COR ET VASA 56 (2014) e274–e278

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Case report

Rare myxoid liposarcoma metastasis to the interventricular septum of the heart

Renata Virtová [a,*], *Tomáš Gazdič* [a], *Jan Pirk* [a], *Radka Kočková* [a], *Otakar Bělohlávek* [b], *Kateřina Kamarádová* [c], *Kateřina Kubáčková* [d], *Dana Kautznerová* [e], *Jan Vydra* [f], *Josef Kautzner* [a]

[a] *Cardiology Center, IKEM, Czech Republic*
[b] *Department of Nuclear Medicine and PET Center, Na Homolce Hospital*
[c] *Institute of Pathology and Molecular Medicine, Teaching Hospital in Motol, Czech Republic*
[d] *Department of Oncology and Radiotherapy, Teaching Hospital in Motol, Czech Republic*
[e] *Radiodiagnostic and Interventional Radiology Department, IKEM, Czech Republic*
[f] *Institute of Oncology and Rehabilitation Na Pleši, Czech Republic*

ARTICLE INFO

Article history:
Received 11 July 2013
Accepted 16 July 2013
Available online 23 July 2013

Keywords:
Myxoid liposarcoma
Heart
Cardiac metastasis
CMR
Echocardiography
PET CT

ABSTRACT

Liposarcomas are malignant tumors of the soft tissue. Myxoid liposarcoma is the second most common subtype of these tumors in adults. It accounts for approximately 20% of all malignant soft tissue tumors [1,2]. Peak of its incidence occurs between 40 to 60 years of age with relatively indolent clinical course Matsumoto et al. (2007) [3], Cho et al. (2010) [4], Faiman et al. (2005) [5]. Typical localizations of myxoid liposarcoma comprise limbs, particularly thighs with a tendency to metastasize into extrapulmonary sites such as retroperitoneum, mediastinum, bones. Cardiac metastases are extremely rare.

We present a case of a 36-year-old man with a history of recurrent myxoid liposarcoma. Primary location was in the left popliteal area. After extirpation of the tumor, metastatic tumor was subsequently revealed in the right axilla. Each surgical extirpation was followed by radiation therapy and brachytherapy. Cardiac metastasis was accidentally diagnosed with PET/CT during the staging process. The patient was asymptomatic and was admitted to our institution for further diagnostics and treatment. After confirmation of its location, the tumor was excised. Histological examination revealed myxoid liposarcoma.

Introduction

Liposarcoma is the second most common type of soft tissue sarcomas in adults after fibrous histiocytoma [3,5]. It accounts for approximately 20% of all malignant soft tissue tumors [3]. We recognize five histologic types: well differentiated, dedifferentiated, myxoid, round cell, and pleomophic. Myxoid variant is the second most common type, which is seen most often in fourth and fifth decade of life. Myxoid liposarcoma is characterized by long disease duration and multiple recurrences [4]. We present

* Corresponding author. Tel.: +420 261365108; fax: +420 26136 2989.
E-mail address: wirtx@seznam.cz (R. Virtová).
http://dx.doi.org/10.1016/j.crvasa.2013.07.004

a rare case of cardiac metastasis from a myxoid liposarcoma.

