Left atrial and ventricular malignant cardiac tumour in a 62-year-old woman: a case report

Złośliwy guz lewego przedsionka i lewej komory serca u 62 letniej chorej. Opis przypadku

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Abstract

Sarcoma is one of the most common malignant primary heart tumours. It accounts for over 76-95% of malignant heart tumours. It can cause serious complications such as pulmonary and peripheral embolism, reduced atrial and ventricular filling, and subsequently reduced cardiac output, arrhythmias, valve dysfunction, and sudden cardiac death. For these reasons early diagnosis and subsequent surgical treatment are the most important. However, complete resection of sarcomas is not always possible and the long-term prospects for resected malignant tumours are adverse. In this article we present a case report of the surgical excision of a left atrial and ventricular sarcoma in a 62-year-old woman.

Key words: malignant heart tumour, sarcoma, surgical management.

Streszczenie

Mięsak jest najczęstszym złośliwym pierwotnym guzem serca. Stanowi on ok. 76–95% wszystkich złośliwych guzów serca. Może być przyczyną wystąpienia zatorowości płucnej, obwodowej, zmniejszonego napływu krwi do przedsionków i komór, co powoduje zmniejszenie rzutu serca, zaburzenia rytmu serca, dysfunkcję zastawkową, nagłe zgony. Z tego powodu wymaga wczesnego rozpoznania i leczenia operacyjnego. Jednakże całkowita resekcja chirurgiczna mięsaka nie zawsze jest możliwa, a rokowanie odległe jest niepomyślne. W artykule przedstawiono przypadek usunięcia operacyjnego mięsaka lewego przedsionka i lewej komory serca u 62 letniej kobiety.

Słowa kluczowe: złośliwy guz serca, mięsak serca, leczenie operacyjne.

Introduction

Primary heart tumours are rare. Sarcoma is one of the most common malignant primary heart tumours [1]. It accounts for over 76-95% of malignant heart tumours [2]. The American Medical Association reported that its incidence, according to autopsy series, is about 0.0017% [3]. Sarcoma can cause serious complications such as pulmonary and peripheral embolism; its mass can obstruct intracardiac blood flow, can lead to reduced atrial and ventricular filling [4], and subsequently cause reduced cardiac output and arrhythmias; it can interfere with valve function, as well as cause cardiac tamponade [5] or sudden cardiac death [6]. Heart tumours require operative excision; however, complete resection of sarcomas is not always possible and the long-term prospects for incompletely resected malignant tumours are adverse [7, 8]. Transplantation may also be an option for those with extensive local disease and malignant tumours [9, 10]. We describe a case of a 62-year-old woman who underwent a surgical excision of a left atrial and ventricular heart tumour. Histopathological examination confirmed the malignant nature of the tumour: pleomorphic sarcoma.

Case report

A female patient, 62 years old, admitted to the Cardiac Surgery Department on 17 August, 2009, was diagnosed

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with a left atrial and ventricular malignant heart tumour. In the medical history, hospitalization on the pulmonary ward of the local hospital (03.08.2009 – 09.08.2009) due to general weakness and persistent cough, and on the cardiological ward of the University Clinical Hospital (09.08.2009 – 16.08.2009). Height – 170 cm, weight – 66 kg, BMI 23. RR 130/70 mm Hg. Morphology – WBC $10.0x10^9$/L, HGB 12.8, PLT 289. Electrolytes in the norm. CRP 25.70 mg/L, procalcitonin (PCT) 0.03 ng/ml, ASO 84 IU/ml. In ECG – normal heart rhythm, regular sinus rhythm of 90/m frequency, Q wave in II, III, V5-V6. In echocardiography examination: in the left ventricle a visible large "cauliflower-like" structure, morphologically similar to myxoma, probably coming out of the anterior leaflet base of the mitral valve, of size 40x25 mm, with substantial mobility, narrowing the left ventricle regurgitation passage up to 3.5 m/s (grad. max. 50 mm Hg). In the left atrium under the anterior leaflet of the mitral valve – a hyperechogenic growth of size 20x10 mm and low mobility. Mitral insufficiency – II/III°. Threadlike structure arising under the anterior mitral valve from the left atrium side. Good systolic function of the left ventricle (LV) EF 58%. Small amount of fluid in pericardial sac. In echocardiographic transoesophageal examination (TEE), the interatrial septum and part of the left atrial wall are "wallpapered" with a jelly-like tissue of thickness about 10 mm and unequal villous surface, an additional structure at the anterior leaflet of the mitral valve of length about 14 mm, on the left ventricular outflow tract – a growth of size 4x2 cm indicating organic continuity with the base of the anterior leaflet of the mitral valve. On the ventricular side the presence of a 7-mm tumour pedicle. Mitral insufficiency of 2nd degree, tricuspid of 1st degree. Left cardiac auricle free, with a flow speed of 80 cm/s. Delamination of pericardial lamellae – 5-6 mm of fluid in pericardial sac (10.08.2009).

