Malignant Fibrous Histiocytoma In The Heart; An Autopsy Case

S Furukawa, L Wingenfeld, I Sakaguchi, T Nakagawa, A Takaya, S Morita, S Yamasaki, K Nishi

Citation

S Furukawa, L Wingenfeld, I Sakaguchi, T Nakagawa, A Takaya, S Morita, S Yamasaki, K Nishi. *Malignant Fibrous Histiocytoma In The Heart; An Autopsy Case*. The Internet Journal of Cardiology. 2012 Volume 10 Number 3.

Abstract

Malignant fibrous histiocytoma (MFH) in the heart is rare. At autopsy, MFH of the left atrium was diagnosed in a 36 year old woman. The mass involved the left left ventricular wall. Histopathologic examination revealed that the tumor consisted of malignant cells with marked atypia. Immunohistochemistry showed a diffuse immunoreaction for vimentin. The histopathologic diagnosis was MFH. We present a case of a 36-year-old woman with histologically confirmed MFH in the left atrium.

INTRODUCTION

Malignant fibrous histiocytoma (MFH) is sarcoma that typically occurs in the extremities, the torso, and the the retroperitoneum [1].

This tumor constitutes less than 3% of primary cardiac tumors. A primary heart tumor is a rare disease, the incidence of which has been reported in only 0.0017% of autopsies [2]. MFH is the most common soft-tissue sarcoma in adults [3]. MFH has been rarely reported as a primary tumor occurring in the heart. It is usually diagnosed when it is locally aggressive or has already metastasized.

CASE REPORT

A 36-year-old woman was found lying on the floor with vomit in her room. She had no past history. At autopsy, the left atrium was opened and a sessile multinodular tumor was found to be attached to the posterior and lateral walls of the atrium. (Figure.1) All parts of the visible tumor were excised. The resected tumor (5×4cm) was yellowish. The tumor was removed and on histological examination showed the characteristic mixture of spindle cells arranged in a storiform pattern, polygonal cells resembling histiocytes, and malignant giant cells, which are all features of MFH. (Figure.2a) Immunohistochemically, the tumor cells stained positive for vimentin. (Figure.2b) Staining for cytokeratin, S-100 protein, and myogloblin were completely negative. The tumor was finally diagnosed as ordinary MFH.

Figure 1

Figure.1 : A sessile tumor measuring 5×4cm was found attached to the left atrium. (Autopsy)

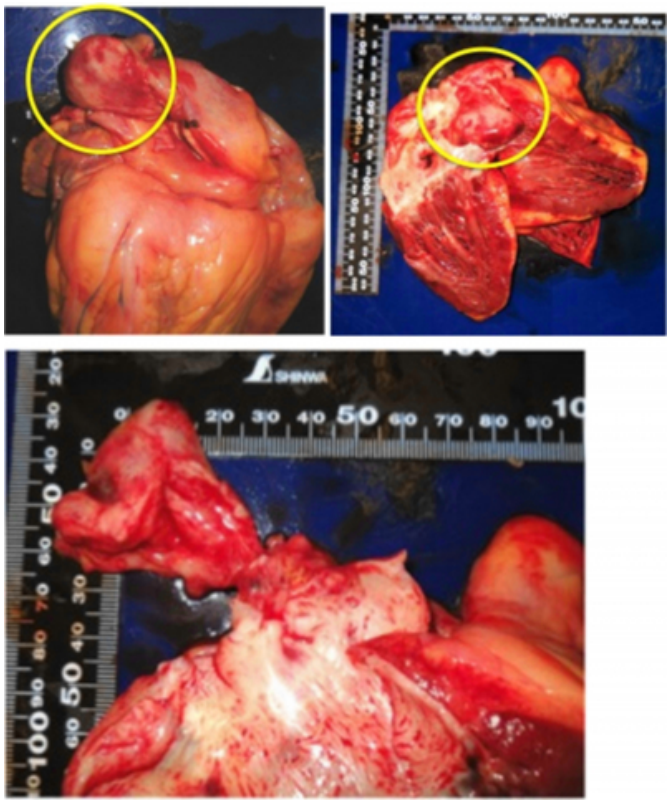


Figure.2 : Histological appearance of MFH showing the mixture of spindle shaped fibroblasts arranged in a storiform pattern, polygonal pleomorphic histiocytes, and malignant giant cells. (2a; Haematoxylin and eosin, 2b; Vimentin, ×400

original magnification)

