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Editorial

Editorial: Primary cardiac malignant fibrous histiocytoma is a rare case of cardiac tumor

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Primary cardiac tumors are rare, with an incidence of 0.001–0.03% in previous autopsy series [1]. The majority of these tumors are benign myxomas, although a quarter of cases are malignant, most of these tumors are sarcomas.

In primary cardiac sarcomas, angiosarcoma is the most common type of sarcoma [2].

Malignant fibrous histiocytoma (MFH) is a type of sarcoma characterized by a high degree of malignant neoplasm. Its origin is uncertain and arises in both bone and soft tissue. Sarcomas, including MFH, comprise the majority of primary cardiac malignant tumors and have a poor prognosis.

The first reported case of cardiac MFH was published in 1978 by Shah et al. [3]. Subsequently, Okamoto et al. [4] reported a case of MFH that arose from the left atrium (LA) and reviewed an additional 46 previously reported cases. An analysis of those 47 cases revealed that 29 patients (62%) were females (mean age of 47.1 years) and cardiac MFH was found to occur most frequently in the LA and to usually attach to the posterior wall; non-cardiac MFH usually occurs in the limbs, trunk, retroperitoneum, bone, or head.

In this issue of *Journal of Cardiology Cases*, Kawarabayashi et al. [5] report a case of primary cardiac MFH (undifferentiated pleomorphic sarcoma). In their case, transthoracic echocardiography revealed cardiac tumors in the LA of a 53-year-old woman with a 3-month history of worsening dyspnea. The largest tumor was found to protrude through the mitral valve in diastole. Three of four tumors were resected during emergency surgery. However, the residual tumor extension into the left pulmonary vein could not be removed. Histological findings of the resected tumors, such as organized thrombus and myxomatous tissue changes, indicated that the tumors were benign at that time.

According to the World Health Organization 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 [6]. According to the main morphological characteristics of tumor cells, MFH can be grouped into four types [7,8].

First is the storiform-pleomorphic type, where the most prominent morphological features include the fact that the tumor cells are pleomorphic and have a spoke wheel-like structure, showing spindle cells arranged in spoke wheel shapes.

A large number of giant cells are stained with a strongly eosinophilic cytoplasm, and single or multiple irregular nuclei are present in the multishaped area.

The second type is the giant tumor cell type. The tumor is nodular and is formed by the fibroblasts, histiocytes, and spread osteoclast-type giant cells. Some tumors present as malignant, giant tumor cells with frequent mitotic activity. The tumors are usually hemorrhaging, exhibit necrosis, and sometimes present with bone-like tissue.

The third type is the inflammatory type. The most prominent feature of this type is a large number of foamy yellow tumor cells in various differentiations mixed with a large number of inflammatory cells, particularly neutrophilic granulocytes. Sometimes poorly differentiated tumor cells have little cytoplasm and large nuclei, and mitotic activity is usually visible.

The last type is the myxoid type, where the rare tumor cells are round- or star-shaped and scattered in the loose mucus matrix. Vacuoles can be seen within the cytoplasm of the tumor cells, which are similar to the lipoblastoma cells, but special staining shows acidic mucus in the vacuoles.

Wang et al. [9] reported two cases of cardiac MFH that reviewed an additional 21 previously reported cases in English in PubMed since 2002 through 2012 that could be categorized as primary cardiac MFH when applying the latest released classification. The tumor was often located in the left atrium (11/23), but it could involve any site, including all chambers, mitral or tricuspid valve, and pericardium. Just as with cardiac myxomas, cardiac MFH often manifested cardiorespiratory symptoms in which dyspnea was most common. Gastrointestinal symptoms, such as abdominal pain, diarrhea, appetite loss, and nausea, which arose in 7 patients, appeared more common than that of myxomas [10].

Surgery is the cornerstone of treatment for all soft-tissue sarcomas. The goal of surgery is to eradicate all of the disease in the affected area.

In Kawarabayashi's case, after 3 months emergency surgery, the patient underwent total resection for a small mass that developed on her right abdominal wall, which was revealed histologically to be

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MFH; additionally, the residual mass in the LA had enlarged progressively. After undergoing radiation therapy without further surgery, she died of cerebral bleeding 6 months after cardiac surgery. Postmortem examination revealed that the tumor in the LA was an MFH.

Although the case reported here included performing surgical tumor resections, the tumor featured rapid disease progression, subsequent relapse, and metastasis. There are few single case reports of successful chemotherapy and radiotherapy [11].

For MFH, the role of chemotherapy in treatment is not entirely clear [7]. Several clinical trials incorporating the chemotherapy drug doxorubicin have shown improved event-free survival without a major impact on overall survival [12]. Radiation clearly improves the incidence of local recurrence and has become an integral part of the treatment for MFH [13,14]. Some factors, however, contribute to the poor prognosis of MFH, including a lack of molecular targets for MFH treatment.

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