In angiocardiography: coronary arteries without essential stenosis. In CT examination of the thorax (06.08.2009) a cord-like, small fibrosis of the base of both lungs was observed. Lack of focal lesions in pulmonary parenchyma, presence of enlarged, not forming clusters, lymph nodes of mediastinum (subcarinal, bilateral lower paratracheal, with left side preponderance, single peribronchial). Remaining visible lymph nodes of the left hilus, mediastinum – not enlarged (06.08.2009). The important, diagnosis-demanding "lymph problem" was noticed by a pulmonary consultant examining the patient; he suggested lymph nodes sampling for pathomorphological examination during cardiosurgery in view of cardiac operation priority.

Thyroid USG: non-homogeneous nodule of 38x29x33 mm in the upper pole of the right thyroid lobe, without infiltration traits of neighbouring structures. Cervical lymph nodes not enlarged (07.08.2009). Histopathological examination of thyroid: benign tumour of the left lobe.

After anaesthesiological-cardiosurgical consultations the patient was qualified for operational treatment of heart tumour on schedule.

Course of surgical anaesthesia and procedure (19.08.2009)

Initial RR 130/70 mm Hg, regular sinus rhythm (RSR) 80/m. Introduction to anaesthesia: fentanyl (FTN) 3.15 ug/kg, Hypnomidate 0.21 mg/kg, pancuronium 0.06 mg/kg. After endotracheal intubation RR was 125/65 mm Hg, RSR 85/m. Maintenance of anaesthesia: FTN in a continuous infusion of 0.087 ug/kg/min, midazolam 1.75 ug/kg/min, fractionated doses of pancuronium. Ventilation with breathing mixture of O_2 45% with air. From the surgical incision to extracorporeal circulation connection – sevoflurane 2-0.5 vol%.

The course of anaesthesia was without any complications, with good, accepted, mid-operational stabilization of the circulatory system before and during the extracorporeal circulation (ECC) as well as post-surgery.

During the cardiosurgical intervention, in extracorporeal circulation conditions, the non-encysted tumour mass was excised from the left atrium and ventricle of the heart. Biopsy material was obtained: tumour and lymph nodes of the mediastinum taken during the surgery were sent for further histopathological examination.

Early post-operative period at the Intensive Therapy Unit (ITU) of the Anaesthesiology Clinic and Intensive Cardiological Care – not complicated. The patient was extubated in the 6th hour after the surgery, respiratory competent, circulatory stable, without a temperature. No episodes of consciousness disorders, and no cardiac arrhythmia or circulatory insufficiency were observed.

On 21.08.2009 – in the second 24-hour post-operative period – the patient was discharged from the ITU. On the 8th day of stay at the Cardiac Surgery Department, the patient was referred to the Oncology Institute for further treatment.

Morphology and Immunohistochemistry

Macroscopic examination of the left atrial cardiac tumour showed four fragments of white-yellowish solid masses measuring: 4.5x3.5x0.5 cm, 5x4.5x1 cm, 6x1.5x1.5 cm, and 6.5x5.5x1.3 cm. The left ventricular cardiac tumour appeared as two yellowish solid bits, the largest of which measured 1.2x1.2x1.3 cm. The outer surface of the masses was smooth. The tissue was fixed in 4% formaldehyde solution, embedded in paraffin, and stained with haematoxylin and eosin. Histologically, tumours located in the left atrium and in the left ventricle were composed of spindle and oval cells with abundant eosinophilic cytoplasm and hyperchromatic nuclei with prominent nucleoli (Fig. 1). Some of the spindle cells were oriented in a storiform pattern with scattered, large round pleomorphic cells. Bizarre multinucleated cells were interspersed with the tumour cells (Fig. 2). Some parts of the tumour consisted of lipocytes and myxoid foci. Mitoses were prominent. Foci of tumour necrosis were also seen. The histological examination was completed with immunohistochemistry using antibodies (DAKO) against: cytokeratin, vimentin, desmin, α-SMA, CD34, and CD68. Visualization of the immunological reactions was performed