Figure 2

Figure.2a

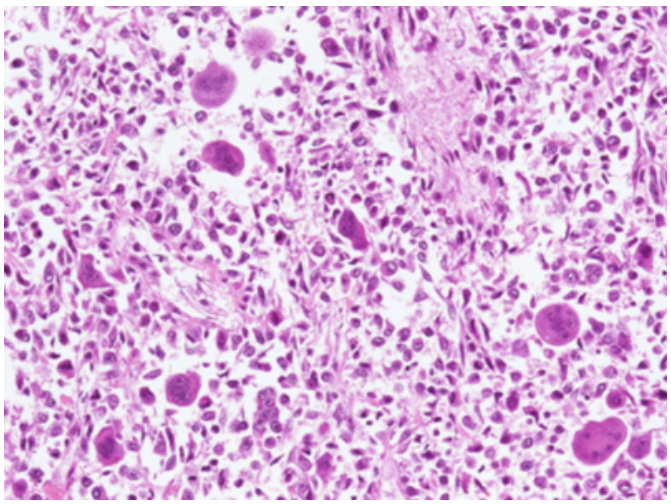
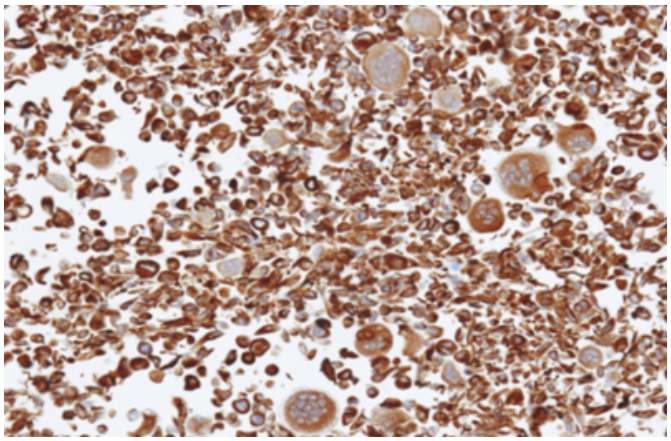


Figure 3

Figure.2b



DISCUSSION

Tumors of the heart are rare. They were first described by Yater [4].Among malignant cardiac tumors, the most frequent one is angiosarcoma. Primary MFH is the second most common primary cardiac sarcoma. When it is small, no clinical manifestations are evident [5]. As the tumor grows, symptoms can vary according to its location. Diagnosis needs confirmation with immunohistochemical and ultrastructural examination [6]. Histologically, MFH is a variously shaped and multilobulated mass, sessile or pedunculated. It is a tumor of the fibroblasts with giant cells and nuclear and cytoplasmic atypical findings. Out of 36 cases in the literature which were histologically examined in detail, 28 cases were storiform variant of ordinary MFH (3 cases), myxoid MFH (4 cases), and giant cell MFH (1 case) were also reported in the heart [7] Thus, the histological features of cardiac MFH are not different from those of extra cardiac MFH which were well described by Weiss and Enzinger.

MFH is a relatively recently recognized diagnostic entity, characterized histologically by the mixture of spindle cells arranged in a storiform pattern, polygonal cells resembling histiocytes, and malignant giant cells. This report presents a rare autopsy case of cardiac tumor MFH.

References

1. Guvendik L, Ross JK, Marshall RJ. Primary aortic malignant fibrous hisiocyoma: a successfully treated case by surgical excision. Ann Thorac Surg., 42 (5): 578-80, 1986.
2. Straus R, Merliss R. Primary tumor of the heart. Arch Pathol Lab Med., 39: 74-78, 1945.
3. Weiss SW, Enzinger FM. Malignant fibrous histiocytoma: an analysis of 200 cases. Cancer., 41: 2250-2266, 1978.
4. Yater WM. Tumors of the heart and pericardium: pathology, symptomatology and report of 9 cases. Arch Intern Med., 48: 627, 1931.
5. Donsbeck AV, Ranchere D, Coindre JM, Le Gall F, Cordier JF, Loire R. Primary cardiac sarcomas: an immunohistochemical and grading study with long-term follow-up of 24 cases. Histopathology., 34: 295-304, 1999.
6. De Jong AS, Van Kessel-van Vark M, Albus-Lutter CE. Pleomorphic rhabdomyosarcoma in adults: immunohistochemistry as a tool for its diagnosis. Hum Pathol., 18: 298-303, 1987.
7. K Okamoto, S Kato, S Katsuki, Y Wada, Y Toyozumi, M Morimatsu, S Aoyagi, T Imaizumi. Malignant fibrous histiocytoma of the heart: case report and review of 46 cases in the literature. Int Med., 40 (12): 1222-6, 2001.

Author Information

Satoshi Furukawa
The Department Of Legal Medicine, Shiga University Of Medical Science

Lisa Wingenfeld
The Department Of Legal Medicine, Shiga University Of Medical Science

Ikuo Sakaguchi
The Department Of Legal Medicine, Shiga University Of Medical Science

Tokiko Nakagawa
The Department Of Legal Medicine, Shiga University Of Medical Science

Akari Takaya
The Department Of Legal Medicine, Shiga University Of Medical Science

Satomu Morita
The Department Of Legal Medicine, Shiga University Of Medical Science

Shigeru Yamasaki
The Department Of Legal Medicine, Shiga University Of Medical Science

Katsuji Nishi
The Department Of Legal Medicine, Shiga University Of Medical Science