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CASE REPORT

Extensive left ventricular, pulmonary artery, and pericardial metastasis from myxoid liposarcoma 16 years after the initial detection of the primary tumour: a case report and review of the literature

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Abstract

Liposarcoma is regarded as the second most common soft tissue malignant tumour. Metastasis of liposarcoma to the heart is very rare, and to date, less than 40 cases have been reported in the literature. We report a 46 year-old male with myxoid liposarcoma of the lower extremity who developed extensive metastasis to the left ventricle, pulmonary artery, and pericardium. The patient presented with acute symptoms of dyspnea 16 years after surgical resection of the primary tumour, and his dyspnea progressed to cardio-respiratory arrest within the first few days of hospital admission.

Keywords: myxoid liposarcoma, cardiac metastasis

INTRODUCTION

Liposarcoma is the second most common malignant soft tissue tumour after malignant fibrous histiocytoma. The main sites of the primary tumour are lower extremity and retroperitoneum. The most common site of metastasis are lungs, retroperitoneum, abdominal cavity and chest wall.[1] Cardiac metastasis is extremely rare, and to date, less than 40 such cases have been reported in English-language literature. The antemortem diagnosis of metastatic liposarcoma of heart and pericardium is difficult to make, mainly due to the indolent course of the lesion and also the fact that small lesions are mostly asymptomatic.[2] Herein we present a rare case of metastatic myxoid-type liposarcoma involving left ventricular, pulmonary artery, and pericardium 16 years after the initial detection of the primary tumour.

CASE REPORT

The patient was a 46 year-old male with history of lower limb liposarcoma 16 years prior to the current admission, when he was 30 years of age. The mass was reported as "myxoid liposarcoma", and it was radically resected with free surgical margins in a hospital at the time of diagnosis. He had no specific complains since then. Sixteen years after the initial detection of the primary tumour, he presented with sudden-onset severe dyspnea and after 3 days, he was brought to our center. Sonography revealed bilateral pleural effusion and hypoechoic lesions in the mediastinum. Due to the poor condition of the patient during hospital course, CT-scan and other radiological examination were cancelled. Within a few hours of hospital admission, the patient developed cardio-respiratory arrest and resuscitation was not successful.

Autopsy findings

Post-mortem examination revealed a giant mass occupying left ventricle measuring 8 x 6 x 3 cm, and occlusion of the pulmonary artery by another mass measuring 4 x 2 x 1 cm, and extensive involvement of the pericardium. The tumour mass was resected and specimen sent for histopathological examination. Grossly, the borders were not apparent, the consistency was mixed gelatinous and rubbery. Microscopical examination showed neoplastic tissue composed of spindle cells and lipoblasts with characteristic cytoplasmic fat vacuoles, and the nucleus which was pushed aside to form a signet ring configuration. In some areas, tumour cells penetrated into the myocardium and were separated from each other by myocardial cells (Figs. 1, 2 and 3). The tumour tissue had a myxoid background with delicate network of

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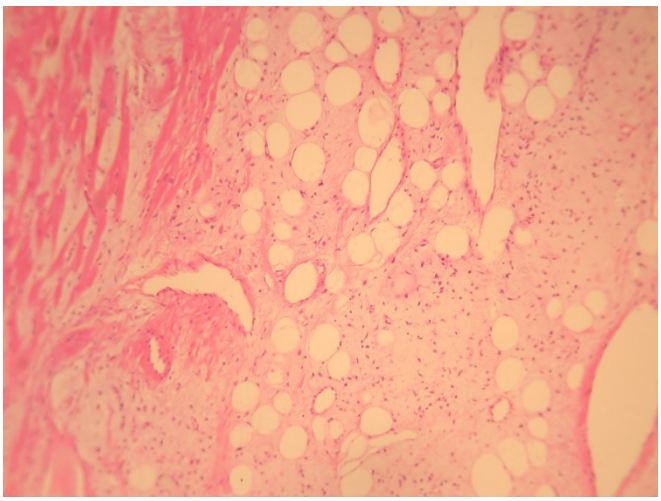