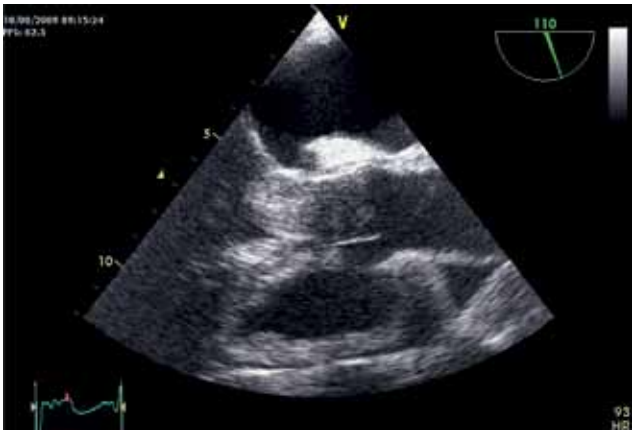

Fig. 1. Transthoracic echocardiography (TTE). The image in long axis, parasternal application of the head. In the left ventricular outflow tract a tumour was observed (Tu 1), partially blocking the outflow tract. In the left atrium a tumour was observed (Tu 2). Further description in the text

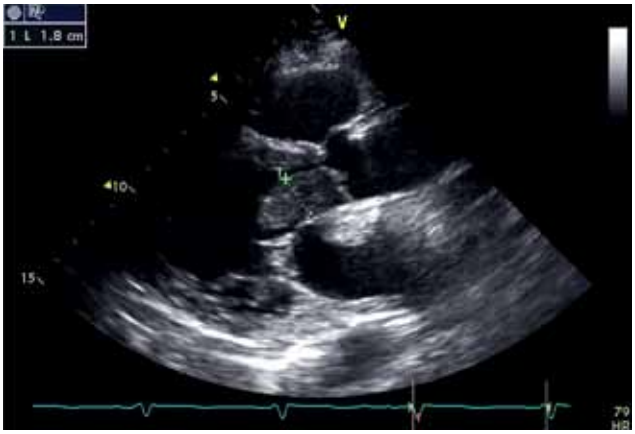

Fig. 2. Transoesophageal echocardiography (TEE). Mid-oesophageal projection of 110°. The presence of tumours registered in TTE confirmed. Further description in the text

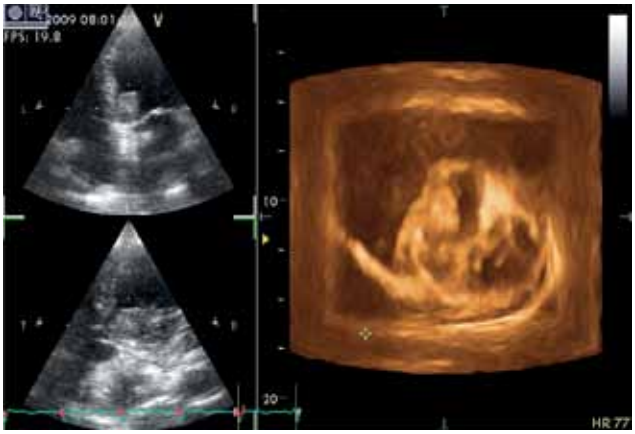

Fig. 3. Three-dimensional echocardiography allowed for a more thorough evaluation of endocardial tumour location

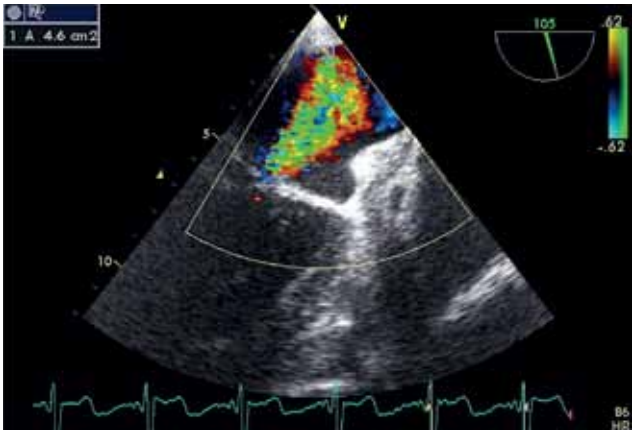

Fig. 4. Transoesophageal echocardiography (TEE). Mid-oesophageal projection of 105°. A substantial mitral incompetence was shown. The wave reaching the left atrium roof