Case presentation

36-year-old man without history of cardiac disease was treated for the first time for myxoid liposarcoma in the left popliteal area in 2007. After surgical resection and radiation therapy, the tumor recurred in the right axilla in 2009. After extirpation of the tumor without adjuvant therapy, another local recurrence was found in 2011. The axillary exenteration was performed repeatedly in April 2011 and March 2012. Positron emission tomography–computed tomography (PET/CT) was carried out as a part of staging process during oncologic follow-up. The scan revealed a hypodense round lesion in the interventricular septum of the heart without abnormal 18F-fluorodeoxyglucose (FDG) accumulation, which meant that glucose consumption was not increased in the lesion. Nonspecifically increased accumulation of the agent was found in the margins of the lesion, which probably represented physiological variant of imaging of increased glycolytic activity in surrounding myocardium (Fig. 1). The patient was referred to our institution (IKEM) for further diagnostic work-up and treatment. He had no symptoms at presentation, and physical examination did not reveal any signs of the disease. ECG was normal. Transthoracic echocardiogram confirmed a nodule 31×25 mm^2 in size in the middle segment of the interventricular septum involving also the basal segment close to the mitral annulus. The tumor protruded into left ventricular cavity and moved with septum. The tumor neither caused hemodynamic compromise nor compressed adjacent structures. Ejection fractions of both ventricles were normal, as well as all valves (Fig. 2A, B). Magnetic resonance imaging confirmed presence of the round lesion 22 mm in diameter with well-defined borders and morphology reminding myxoid tissue (Fig. 3). Subsequently, the patient underwent operation, and the tumor of the septum was resected (Fig. 4A, B, C). The operation and early postoperative course was uncomplicated without arrhythmias during monitoring. Histological examination confirmed a metastasis of myxoid liposarcoma (Fig. 5). The patient has been followed up for 10 months and has had no signs of disease recurrence according to MRI and PET/CT scan (Figs. 6A, B, and 7).

