Kardiologia Polska 2013; 71, 12: 1296–1298; DOI: 10.5603/KP.2013.0328 ISSN 0022–9032

OPIS PRZYPADKU / *CASE REPORT*

Angiosarcoma of the heart — a diagnostic pitfall

Naczyniakomięsak serca — pułapki diagnostyczne

Piotr Gajewski[1], Wiesława Błaszak-Ciećwierska[1], Sławomir Chojnowski[1], Artur Bartczak[2], Jacek Różański[3], Marek Dąbrowski[1, 4]

[1]Cardiology Clinic of Physiotherapy Division, 2nd Faculty of Medicine, Medical University of Warsaw, Warsaw, Poland
[2]Department of Pathomorphology, Medical Centre of Postgraduate Education, Warsaw, Poland
[3]Cardiosurgery and Transplatology Clinic, Institute of Cardiology in Warsaw, Warsaw, Poland
[4]Cardiac Unit for Diagnosis and Therapy, M. Mossakowski Medical Research Centre, Polish Academy of Science in Bielanski Hospital, Warsaw, Poland

Abstract

Primary tumours of the heart are extremely rare. Sarcomas are considered to be the most frequent histological type. This study presents two cases with a two-week and two-month history of fatigue, fever and shortness of breath. In both cases, cardiac tumours with pericardial effusion were diagnosed by means of echocardiography, being responsible for the above-mentioned symptoms. At the beginning of the diagnostic process, sarcomas seemed to be the likeliest cause of these symptoms. Establishment of the histopathological diagnosis based on the tumour biopsy turned out to be very difficult, and this delayed further therapeutic procedures.

Key words: angiosarcoma, heart, echocardiography, 'cardiac MICE', open heart biopsy

Kardiol Pol 2013; 71, 12: 1296–1298

INTRODUCTION

Heart tumours may be divided into primary tumours and metastatic lesions, which occur more often. About 25% of lesions are considered to be malignant, with angiosarcoma being the most commonly diagnosed histological type [1]. Angiosarcomas are rare primary cardiac tumours occurring in 1.7/100,000 cases, more often in male patients [2]. The peak incidence of angiosarcoma is observed in the fourth decade of life. In most cases, the tumour is located in the right atrium [3]. Initial non-specific symptoms, such as shortness of breath, (64%), loss of body weight, fever, night sweating (43%), pain (43%), deterioration in exercise tolerance (29%), tachycardia (21%), cough (21%) and neurological symptoms develop in the advanced stage of the disease [3].

The aim of this study was to present a case of angiosarcoma and draw attention to the rapid course of the disease, degree of malignancy, as well as clinical and pathomorphological diagnostic difficulties.

CASE 1

A 66-year-old male patient with a history of myocardial infarction treated by means of angioplasty of the right coronary artery with stent implantation (2002), permanent atrial fibrillation and hypertension, was admitted to the Department for diagnostics of deteriorating exercise tolerance of several weeks' duration. Transthoracic echocardiography (TTE) revealed the presence of 40 mm of fluid behind the left ventricle. An additional lesion 36 × 24 mm in diameter was observed from the side of the right atrium (RA). Computed tomography scan showed the presence of pericardial fluid and a 43 mm layer of right-sided pleural effusion. On the fourth day of hospitalisation, we evacuated 1,200 mL of pericardial haemorrhagic fluid, obtaining an improvement in the patient's general condition and a reduction in exertion dyspnoea. The cytological examination of the fluid showed no abnormalities. Transoesophageal echocardiography (TEE) revealed the presence of a large polymorphic tumour located in the upper-posterior part of the RA. The tumour had infiltrated the atrial walls, being partly located in the pericardium (Fig. 1).