FIG. 1: Infiltration of myocardium by tumour cells. (Magnification 100X) H&E

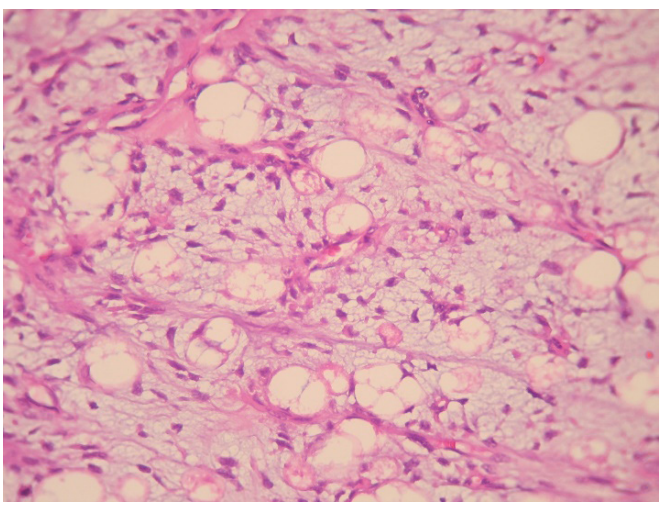

FIG. 2: Neoplastic tissue composed of spindle cells and lipoblasts exhibiting characteristic cytoplasmic fat vacuoles, and the nucleus pushed aside to form a signet ring configuration, in a myxoid background with delicate network of thin-walled vessels. (Magnification 200X) H&E

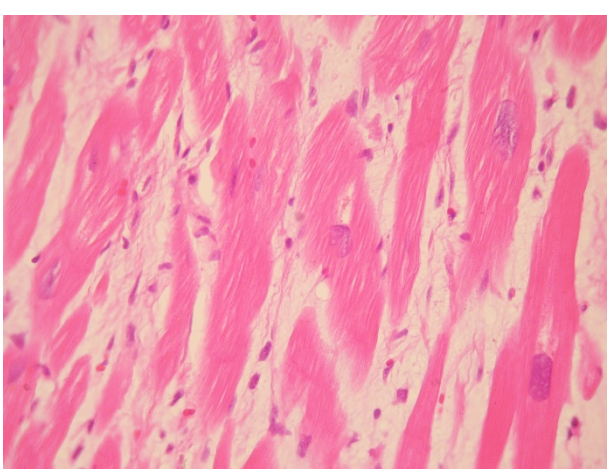

FIG. 3: Tumour cells penetrated into the myocardium and cardiac muscle cells were separated from each other by tumour cells. (Magnification 400X) H&E

thin-walled vessels.

DISCUSSION

Metastatic cardiac liposarcoma is a rare condition. The first reported case was by Scott and Garvin in 1938, in an article regarding tumours of heart and pericardium. Tong *et al* in 1968 specifically described a case of myxoid liposarcoma with left ventricular metastasis, and his treatment response to radiotherapy.[2] Since then only less than 40 cases of metastatic cardiac liposarcoma have been reported in English language literature (Table 1).

