Clinical Case Report

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Primary atrial fibromyxosarcoma with multiple-system metastases

A case report

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Abstract

Rationale: Fibromyxosarcoma is common in head and neck, vessel, omentum, and reproductive system, with low-grade malignant behavior. However, primary atrial fibromyxosarcoma with highly malignant behavior is extremely rare.

Patient concerns: A 34-year-old female presented with oppression in the chest, short breath, and onset of headache as initial symptoms. The preoperative echocardiogram showed a medium-size echogenic mass close to the posterior leaflet of the mitral valve in the left atrium.

Diagnosis: Primary atrial fibromyxosarcoma with multiple-system metastases.

Interventions: The patient underwent surgery, and the tumor was removed completely. The diagnosis of left atrium fibromyxosarcoma was confirmed through postoperative histopathological examination. Positron emission tomography/computed tomography scan was performed, which revealed multiple metastases to left adnexa, bilateral adrenal glands, left iliacus, right lateral ventricle, and skeletal system.

Outcomes: The patient died of cerebral hernia caused by hemorrhage from the metastatic brain tumor, 30 days after the surgery, without receiving chemotherapy or radiotherapy.

Lessons: Cardiac fibromyxosarcoma is a rare primary malignant cardiac neoplasm, probably with systemic metastases. The possibility of malignancy should be considered as differential diagnosis for cardiac mass.

Abbreviations: MRI = magnetic resonance imaging, PET/CT = positron emission tomography/computed tomography, SUV = standardized uptake value.

Keywords: cardiac connective tissue, differential diagnosis', fibromyxosarcoma, heart neoplasms, metastases

1. Introduction

Cardiac primary malignancies are rare,[1,2] presented mainly as sarcoma of progressive onset, and a pathological diagnosis assisted by immunohistochemical staining is needed for definitive diagnosis. Fibromyxosarcoma is uncommon in heart, and usually manifests as low-grade malignancy. We report a case of left atrial fibromyxosarcoma featured with a highly malignant biological behavior, as evident from acute onset, multiple metastases, and short lifespan. The patient died of cerebral hernia led by hemorrhage of the metastatic brain tumors.

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2. Case report

The patient was a 34-year-old female of Han nationality. She was a company staff with no history of allergic diseases; no specific menstrual history, medical history, family history, or psychosocial history including comorbidities; and no relevant genetic information. In May 2016, she presented with oppression in the chest, short breath, inability to prostrate at night as exacerbated by exercise, and occasionally stimulating dry cough, without fever. chest pain, or phlegm. These symptoms were slightly relieved after she received treatment at a local hospital. On May 29, she suddenly had a headache, most severe in the occipital region, along with nausea, vomiting, and inaccurate visual sense of distance, without fainting, unconsciousness, or limb disorder. On May 30, she underwent echocardiogram (Fig. 1A), which revealed that the left atrium had a medium echogenic mass, with high possibility of myxoma. Moreover, mitral orifice was severely obstructed by the big left atrium. The patient also had pulmonary hypertension and mild tricuspid regurgitation. Brain contrast-enhanced magnetic resonance imaging (MRI) (Fig. 1B) showed a hemorrhagic focus in the right parietooccipital lobe, indicating a space-occupying lesion accompanied by apoplexy and a small intracranial ischemic focus. The preoperative diagnosis indicated a left atrium myxoma, with cardiac insufficiency (*New York Heart Association* class III), and intracranial space-occupying lesion coupled with cerebral hemorrhage and infarction. On June 3, the patient underwent resection of the left atrium myxoma and mitral valvuloplasty under general anesthesia with extracorporeal circulation by the Department of Cardiac Surgery in the First