A magnetic resonance imaging (MRI) examination confirmed the presence of a RA tumour, 90 × 91 × 54 mm in diameter. The image suggested the possibility of an angiosarcoma. There were no abnormalities in abdominal ultrasonography. The patient was disqualified from cardiosurgery aimed

Address for correspondence:
Piotr Gajewski, MD, Cardiology Clinic of Physiotherapy Division, 2nd Faculty of Medicine, Medical University of Warsaw, ul. Cegłowska 80, 01–809 Warszawa, Poland, e-mail: gajewskipiotr10@gmail.com
Received: 12.06.2012 **Accepted:** 10.09.2012

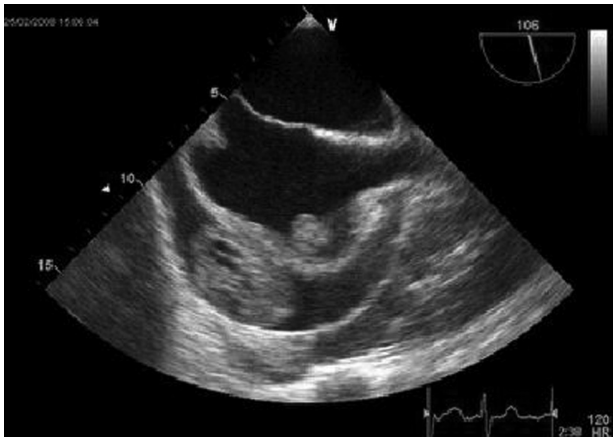

Figure 1. Transoesophageal echocardiography. Large tumour located in the pericardiac space infiltrating the atrial wall, also visible in its lumen

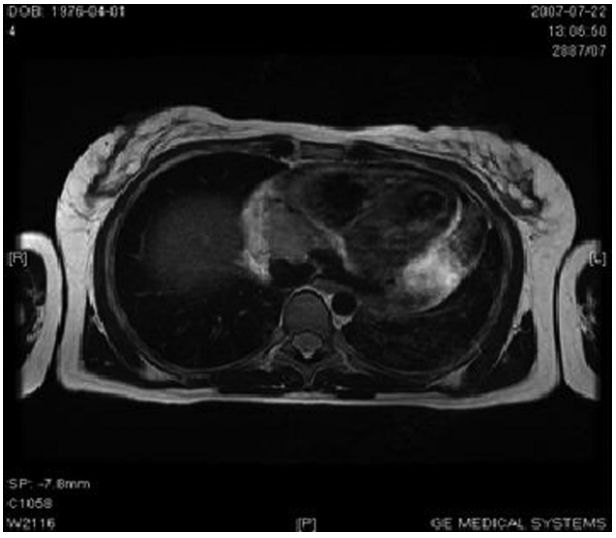

Figure 2. Magnetic resonance imaging scan. Visible polymorphic tumour (36 × 28 × 30 mm) located in the pericardial sac filled with fluid, located at the border of the inferior caval vein and right atrium

at radical excision of the neoplastic lesion, due to the stage of the disease. However, he was qualified for open heart biopsy, in order to obtain samples for pathomorphological evaluation, which would constitute the basis for determining further oncological treatment. Due to difficulties in interpreting the results of the collected samples, the above-mentioned were consulted in a foreign centre. Thus, on the basis of a wide immunohistochemical panel, the following diagnosis was established: mesothelial/monocytic incidental cardiac excrescence — cardiac MICE. Due to the histopathological diagnosis suggesting the benign character of the tumour, the patient was disqualified from oncological treatment. Due to increasing symptoms of right-ventricular heart insufficiency and recurrent fluid in the right pleural cavity, the patient was considered for heart transplantation. Due to the anatomical conditions, the possibility of heart transplantation was abandoned. In spite of the clinical suggestions of tumour malignancy, the oncologists decided against chemotherapy without histopathological confirmation. In spite of therapy, the patient's condition deteriorated, and he died. Due to the unclear clinical picture and cardiac MICE diagnosis, an autopsy was performed. This confirmed the diagnosis of angiosarcoma.