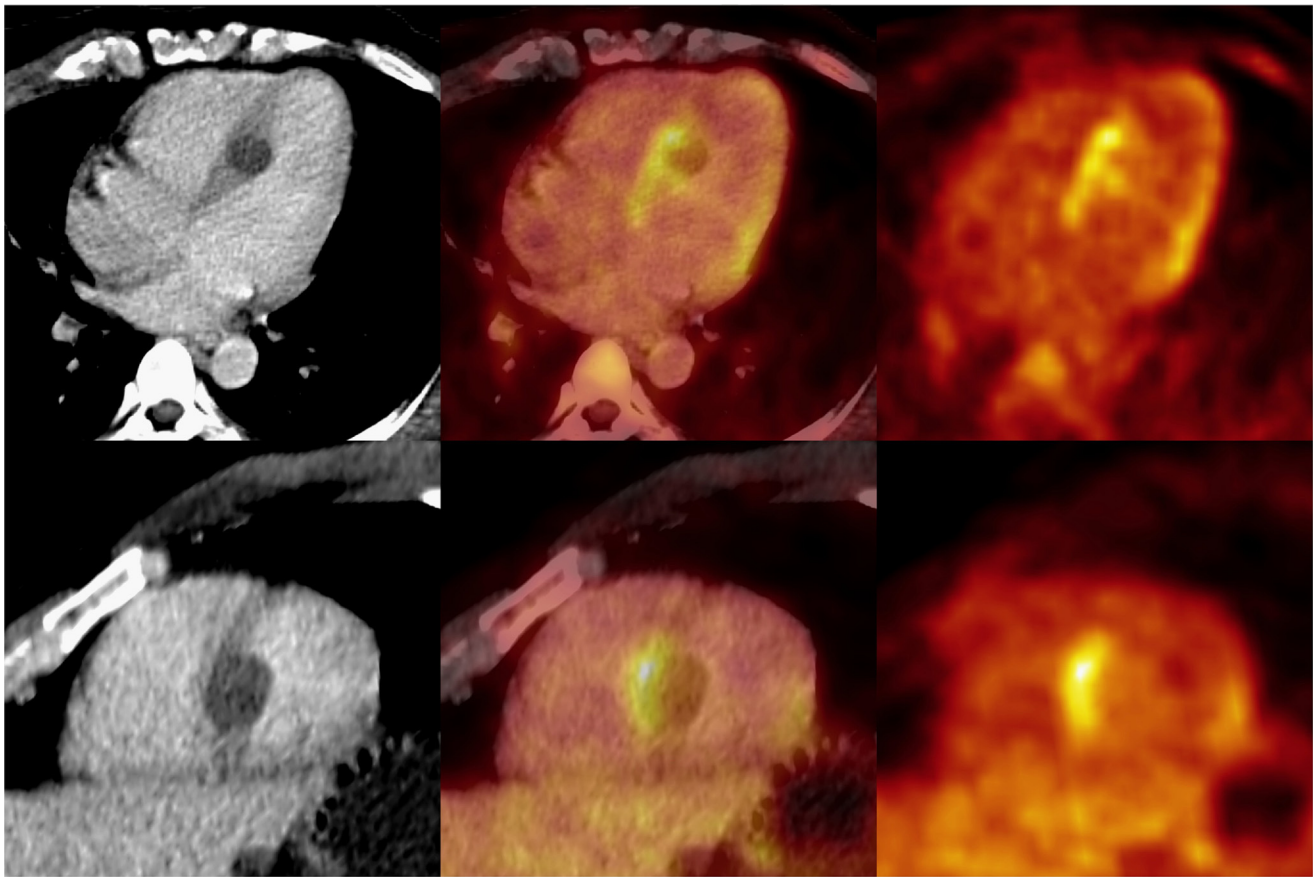

Fig. 1 – PET/CT scan (performed at Na Homolce Hospital). CT component (left panel), PET component (right panel), and FDG-PET/CT fused images (middle panel); top panels shows horizontal long axis slices (viewed from below), and bottom panels show short axis slices (viewed from apex). A round hypodense nodule of 22 mm in diameter is visible in the interventricular septum on the CT scan. The nodule does not demonstrate increased glycolytic activity on the FDG-PET scan. Such findings are usual in low grade liposarcoma. Slightly increased FDG accumulation in surrounding tissues represents physiological inhomogeneous distribution of glycolysis in myocardium.

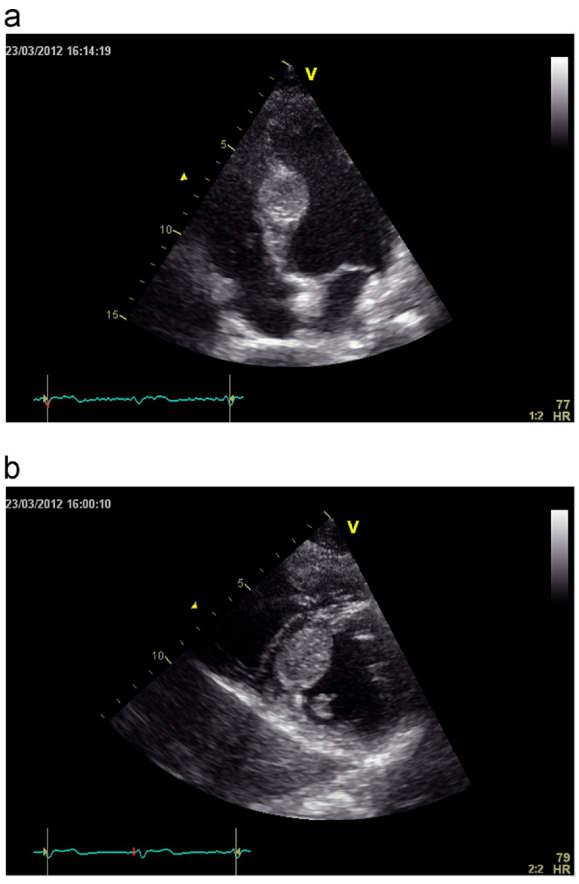

Fig. 2 – Echocardiography – (A) apical four chamber view and (B) short axis view.

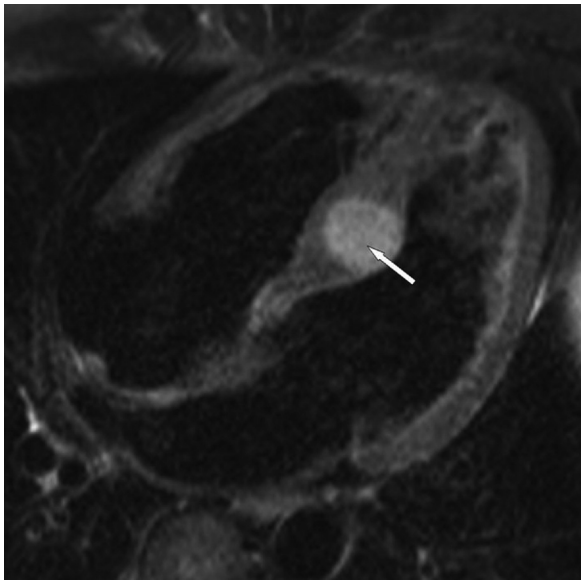

Fig. 3 – Magnetic resonance imaging: a hyperintense nodule in the interventricular septum (native 2D T2-weighted TSE imaging).