using the immunoperoxidase method with LSAB/Universal Kit (DAKO). As a chromogen diaminobenzidine (DAB, DAKO) was used. The tumour was immunoreactive for vimentin (Fig. 3) and CD68 (Fig. 4). Immunohistochemical stains for cytokeratin, desmin, α-SMA, and CD34 were negative. Based on the morphology and immunohistochemistry, undifferentiated (high-grade) pleomorphic sarcoma was diagnosed.

Discussion

The estimated frequency of primary heart tumours is 0.0017% [3], 75% of which are benign heart neoplasms, and 25% malignant neoplasms, most often sarcomas, which constitute 1% of all malignant human neoplasms [11].

Primary cardiac sarcomas may occur practically in every cardiac cavity [12]. Statistically they are found most often in the left atrium and left cardiac ventricle. They occur at every age, affecting women and men equally. They grow from undifferentiated mother cells resulting from multistage genetic aberrations of these cells [13].

Reports describing chromosomal disorders concerning cardiac sarcoma are scarce and they refer to chromosomes 11p11 and 19p13 and "overproduction" of the genes MDMD2, CDK2, SAS, and CHOP of chromosome 12q 13-14 [11, 14], and they do not have established diagnostic and prognostic significance.

Sarcoma aetiology, in most cases, is unknown, and it is assumed that the development of some of them may have an influence on virus infections, lowering of immunity, irradiation, and presence of proto-oncogenes SAS, MDMD2, CDK4, and CHOP [11].

Intravital diagnostics of malignant cardiac neoplasm is difficult due to the lack of characteristic clinical symptoms [15].

Clinical symptoms of a primary pleomorphic cardiac sarcoma are not characteristic. The most frequent

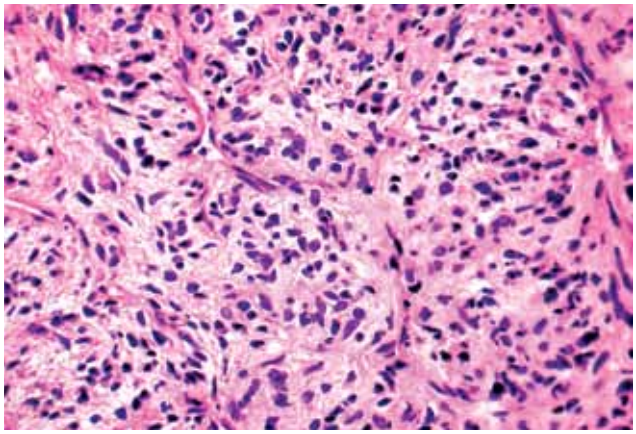

Fig. 5. Cluster of spindle and oval cells with hyperchromatic nuclei and irregular coarse nuclear chromatin. H&E staining, magnification 200x

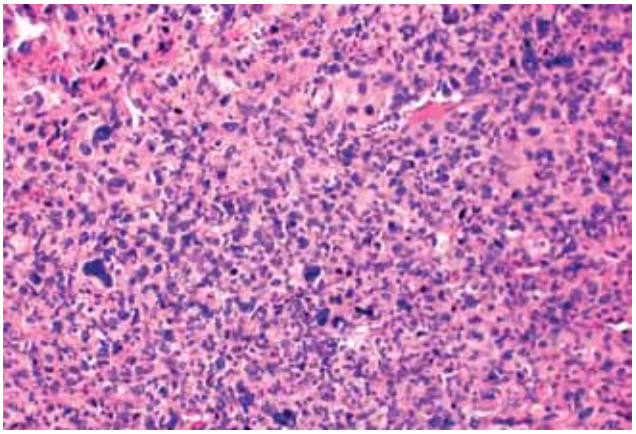

Fig. 6. Multinucleated giant cells scattered within pleomorphic tumour cells. H&E staining, magnification 200x