TABLE 1: Reported cases of metastatic cardiac liposarcoma

Author	Age	Sex	Primary site	Interval	Symptom	Metastasis	Year
Tong *et al*[2]	35	M	Left upper thigh	7 years	Progressive dyspnea	Left ventricle	1968
Godwin *et al*[9]	59	M	Right thigh	25 years	Asymptomatic systolic murmur	Right ventricle, pericardium	1981
Mavroudis *et al*[10]	59	M	Thigh	25 years	Congestive heart failure	Right ventricle	1981
Ravikumar *et al*[6]	79	M	Thigh	25 years	Congestive heart failure	Left ventricle, pericardium	1983
Pizzarello *et al*[11]	61	M	Not reported	20 years	Recurrent metastasis, friction rub	Right atrium and right ventricle, pericardium and pleura	1985
Lagrange *et al*[12]	46	M	Left thigh	7 years	Right-sided Congestive heart failure	Right ventricle	1986
Bartels *et al*[13]	64	M	Left scapular area	3 years	Right-sided Congestive heart failure	Right ventricular apex	1988
Ozoux *et al*[14]	60	F	Thigh	17 years	Murmur	Left ventricle	1988
Schrem *et al*[15]	41	M	Left knee	7 years	Superior vena cava syndrome	Right atrium	1990
Maloisel *et al*[16]	28	F	Abdomen	4 years	Lower extremity edema	Embolization of the foramen ovale	1991
Langlard *et al*[17]	54	F	Thigh	5 years	Right-sided heart failure	Right atrium, right ventricle	1992
Oshima *et al*[7]	37	M	Left thigh	5 years	Not provided	Left ventricle	1993
Papa *et al*[18]	45	M	Retroperitoneal	15 years	Dyspnea	Left ventricle	1994
Hatton *et al*[19]	39	M	Thigh	4.5 years	Chest pain	Pericardium	1997
Sugiyama *et al*[20]	61	F	Right knee	11 years	Congestive heart failure	Right ventricle	2000
Gacem *et al*[21]	42	M	Right inguinal	15 years	Friction rub	Pericardium	2000
Ng *et al*[22]	45	M	Thigh	3 years	Arrhythmia	Interventricular septum	2001
Lee *et al*[23]	53	F	Thigh	5 years	Biventricular failure and constrictive hemodynamics	Pericardium	2002
Wong *et al*[3]	54	M	Left chest wall	7 years	Dyspnea	Right ventricle	2002
Fairman *et al*[24]	56	F	Thigh	12 years	Angina and syncope	Left ventricle	2004
Kono *et al*[25]	60	M	Right foot	13 years	Superior vena cava syndrome	Superior vena cava (SVC), right atrium, and right ventricle	2005
Aoyama *et al*[26]	63	F	Left thigh	1 year	Cough and dyspnea	Pericardium	2005
Matsumoto *et al*[27]	55	M	Thigh	2.5 years	Dyspnea	Right ventricle, pericardium, pulmonary artery	2007
Chughtai *et al*[28]	46	M	Left shoulder	3 years	Dyspnea and fatigue	Right ventricle	2007
Komoda *et al*[29]	52	M	Left thigh	17 years	Dyspnea	Right atrium, right ventricle, atrioventricular sulcus	2009
Dogan *et al*[30]	54	F	Left thigh	4 years	Dyspnea	Left atrium and left upper pulmonary vein	2011
Lazopoulos *et al*[31]	63	M	Thigh	13 years	Congestive heart failure	Intracardial and pericardium	2011
Markovic *et al*[8]	45	F	Thigh	5 years	Chest pain	Pericardium	2012

Author	Age	Sex	Primary site	Interval	Symptom	Metastasis	Year
Fernández-Golfín *et al*[32]	68	M	Lower right limb	Not provided	Right lower extremity	Pericardium	2012
Pino *et al*[33]	36	M	Left shoulder	2 years	Dyspnea	Left atrium and left upper pulmonary vein	2013
Pino *et al*[33]	35	M	Lung	3 years	Congestive heart failure	Right atrium	2013
Pino *et al*[33]	23	M	Popliteal fossa	3 years	Right-sided heart failure	Pericardium	2013
Mottahedi *et al*[4]	50	F	Left knee	4 years	Dyspnea	Right atrium and right ventricle	2013
Virtová *et al*[34]	36	M	Left popliteal area	5 years	Incidentally	Interventricular septum	2014
Xu *et al*[5]	60	M	Left thigh	20 years	Dyspnea	Right ventricle and pulmonary artery	2014

The time intervals between the initial detection of primary tumour and cardiac metastasis ranged from 1 year to 25 years (mean = 9 years). Most reported cases were males (71%) with a mean age of 50 years. Patients with cardiac metastasis are asymptomatic for many years and sometimes the tumour is an incidental post-mortem finding. The presentation of cardiac metastasis depends on which part of the heart is involved. The most common presentations were dyspnea and congestive heart failure.

Histologically, liposarcoma has four subtypes: well-differentiated, dedifferentiated, myxoid/round cell, and pleomorphic. The myxoid/round cell is the most frequent subtype.[1] It is important to specify the subtype of the tumour, since myxoid liposarcoma has an expansive growth nature with less infiltrative behavior and usually causing obstruction at the site of metastasis and leads to an aggressive course of rapidly progressing heart failure in many reported cases.[3]

Most of the cases are diagnosed post-mortem, but in cases with early detection of cardiac metastasis both radiotherapy and surgical resection have been reported to be successful.[3-6] In non-resectable cases radiotherapy helps to reduce disease progression to heart failure.[2]

There are many imaging techniques available for diagnosis of cardiac metastasis. Two dimensional echocardiography can help to detect pericardial effusion, intracavitary masses, and ventricular function, and it is the first imaging techniques for diagnosis of such lesions. CT is usually used for cancer staging, and MRI with a better contrast has advantage for clarifying anatomic details. In some patients PET examination is reported to be the most effective diagnostic procedure, since, PET is a unique diagnostic tool for detecting infiltration of tumour into myometrium.[4,7,8]

In this article we emphasize on regular check-up of patients diagnosed with liposarcoma including examination for cardiac lesions even long after resection of the primary lesion.

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