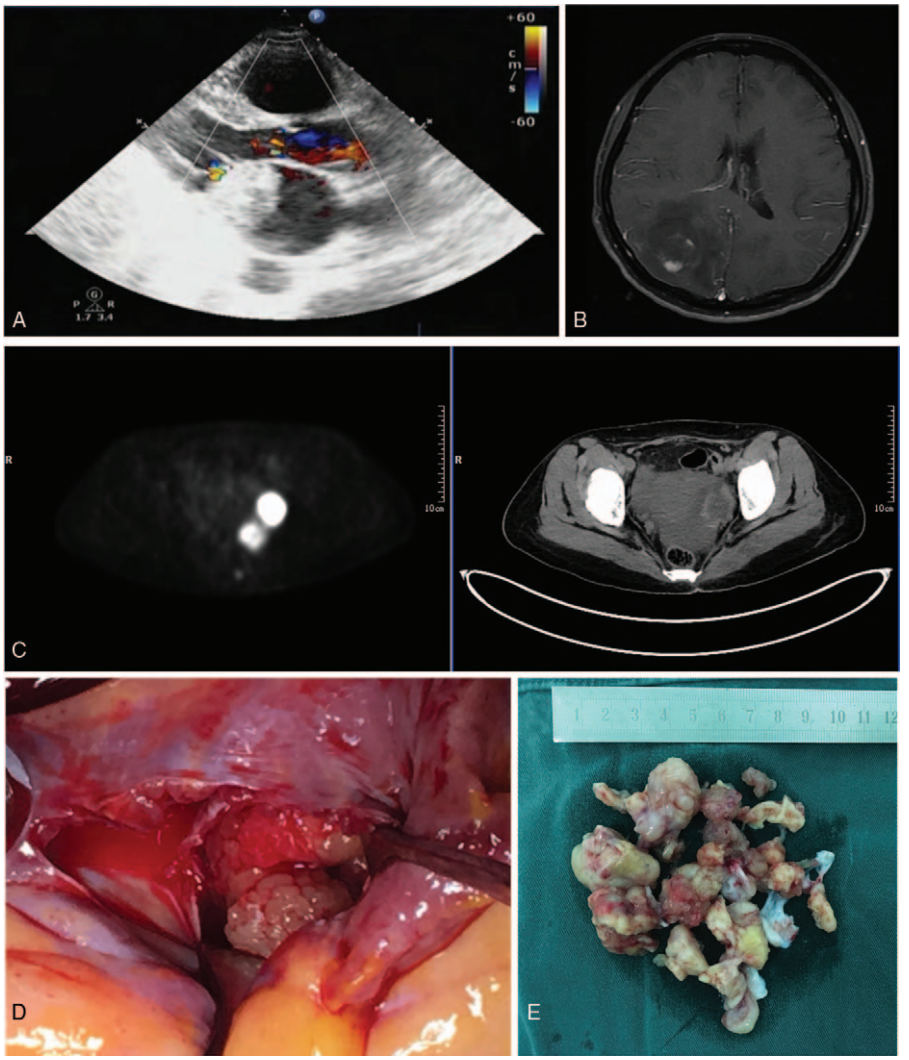

Figure 1. (A) Preoperative echocardiography: The mass was of irregular shape, lobulated with obvious deformation, closely related to posterior leaflets of the mitral valve. (B) Preoperative contrast-enhanced MRI scan (2016-5-31): The enhanced scan revealed small strip-sheet enhancement, with edema surrounding the lesion, and the right ventricle was compressed and deformed. (C) PET/CT (2016-6-27) revealed 2 massive foci of increased FDG uptake were in the left adnexal area (SUVmax = 16.4). Low-density foci surrounded by lesions of increased density were found in the corresponding site on CT. (D) The intraoperative image revealed that the pedicle was broad-based located on the posterior wall of the left atrium and posterior leaflets of the mitral valve P2 and P3. (E) The gross appearance was a heap of gray and yellow broken tissues, $5 \times 4 \times 3\,cm^3$ in size, tough, delicate, and rich in mucus. MRI = magnetic resonance imaging, PET/CT = positron emission tomography/computed tomography, FDG = fluorodeoxyglucose, SUV = standardized uptake value.

