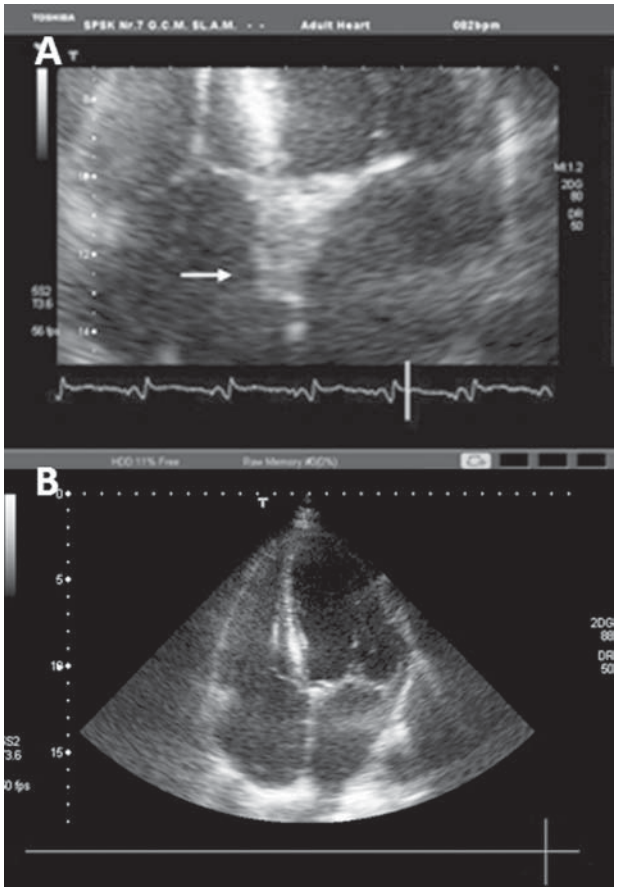

Figure 4. Transthoracic echocardiography, four-chamber view; **A.** Zoom scan; significant regression of the tumour limited to the inter-atrial septum (arrow) after the first cycle of treatment; **B.** Complete echocardiographical regression of the heart tumour

which was also confirmed during aortic valve replacement cardiosurgery performed two years after the chemotherapy.

DISCUSSION

The reported case and the sequence of clinical findings strongly suggest that cardiac infiltration was the first presentation of BL and preceded peripheral lymphadenopathy. Pericardial effusion with atypical cells and a III° A-V block were probably the first clinical signs of the disease. It is worth noting that while initial TTE and TEE revealed no structural abnormalities, it was the 64-MSCT examination performed due to pericardial fluid cytology that showed the infiltrative structure within the atria.

Available case reports have described incidentally found primary malignant heart lymphomas confirmed after its resection [1–5]. Primary cardiac non-Hodgkin's lymphomas are extremely rare and occur mainly in immunocompromised patients [6]. Of 35 cases of primary cardiac non-Hodgkin's lymphoma reported [7], 22 of them had DLBCLs.

In the presented case, tumour biopsy was temporarily delayed (double antiplatelet treatment, implanted stent). The nature of pathology (benign, malignant) was unknown, which convinced us that continuation of diagnostic procedures and possible cardiosurgical intervention would be the optimal solution. According to the recent update of the WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues [8], an eventual diagnosis of B-cell lymphoma unclassifiable with features intermediate between DLBCL and BL was established. Several factors suggested the diagnosis of intravascular large B-cell lymphoma: advanced age, potentially primary heart tumour, short medical history, aggressive course, and initial histopathological lymph node evaluation. However, there were no skin abnormalities, or infiltration of central nervous system and the dermatomuscular biopsy did not reveal pathology. Moreover, high percentage of active cells in the lymph node examined — Ki67 > 95% did not support the diagnosis of DLBCL [8, 9].

We cannot unequivocally exclude a potential coincidence of lymphoma and another heart tumour. However, the sequence of presented events suggests the primary heart localisation. According to the available evidence based on case reports, only one third of patients survive longer than one year [10]. Our patient underwent all required chemotherapy cycles and further clinical observation revealed complete tumour regression.

Conflict of interest: *none declared*

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