CASE 2

A 31-year-old female patient without a significant medical history was admitted to the Department of Cardiology, due to increasing general weakness and symptoms of right-ventricular insufficiency. TTE demonstrated the presence of pericardial fluid (35–42 mm) without signs of cardiac tamponade, and a polycyclic tumour (35 × 35 mm in diameter) in the pericardial sac, infiltrating the RA. Pericardiocentesis was performed, obtaining 700 mL of haemorrhagic fluid. The histopathological examination showed no presence of neoplastic cells, and fluid culture was sterile. TEE performed after the pericentesis confirmed the presence of a polycyclic tumour. A series of examinations excluded the metastatic character of the tumour. MRI revealed the presence of a polymorphic tumour, as described in the Figure 2 legend.

The patient was qualified for surgery. The histopathological result was as follows: rabdomyosarcoma, GI malignancy, small lymph nodes without neoplastic cells. According to the opinion of the consulting oncologist, the patient at this stage did not require active treatment, only echocardiographic examinations every three months, and chest and abdominal computed tomography (CT) every six months during the initial two years, and once a year thereafter. The TTE as well as control chest CT examination performed six months after surgery showed no significant abnormalities. After another month, the patient reported back because of a painful infiltration located in the postoperative scar. The histopathological diagnosis of the samples was again: rabdomyosarcoma. However, due to the relatively rapid progression of the disease, the oncologist's previous decisions were verified, including the examination of tissue samples collected during the procedures, and sent to the Institute of Oncology, Warsaw. Thus, an angiosarcoma diagnosis was established and the patient was qualified for chemo- and radiotherapy. Detailed investigations conducted at the Institute demonstrated metastatic lesions in the spine, cranium and sternum. The patient died after three years of intensive therapy as a result of subsequent metastases to the central nervous system. Repeatedly consulted at that time in our centre, she did not present characteristics of recurrence at the site of the removed primary tumour.

DISCUSSION

The first patient complained of typical, although non-specific, symptoms. The diagnosis (a high probability of angiosarcoma)

was rapidly established after performing initial imaging and laboratory examinations. The evacuation of pericardial and pleural fluid did not confirm the presence of atypical cells. The patient was disqualified from surgery due to the size of the tumour, while the decision concerning the implementation of chemotherapy depended on the histopathological result. Open biopsy was performed and the final diagnosis was surprising, but because of the benign character of the tumour the patient did not receive chemotherapy. The subsequent clinical course cast doubt on the established histopathological diagnosis, since 'cardiac MICE' cases mentioned in the literature data did not lead towards haemodynamic disturbances [4]. During the open biopsy, the surgeon was probably afraid to penetrate too deeply into the mass taking superficial samples. This possibly led to an incorrect histopathological diagnosis. A possible solution, as suggested by the treating physicians, was the collection of samples from the atrium, from which the surgeon withdrew on seeing the nature and extent of the infiltration.

In the second case, delayed diagnosis was also connected with difficulties in the proper interpretation of the histopathological result of the removed tumour tissue. This might be evidence of diagnostic difficulties at the level of sarcoma pathomorphology, even in the case of experienced pathologists. The histopathological result probably 'calmed down' the consulting oncologist, who did not recommend treatment, only patient observation. This caused a delay in treatment of nearly six months, implemented at the time of metastases diagnosis.

It seems that in difficult cases there should be closer co-operation between the surgeon and the pathologist, perhaps with the presence of the latter during the surgical intervention. Based on the experience in the treatment of the above-mentioned patients, in case of a suspicion of cardiac sarcomas, one should not solely depend on the opinion of the 'general' oncologist. Such patients should be consulted by teams experienced in the diagnostics and treatment of sarcomas.

Conflict of interest: *none declared*

References

1. Silverman NA. Primary cardiac tumors. Ann Surg, 1980; 191: 127–138.
2. Burke A, Cowan D, Virmani R. Primary sarcomas of the heart. Cancer, 1992; 60: 387–395.
3. Mayer F. Aerbert H. Primary malignant sarcomas of the heart and great vessels in adult patients: a single center experience. Oncologist, 2007; 12: 1134–1142.
4. Veinot JP, Tazelaar HD, Edwards WD. Mesothelial/monocytic incidental cardiac excrescences: cardiac MICE. Mod Pathol, 1994; 7: 9–16.