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Short communication

Primary pleomorphic malignant fibrous histiocytoma of the heart

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ABSTRACT

Primary pleomorphic malignant fibrous histiocytoma of the heart is rare. The present study was performed to study the clinical and pathological features of the disease. We describe two rare cases of primary cardiac malignant fibrous histiocytoma and review the published individual data of the patients. Both patients complained of dyspnea, and underwent palliative tumor resection. However, they died several months after surgery. A thorough literature review with clinical presentations, diagnostic features, treatment, and outcomes was done. We have for the first time analyzed the factors related to the survival of malignant fibrous histiocytoma. It is usually difficult to make an appropriate preoperative diagnosis. Despite complete surgical resection and aggressive chemotherapy and radiotherapy, the prognosis is still poor.

Introduction

Primary cardiac sarcomas are rare. Angiosarcoma presents the most common type of sarcoma, and the second most common sarcoma is malignant fibrous histiocytoma (MFH) (Burke et al., 2004). According to the World Health Organization (WHO) classifications of soft tissue tumors released in 2002, the term pleomorphic MFH is synonymous with undifferentiated pleomorphic sarcoma, which manifests a broad range of histological appearances with three-types described: storiform-pleomorphic, giant cell, and inflammatory. Myxofibrosarcoma (formerly defined as myxoid variant or myxoid MFH) remains a distinctive and discrete entity (Fletcher et al., 2002).

To the best of our knowledge, there have been only 21 cases reported in English in PubMed since 2002 that could be categorized as primary cardiac MFH when applying the latest released classification. Although previous reports have documented a series of cases, quite a few could not meet the criteria "primary" or be categorized as other malignancies according to the definition of 2002. In the present study we report two cases of primary cardiac MFH and have carried out a literature review with 21 reported cases.

Methods

Two cases of primary cardiac MFH were identified by searching our surgical pathology database during the ten-year period between 2002 through 2012. The medical records were reviewed with clinical presentations, methods of diagnosis, and management. Follow-up was made by telephone.

We suggest that primary cardiac MFH should meet the following criteria: (a) be located in the heart chambers, myocardium or pericardium; (b) exclude metastasis: absence of history of MFH, with no MFH in other locations at the time of diagnosis; (c) morphological features should conform to the description of WHO classification. Okamoto et al. (2001) and Toda et al. (2002) have reviewed cases of cardiac MFH in the literature published before 2002, however, some cases do not conform with the above criteria. We undertook a PubMed search using the terms "cardiac malignant fibrous histiocytoma", "malignant fibrous histiocytoma AND heart", "cardiac malignant fibrohistiocytoma", and "malignant fibrohistiocytoma AND heart". Studies cited by each of these reports were also evaluated to identify any cases that may have been missed. In order to make a more accurate identification, we have carefully re-reviewed all available descriptions and figures. A total of 21 instances were included in the present study (Table 1).

All patient data were collected and recorded using Microsoft Excel (Microsoft, Redmond, WA, USA). Statistical analyses and graphing were performed using Prism 5.0 (Graphpad Software, La Jolla, CA, USA). For analyses of factors affecting survival times,

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Table 1
Reported cases of primary cardiac malignant fibrous histiocytoma.