Hospital of China Medical University. The right atrium and interatrial septum were incised during the surgery, presenting a big tumor in the whole left atrium (Fig. 1D, E). The posterior leaflet of the mitral valve was found to be poorly developed due to heavy oncothlipsis with backflow after carefully separating and completely excising the tumor and pedicle, and flushing with large amounts of saline. Postoperative histopathological diagnosis indicated left atrium sarcoma, and immunohistochemistry analysis showed fibromyxosarcoma (Fig. 2). Light microscopy revealed spindle cell proliferation with myxoid stroma, hyperchromatic nuclei, and atypia. The immunohistochemistry results were actin (SM −), CD34 (blood vessel +), CK (+), D2–40 (focal +), desmin (−), Ki-67 (30%), myogenin (scattered weak +), S-100 (weak +), vimentin (+), EMA (−), WT1 (+), and CD68 (partial +). On June 27, the positron emission tomography/computed tomography (PET/CT) scan (24 days after the surgery) (Fig. 1C) revealed a low-density lump in the left adnexal area with a hypermetabolic activity; thus, malignant disease was considered. The bilateral adrenal gland was hypermetabolic, and the skeleton had multiple hypermetabolic foci with partial destruction of bones. Also, the left iliacus had a low-density mass with a hypermetabolic activity; following these findings, malignant disease metastases were considered. The posterior crus side of the left lateral ventricle had a nodule of increased density without hypermetabolic activity, while the posterior crus side of the right lateral ventricle had an irregular focus of increased density surrounded by a low-density focus; these findings were also indicative to malignant disease metastases.

On June 27 (24 days after the surgery), the patient presented with aggravated headache, nausea, vomiting, and inaccurate visual sense of distance accompanied by ptosis. Plain CT scan of the head showed hemorrhagic foci of the bilateral parietooccipital lobes; the right hemorrhagic focus decreased obviously, but the left side revealed a new hemorrhagic focus. Moreover,

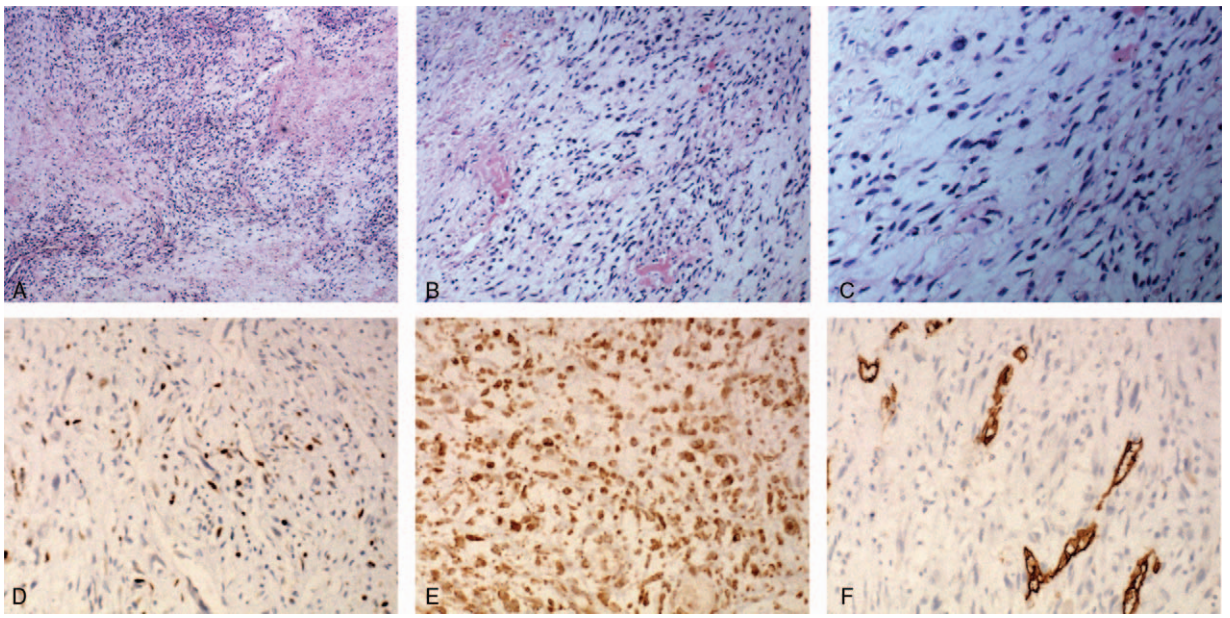

Figure 2. Histological image showed spindle cell proliferation with myxoid stroma, hyperchromatic nuclei, and atypia (A, ×40, B, ×100, C, ×200, H&E). Immunohistochemical staining shows Ki-67 (30%) (D, ×200), vimentin (+) (E, ×200), and CD34 (blood vessel +) (F, ×200).