Discussion

Malignant heart tumors are very rare. The most common primary tumors occurring in heart are sarcomas. Secondary heart tumors, i.e., metastases of other malignancies as melanoma, lung carcinoma, lymphoma, or breast cancer, are 20–40 times more often than primary lesions [5]. Liposarcoma is rarely occurring mesenchymal tumor belonging to soft tissue sarcomas. It is the second most common malignity of this type in adults after fibrous histiocytoma [3,5], which accounts for about 20% of all malignant soft tissue tumors [1]. The incidence of liposarcoma peaks between 40 and 60 years of age and it usually has relatively long clinically silent course [4,7]. Myxoid liposarcoma is typically found in the extremities, particularly legs; it also occurs quite often in the buttock, retroperitoneum, abdominal cavity, or chest wall [4,5]. Primary tumor in our case was localized in the popliteal area of the left lower extremity. Although it is typical for liposarcomas that they usually metastasize to the lungs, myxoid liposarcoma tends to spread also to extrapulmonary sites as soft tissues, mediastinum, bones, and heart [4,8]. Metastases to the spine are extremely rare [4]. In our case, the first metastasis of the tumor was found in the right axilla.

Heart metastases are also very rare, and their clinical presentation is heterogeneous. Many patients have no symptoms, and the diagnosis is not usually made until post mortem [9]. In our patient, the diagnosis was established during staging for primary tumor. The most common sign in symptomatic patients is pericardial involvement with pericardial effusion. Other usual signs are heart failure, conduction abnormalities, tachyarrhythmia, mechanical obstruction, valve disease, or myocardial rupture [3,7,10]. Symptoms are dependent upon the site and size of the tumor. Tumors in the right side of the heart cause symptoms such as dyspnea and hemoptysis. The ones in

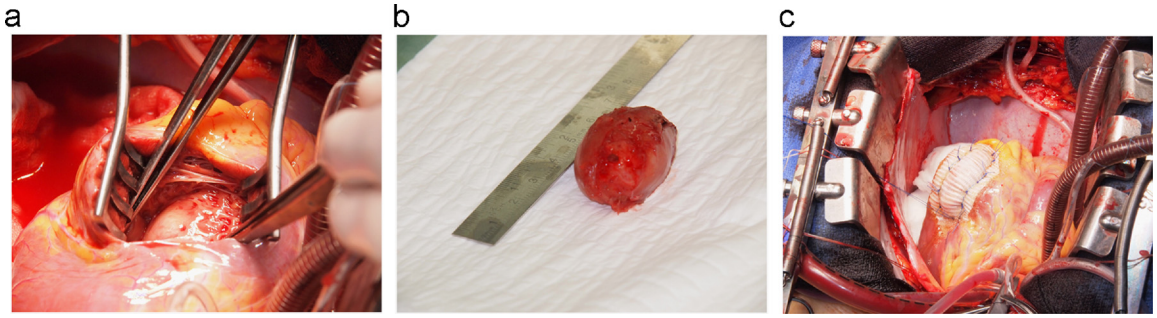

Fig. 4 – View of (A) the interventricular septum with the tumor after ventriculotomy, (B) the excised tumor, and (C) the interventricular septum after ventriculotomy closure with a prosthetic patch.