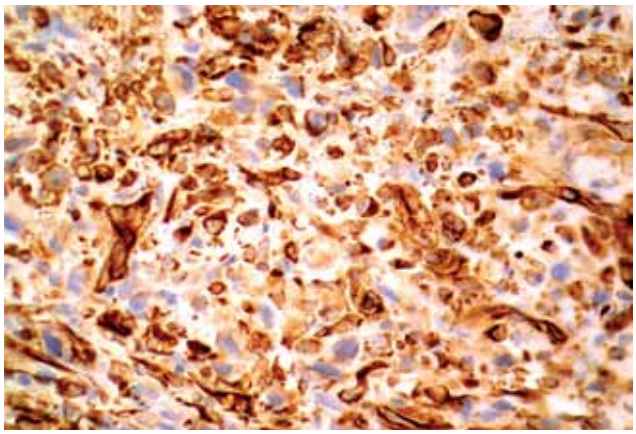

Fig. 7. Immunopositivity of vimentin in neoplastic cells. Magnification 400x

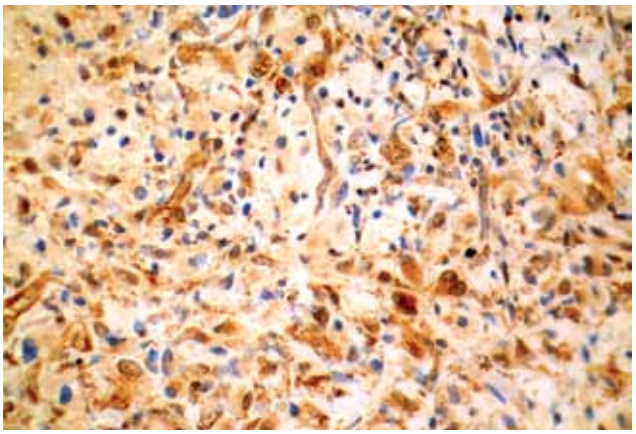

Fig. 8. Cytoplasmic immunoreactivity for CD68 in some neoplastic cells. Magnification 400x

complaints are: paroxysmal dry cough, effort and night dyspnoea, temperature, intensive perspiration, pain in the chest, fainting, epileptic seizures, spitting blood [6], heart palpitation, lower extremities oedema, lesions on skin (rash, pruritus, dry skin, extravasation), and body mass loss. This sarcoma can cause occurrence of intracardiac shunt [15], cardiac arrhythmia, and cardiac insufficiency [4], and can contribute to sudden cardiac death [1], or sudden death without noticeable cause [8, 15].

Sarcomas may produce metastases to various organs. The most common sites of primary sarcoma metastasis are, according to frequency of occurrence: lungs, lymph nodes, pleura, thoracic wall, diaphragm, skeletal system, liver, skin, brain, spleen, testicles, kidneys, and suprarenal glands [1, 15]. Metastasis can sometimes be asymptomatic and is discovered incidentally during surgery, most often in the thoracic region or during autopsy [15]. A key role in the intravital diagnosis of cardiac sarcomas is played by transoesophageal echocardiography (TEE), which allows the tumour to be revealed and located [15-17]. Less helpful are: computed tomography (CT), magnetic resonance imaging (MRI), angiographic examination, x-ray examination of the thorax, and laboratory investigations [16]. Prognosis in cases of primary cardiac sarcoma is unfavourable due to the high frequency of local recurrences with infiltration of neighbouring organs [8], as well as distant metastasis [1, 15]; and surgical treatment relying on complete or partial tumour excision is only possible in some cases [7, 8, 12]. Sporadically, attempts of heart or heart and lung transplantation are undertaken [8-10, 16, 18]. Efficiency of complementary therapy in the form of radiotherapy and/or chemotherapy is not satisfactory. Mean survival rate is 6-16.5 months [15, 19].

Conclusion

The described case shows the role which is played by transoesophageal echocardiography (TEE) in an intravital diagnosis of cardiac sarcoma. TEE allowed us, despite the lack of characteristic clinical symptoms (weakness, persistent cough), as well as non-homogeneous thoracic CT examination results and laboratory investigations, to establish the diagnosis which consequently allowed us to take the decision of operational excision of left atrial and ventricular heart tumour. Unfortunately, the definition

of tumour histological type (pleomorphic sarcoma) is not, in this case, and in spite of good early surgical treatment results and referral of the patient for additional oncological therapy, a favourable sign of prognosis as to health.

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