Author/year	Age (year)/sex	Symptoms	Location	Size (cm)	Surgery	Chemo/radio therapy	Follow-up/outcome
Toda et al. (2002)	16/M	Dyspnea	LA, PV	10 × 7.5 × 5	PR	N/N	9 months/uneventful
Akhter et al. (2004)	32/M	Dyspnea, fatigue	RV	6.2 × 3.5 × 8.5	OCT	Y/N	8 years/lung metastasis, RA recurrence, uneventful after the third surgery
Dorobantu et al. (2005)	53/F	Dyspnea, chest pain, cyanosis, and fatigue	LA, LV, PC	2.6 × 2.1, 2.4 × 3, 1.7 × 3.8	NA	Y/N	6 months/died
Novelli et al. (2005)	80/F	Dry cough	LA, LV	7 × 6 × 5	NA	Y/N	15 months/died
Uezu et al. (2005)	16/F	Headache, vertigo, and headedness	LA	NA	PR	NA	7 months/died
Kim et al. (2006)	34/F	Dyspnea	RA, TV	7 × 6 × 4.5, 2.7 × 2.2 × 2	NA	Y/N	4 months/recurrence
Chung et al. (2007)	39/M	Fever, chills, watery diarrhea, abdominal pain, and lethargy	LV	2.6 × 1.8 × 1.8	NA	N/N	1 month/uneventful
Milicic et al. (2007)	58/F	Congestive heart failure, pleural effusion	LA	4 × 2	NA	Y/N	3 years/recurred and distal metastasis
Skarysz et al. (2007)	72/M	Dyspnea	RA, RV	5.4 × 2.2	PR	N/N	39 months/uneventful
Sato et al. (2007)	72/M	Appetite loss	LA	NA	PR	N/N	22 months/died
Matsukuma et al. (2008)	72/F	Dyspnea	PC	10 × 10 × 5	PR	Y/N	2 months/died
Yaliniz et al. (2008)	28/M	Dyspnea, fatigue, and epigastric pain.	LV, RV, PA	NA	PR	N/Y	NA/NA
Inoue et al. (2009)	15/M	Dyspnea	LA, PV	10	NA	Y/Y	4.5 years/died
Iwa et al. (2009)	31/F	Fatigue, back pain, and nausea	RV	6 × 3 × 2	NA	NA	NA/died
Balaceanu et al. (2010)	22/F	Headedness, nausea, vomiting, loss of awareness in orthostatism	RV	5	NA	Y/N	NA/NA
Fontana et al. (2010)	55/F	Ictus cerebri	MV	8 × 2	CR	Y/Y	NA/NA
Abdullgaffar (2010)	30/M	Fever, shortness of breath, fatigue, abdominal pain, bilateral lower limb swelling	RA, IVC	6 × 4.5	PR	N/N	15 days/died
Sawaed et al. (2010)	22/M	Dyspnea	LA	3.4 × 3.4 × 3.5	NA	Y/Y	20 months/recurrence, uneventful after the second surgery
Dorobantu et al. (2011)	22/F	Abdominal pain, nausea, and consciousness loss	RV	5.0 × 4.5 × 3.5	CR	Y/N	1.5 years/uneventful
Lee et al. (2011)	42/M	Chest discomfort, palpitation, and dyspnea	LV	3.0 × 2.5	OCT	N/N	11 months/died
Rashidi et al. (2011)	59/M	Asymnostic	LA	NA	NA	Y/N	NA/NA

Abbreviations; M, male; F, female; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; PA, pulmonary artery; PV, pulmonary vein; TV, tricuspid valve; MV, mitral valve; PC, pericardium; NA, not available; PR, partial resection; IVC, inferior vena cava; C, complete resection; OCT, orthotopic cardiac transplantation; Y, yes; N, no.

patients were included only if they were diagnosed pre-mortem and had reported survival data.

Results

Case 1

A 45-year-old woman was admitted to our hospital with dyspnea for 20 days. She complained of pink bubble sputum cough and difficulty in lying down to sleep at night. On admission, her blood pressure was 60/40 mmHg. On physical examination, she showed acute facial features, somnolent and peripheral edema. Low breath sound and moist rales were recorded. The apex impulse was unclear, and a slight diastolic rumbling murmur and enhanced second sound of pulmonary area were heard. Transthoracic echocardiography revealed a large, partially mobile left atrial tumor mass, about 3.5 cm × 2.8 cm in size, attached to the interatrial septum with a short stalk. The pulmonary arterial pressure was calculated to be about 70 mmHg. Blood tests revealed a high white cell count ($11.69 \times 10^9\ L^{-1}$), elevated alanine aminotransferase (662 U/L), aspartate aminotransferase (660 U/L), blood urea nitrogen (8.4 mmol/L). After a preliminary diagnosis of left atrial myxoma, the patient subsequently underwent open heart surgery under cardiopulmonary by-pass. An oval mass measuring 2 cm × 2 cm was observed in the top of right atrium. A further two masses (5 cm × 3 cm × 3 cm, 5 cm × 7 cm × 8 cm) originating from the anterior leaflet of the mitral valve, were protruding into the left atrium. The tumors were partially resected together with the attached endocardium. Histological examination revealed it was mainly composed of fusiform fibroblast-like cells, forming a storiform pattern in some areas (Fig. 1A and B). Multinuclear giant cells could be observed. The tumor cells were immunoreactive for CD68 and α-1-antichymotrypsin (Fig. 1C and D). A diagnosis of pleomorphic MFH was made. The patient was discharged after surgery without further chemotherapy or radiotherapy, however, she died of congestive heart