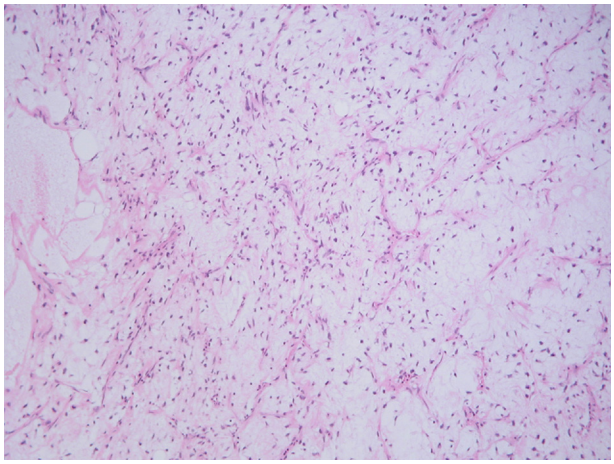

Fig. 5 – Microscopic findings (Institute of Pathology, Teaching Hospital in Motol), hematoxylin and eosin stain, magnification 100×. The tumor contains relatively small amount of cells that are oval, slightly elongated, or star shaped embedded in an abundant matrix with network of thin capillaries. Cells containing vacuoles and resembling lipoblasts are present.

the left side of the heart cause dyspnea, chest pain, or syncope due to left ventricular outflow obstruction. Also neurological symptoms such as movement disorders, vision disorders, and sensory symptoms can be seen [5,6,7].

In our patient, histological examination demonstrated a tumor surrounded with myocardium and partially with a thin fibrous capsule. The tumor contained relatively small amount of cells that were oval to slightly elongated or star shaped embedded in an abundant myxoid matrix (Fig. 5). No mitotic activity was found. Proliferation rate assessed by expression of the antigen Ki-67 was about 2%. The microscopic findings were similar as in the previous histological examinations of specimens from the left popliteal area and right axilla. It is of note that both myxoid and round cell liposarcoma are characterized by chromosomal translocations – fusion of the gene CHOP on the chromosome 12p 13 and the gene FUS on the chromosome 16q 11. These translocations were also detected in our case. The same chromosomal translocations were also retrospectively found in the previous specimen from the right axilla. Those findings confirmed unequivocally that the nodule in the septum in our patient was a metastasis of myxoid liposarcoma.

Diagnostic workup is based on medical history and physical examination complemented with noninvasive imaging techniques. The most important imaging methods are CT angiography and magnetic resonance imaging, which are able to characterize in detail pathological tissue. Both methods bring important information on tumor homogeneity, presence of adipose cells and calcification, percentage of necrotic or cystic components, tumor vascularization, size, location and relation to surrounding structures [3,9]. In our case, magnetic resonance imaging described size and location of the tumor and also its characteristics reminding myxoma (Fig. 3).

FDG-PET or PET/CT is increasingly used in clinical staging of most tumors. This method enables detection of metabolic activity of viable neoplastic tissue based on increased

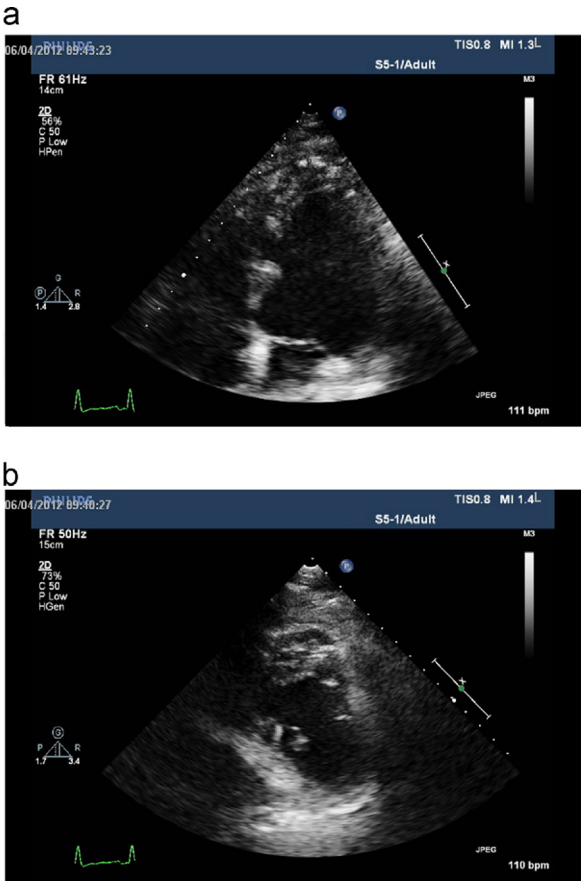

Fig. 6 – Echocardiography after tumor excision – (A) apical for chamber view and (B) short axis view.