no positive outcomes were observed following decreasing intracranial pressure treatment. On July 1 (28 days after the surgery), the patient had sudden unconsciousness, slow pupil reaction to light, and no response to tickling. On July 3 (30 days after the surgery), she died.

The local institutional review board approved this case report.

3. Discussion

Fibromyxosarcoma has been considered to be related to immune function disorder evoked by various viral infections; it may also be congenital[3] and induced by radiation.[4] In 1977, Hirano et al[5] evoked various stromal neoplasms, including fibromyxosarcoma, using the Moloney murine sarcoma virus. In 1987, Strayer and Leibowitz[6] discovered that the infection by malignant rabbit fibroma virus could cause severe immune function disorder, finally inducing deadly fibromyxosarcoma. This patient was HIV negative and without medical history of immunosuppressive agents. Therefore, no evidence is shown that the patient's tumorigenesis was associated with immune dysfunction.

Primary cardiac neoplasms are rare with the incidence rate lower than that of metastatic neoplasms. Except for atrial myxomas, the incidence rate is only 0.001% to 0.28% with 25% malignant neoplasms, of which 75% to 95% are sarcomas.[1,2] Neragi-Miandoab et al[7] retrospectively analyzed about 110 studies published during 1973 to 2006 and performed statistical analysis of 117 primary cardiac malignant neoplasms of adults. Malignant neoplasms with the first 5 highest incidence rates were angiosarcoma (33%), rhabdomyosarcoma (21%), mesothelioma (16%), fibrosarcoma (11%), and lymphoma (6%).[7] Zajonz et al[8] proposed that the fibromyxosarcoma generated from the fibroblasts in the heart accounted for about 10% of all malignant cardiac neoplasms.

A total of 37 fibromyxosarcoma case reports were collected by searching the PubMed database. Fibromyxosarcoma, reported in 1932 for the first time, may occur in various parts of the body as follows: commonly in head and neck (8, 21.6%[4,9–15]), vessel, omentum (11, 29.7%),[16–26] and reproductive system (7, 18.9%),[27–33] and seldom in parenchymal organ.[34] So far, only 2 cardiac fibromyxosarcoma cases have been reported in the literature. Dallocchio et al[29] first reported a case of fibromyxosarcoma located in the heart and ovary, in 1970. Zajonz et al[8] reported a case in which a 58-year-old female patient was diagnosed with lower-level primary cardiac fibromyxosarcoma accompanied by multiple pulmonary and bone metastases, who survived for more than 6 years through surgery and chemotherapy. This is the third case report of fibromyosarcoma of the primary heart, but the novelty of this case is the system metastases and consequently short survival.

The limitations of this case report were, first, due to epidemiological quantities, there were no denominator to assess rate; second, causal inference is not possible, that is, the hypothesis that immunodeficiency defects were the cause of cardiac fibromyxosarcoma cannot be rejected or accepted; third, bias, publication bias favored the novelty and atypicality, but recall bias might limit the complement of information. Despite the limitation, this report could hopefully assist in accumulating scientific data about rare disorders and serving as an educational tool.

Cardiac fibromyxosarcoma is a rare primary malignant cardiac neoplasm. Still the potential clue for differential diagnosis from cardiac myxoma might be the broad-based pedicle appears in the ultrasonic cardiogram. For another, the characteristic of this case was systemic metastases. Therefore, we present this case report to call attention to the possibility of primary cardiac malignancy with metastases for cardiac mass. Despite surgery is inevitable in a critical situation, a comprehensive treatment should be determined based on the neoplasm growth and metastasis.

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