CASE REPORT

Primary cardiac angiosarcoma: presenting with cardiac tamponade followed by cerebral hemorrhage with brain metastases

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Abstract A 49-year-old man presented with palpitation and shortness of breath. He was seen to have a massive pleural and pericardial effusion on radiography and echocardiography. Computed tomography (CT) scanning showed that cardiac tumors arose from the right atrium with epicardial and endocardial extension. Pathology examination of samples at pericardiotomy revealed them to be angiosarcoma. Two days after the surgery, he developed left hemiparesis. CT scans showed a large cerebral hemorrhage on the right temporal lobe with midline shift by brain metastases. He died 37 days after the surgery. At autopsy, he had metastases in the brain, multiple bones, and soft tissues but no lung or leftside heart involvement. Primary cardiac angiosarcoma is rare, and mostly arises from the right side of the heart. Common metastatic sites are the lungs and liver. There are only a few reports of brain metastases. In conclusion, this is a rare report of cardiac angiosarcoma presenting with pericardial tamponade. There were rapid brain and multiple bone metastases but no lung or left-side heart lesions.

Key words Angiosarcoma · Cardiac tamponade · Brain metastases

Introduction

Primary cardiac sarcomas are rare and constitute less than 25% of primary cardiac tumors. Metastases are common and are often widespread. Most patients die within a few months after symptom onset. We report a patient with an angiosarcoma that originated in the right atrium who presented with cardiac tamponade followed by cerebral hemorrhage with rapid brain metastases.

Case

A 49-year-old man presented with a 2-month history of palpitation and shortness of breath. He had an episode of hemorrhage of hypodermal bleeding in his left chest. He was a regular worker and had no special medical history. His laboratory tests revealed a slight tendency of prothrombin time prolongation (14.4; normal 10.7–

One month later, he was seen to have a massive pleural effusion and cardiac tamponade on chest radiography and echocardiography. Chest computed tomography (CT) scans showed cardiac tumors that arose from the right atrium with epicardial and endocardial extension (Fig. 1a). We performed exploratory pericardiotomy and removed 400 ml of sanguineous fluid. Multiple tumors were found over the right atrium and

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occupying the pericardial space. The tumors were dark red and easily hemorrhaged. We chose palliative therapy and relieved the cardiac tamponade. Two days after surgery, he suddenly developed left hemiparesis. The head CT scan showed a large cerebral hemorrhage on his right temporal lobe with midline shift presumably caused by a metastatic brain lesion (Fig. 2b). One month



Fig. 1 Chest Computed tomography (CT) scan showed that the cardiac tumor (*arrow*) arose from the right atrium and exhibited epicardial and endocardial extension

after surgery, he was found to have multiple bone and soft tissues metastases by CT. He died 37 days after surgery.

At autopsy, cardiac tumors were found to have invaded the right atrium, especially the lateral wall, but not the left atrium or interatrial septum. Disseminated nodules were present in the right ventricle wall and pericardium (Fig. 3a). Multiple metastases were found in the right temporal lobe of the brain, subcutaneous tissue of the left shoulder area with left subclavian artery invasion, the right psoas muscles, and multiple bones (left clavicle, several lumbar vertebrae) but not the lungs, the cardiac left atrium, or the cardiac left ventricle. Microscopicably, the right atrium tumor was composed of a proliferation of atypical spindle cells forming thinwalled blood vessels filled with numerous erythrocytes (Fig. 2b). This tumor was also stained positively with the antibodies for factor VIII-related antigen and CD31 as the endothelial markers, which was compatible with angiosarcoma.

Discussion

Primary cardiac angiosarcoma is a rare tumor that usually originates in the right side of the heart. In adults,

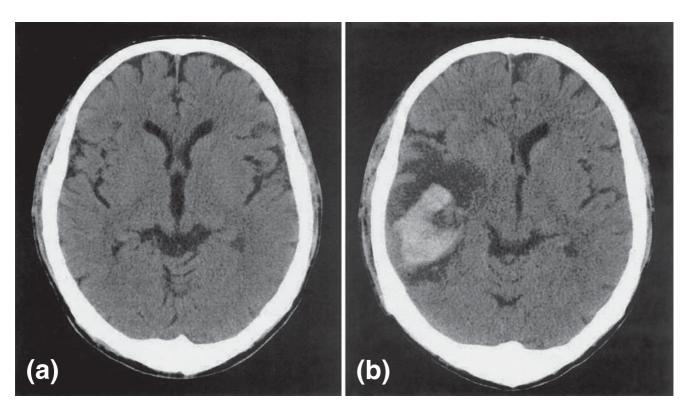
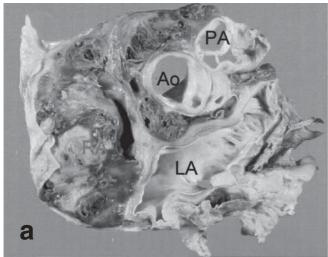


Fig. 2 At admission, there was no significant lesion apparent on his head CT scan (a). Only 8 days later his head CT scan showed a large cerebral hemorrhage on the right temporal lobe with a midline shift caused by brain metastasis (b)



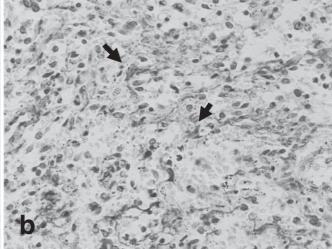


Fig. 3 a At autopsy, cardiac tumors were seen to have originated from the right atrial wall and invaded the right ventricle wall and pericardium. PA, pulmonary artery; Ao, aorta; LA, left atrium b Angiosarcoma is composed of a proliferation of atypical spindle

cells forming thin-walled blood vessels filled with numerous erythrocytes and stained positively with the antibodies for factor VIII-related antigen (*arrows*)

approximately 25% of primary cardiac tumors are malignant, and one-third of these malignant tumors are angiosarcomas.² A review of cardiac angiosarcomas by Janigan et al. showed that right and left atrium involvement was 93% and 7%, respectively. Central nervous system metastases were reported in three of nine cases in their review.⁵

During our search of articles and reports about primary cardiac angiosarcomas, we found only nine cases of brain metastasis since the report of Janigan et al. There were five males and four females (15–58 years old). Six of the lesions originated in the right atrium, two in the left atrium, and one in the pericardium. There were seven angiosarcomas, one fibrosarcoma, and one sarcoma. Five patients had brain metastases after cardiac surgery with cardiopulmonary bypass, which could have caused systemic dissemination of tumor cells. Among the other four patients, three had lung metastases, and one originated in the left atrium. Therefore, all nine cases had sufficient cause for systemic metastasis of the tumor cells.

In our case, on admission to the hospital, the head CT scan showed no metastases (Fig. 2a). However, cerebral hemorrhage (probably at a brain micrometastatic site) occurred only 8 days later without lung or left-side heart metastatic lesions.

The mechanism of the brain metastasis in our patient is still unknown. Even after vigorous pathology examination, there was no evidence of direct invasion of the tumor into the left ventricular wall or the aortic wall. However, these tumors existed in the pericardial space.

There is still a possibility that the tumor cells invaded the coronary circulation from the epicardial space. Furthermore, generalized metastasis might occur via micrometastases reaching cardiac perforated arterioles from the coronary circulation to the left ventricular chamber. Needless to say, this speculation is but one of the possible mechanisms that can produce of generalized metastases.

In conclusion, this is a rare case of angiosarcoma that originated in the right atrium. It presented with massive pleural effusion, pericardial tamponade, and rapid brain and multiple bone metastases but with no lung or left-side heart lesions.

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