glycolysis in tumor cells. It significantly helps to assess clinical stage of the disease, extent of the metabolically active tumor compartment in large tumors, and prognosis. In case of sarcomas, including myxoid sarcoma, this method is more suitable for grading. In our patient, the CT component of the CT/PET scan revealed accidentally a nodule in the interventricular septum, and the negative PET component indicated rather low grade tumor (Fig. 1). Postoperative examination still has not shown any signs of tumor recurrence [11]. The prognosis of the disease is dependent on several factors such as the size of the primary tumor, its location, histology, and the presence of metastases [12]. The primary treatment of liposarcoma is radical resection with adequate margin of normal tissue [5,13]. Adjuvant chemotherapy is not recommended as standard treatment, but it can be considered in individual cases when the tumor is very large or not removed completely, or risk factors are present. Systemic chemotherapy is indicated in generalized disease. The first choice treatment is doxorubicin with ifosfamide; the second choice is trabectedin, ifosfamide, or tyrosine kinase inhibitor pazopanib. Dacarbazine represents further treatment option in selected pretreated patients. When anthracyclines are administered, initial ejection fraction of the left ventricle needs to be established, and the cumulative dose, which represents the limiting factor of doxorubicin therapy continuation, has to be monitored [6]. The goal of radiation therapy is reduction of local recurrence in patients with large tumors and non-cardiac

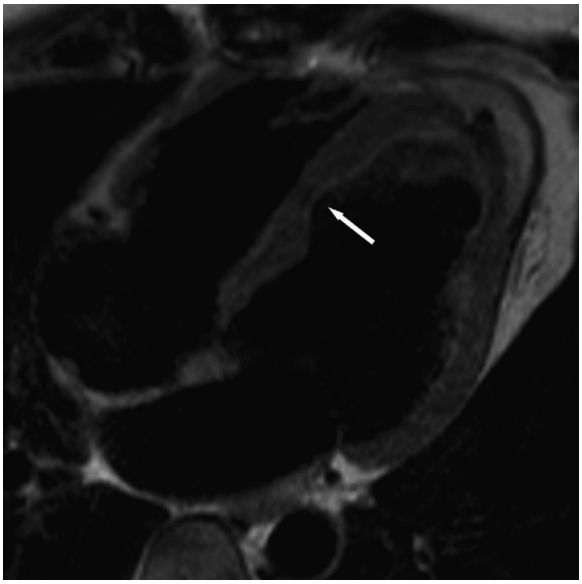

Fig. 7 – Magnetic resonance imaging: after excision of the tumor in the interventricular septum (native 2D T2-weighted TSE imaging).

surgical excisions [2]. Contrary to other more often seen sarcomas of soft tissues, myxoid and round cell form of liposarcoma are highly radiosensitive [4,5,13]. However, considering the fact that the heart should not be treated with radiation, and the discussed tumor has low chemosensitivity, surgery is the only option in such cases. Our patient was also treated solely surgically. No radiation therapy or chemotherapy has been indicated in this case since the tumor had very low proliferation activity, and no signs of recurrence have been identified so far.

Conclusion

Myxoid liposarcoma is the second most common type of soft tissue sarcoma. Its main features are long disease course and multiple recurrences. Diagnostic workup is based on noninvasive imaging techniques, particularly magnetic resonance imaging. Surgical resection is the treatment of choice, and is sometimes accompanied by radiotherapy. The prognosis of the disease is uncertain.

Conflict of interest

None to declare.

Funding body

No financial support.

Ethical statement

The research was done according to ethical standards.

Informed consent

The patient agreed with publication of this article, and signed written informed consent.

Appendix A. Supplementary data

Supplementary material related to this article can be found, in the online version, at http://dx.doi.org/10.1016/j.crvasa.2013.07.004.

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