Heart Metastasis of Extraskeletal Myxoid Chondrosarcoma

Ljiljana Banfić, Ivan Jelić1, Dražen Jelašić2, Snježana ČuZić2

Departments of Cardiovascular Diseases, 1Cardiac Surgery, and 2Pathology, Zagreb University Hospital Center, Zagreb, Croatia

Isolated, remote heart metastasis of myxoid chondrosarcoma is extremely rare. We present an unusual case of isolated remote metastasis of extraosseus myxoid chondrosarcoma from the right ankle region to the right ventricle, its clinical course, and treatment in a 46-year-old woman. Although heart biopsy done before the surgery revealed myxoma, histopathologic diagnosis of the heart tumor was confirmed after its surgical resection from the right ventricle. Nine months after the surgery the patient was doing well but, seven months later, she died in the local hospital because of global heart failure. On the autopsy, the only metastatic lesion found was in the heart. The pericardium and heart muscle were infiltrated with the tumor, whereas all other organs were free from malignant dissemination.

Key words: chondrosarcoma; echocardiography; heart neoplasms; neoplasm, metastasis; soft tissue neoplasms

Primary malignant cardiac tumors occur extremely rarely. Among them, sarcomas are the most frequent (1). Primary chondrosarcoma of the heart has been previously reported, but it is an extremely rare finding (2,3). There are few reports of the metastatic chondrosarcoma in the heart (4,5).

Metastatic tumors of the heart, especially of the pericardium, are more common than primary malignant tumors (6). Carcinoma of the bronchus, melanoma, breast carcinoma, and lymphoma are the most common primary tumors that produce cardiac metastases. It is extremely rare for a soft-tissue mesenchymal tumor of low malignant potential, such as myxoid chondrosarcoma, to give isolated heart metastases (7). It is even more unusual to have a solitary metastasis in the myocardium, since pericardium is more frequently affected with metastatic dissemination (6).

Case report

A 46-year-old woman was referred to our echocardiography laboratory because of the second incident of pulmonary embolism. She had developed exertional dyspnea and non-productive cough during the previous month. The transthoracic echocardiography revealed great tumorous mass in the right ventricle.

Her previous medical history was unremarkable until the age of 40, when the extraosseus tumor was discovered slightly above the right ankle. The histology revealed soft tissue myxoid chondrosarcoma. After it was surgically treated, the patient was in remission for three years. Then, because of the local recurrence of the tumor, the patient underwent the second surgery, followed by local irradiation therapy. X-ray revealed that there was no involvement of the bone in the right ankle. Nevertheless, because of the reappearance of the tumor, the patient was scheduled for below-knee amputation to prevent local and remote dissemination. Chemotherapy was not suggested, since this type of tumor is resistant to chemotherapy (8).

While waiting for the amputation, the patient was admitted to the local hospital because of the pulmonary embolism. Perfusion lung scan was positive. Ventilation scan was not done at that time. There was no lymphadenopathy and no evidence of pulmonary parenchymal infiltration on the chest X-ray. Echocardiography revealed only mild left ventricular hypertrophy. Below-knee amputation had to be postponed because of the pulmonary embolism, and the patient refused the surgery herself. One year later, she suffered pulmonary embolism again and was referred to our echocardiography laboratory for examination.

On admission, she complained of dyspnea and mild non-productive cough. She was obese and hypertensive. Neck veins were moderately engorged and liver was tender and enlarged. She had right basal pulmonary rales. Her right ankle was edematous and deformed after the repetitive surgery and irradiation of the tumor. The laboratory findings were in normal ranges except for high lactic dehydrogenase levels. Chest X-ray showed right
lower lobe effusion. Lung perfusion scans confirmed multisegmental perfusion defects in both lungs and segmental perfusion defect in the right lower lobe. Pulmonary angiography revealed embolic occlusion of the pulmonary artery branch for the right lower lobe.

Echocardiography revealed a big tumor in the right ventricle. Most of the right ventricle cavity was occupied with immobile tumor masses, which grew from the right ventricular free wall and invaded the part of the right ventricular apex and the apical part of the interventricular septum. The tumor pushed the tricuspid valve into the right atrium, which was free from masses, except for the small area of the right side of the interatrial septum (Fig. 1). The interventricular septum bulged into the left ventricular cavity and compressed it. Tumor was lobulated and consisted of acoustically inhomogeneous tissue. There was no evidence of calcification in it. Transesophageal echocardiography confirmed the pulmonary artery trunk and pulmonary arteries both empty.

The findings on transthoracic echocardiography were correspondent (Fig. 1).

There was no evidence of leg venous thrombosis on a Duplex scan. Abdominal ultrasound revealed only moderately enlarged liver with dilated intrahepatic veins. Chest computerized tomography (CT) scan confirmed the big tumor in the right heart (8×6 cm) and no involvement of the pericardial sac or big vessels. There was no evidence of a mediastinal enlargement. Abdominal CT-scan showed only dilated hepatic veins. The only expansive formation was located in the right ventricle.

We assumed that the big solitary mass in the right ventricle was the metastasis of the chondrosarcoma originating from the right leg. Another hypothesis was that it could be another tumor or even an old large thrombus. Since there was no evidence of local or remote dissemination of chondrosarcoma, we did the biopsy of the intracardiac mass. Five tissue specimens were taken by transjugular approach. The histology revealed ventricular myxoma or myxomatous degeneration of the old thrombus.

The patient was than referred to a heart surgeon because of deteriorating right heart function and decreased cardiac output. She had an open-heart surgery with the standard cardiopulmonary bypass. She recovered normal sinus rhythm and was on the ward on the second postoperative day.

The intraoperative surgical finding was impressive and corresponded with the echocardiographic findings. An extremely big tumor, growing from the lateral wall of the right ventricle opposite the interventricular septum, filled the cavity. It was resected with the belonging myocardium invaded with the tumor. The most of the tumor was removed, except the part that invaded the right ventricular free wall.

**Pathological Findings**

Macroscopically, the tumor was soft to firm, ovoid, lobulated, and generally well circumscribed, with a distinct fibrous capsule. On a cross section, it was lobulated, with foci of hemorrhage and necrosis (Fig. 2). Its surface was gelatinous, grey to tan brown. It measured 8×3 cm (Fig. 2).

Tumor tissue was fixed in 10% neutral buffered formalin and processed for routine paraffin-embedding. Five
m sections were stained with hematoxylin-eosin (HE), periodic acid Schiff reagent (PAS), PAS diastase (PAS-d), and treated with hyaluronidase. Immunohistochemical staining with avidin-biotin complex (ABC) method, using antibodies against vimentin, cytokeratin, EMA and S-100 protein, and CD68 (DAKO, Glostrup, Denmark) was done. Histologically, the tumor consisted of rounded or slightly elongated cells in lacunas (typical for chondrocytes), separated by variable amounts of mucin positive (hyaluronidase treated) intercellular matrix (Fig. 3). The cells had small hyperchromatic oval nuclei and narrow rim of eosinophilic and PAS positive cytoplasm. Mitotic figures were rare (Fig. 3).

Immunohistochemically, they were intensively vimentin and pale S-100 positive; and cytokeratin, EMA, and CD68 negative. The histologic diagnosis was myxoid chondrosarcoma. Histological finding in the heart was the same as the primary tumor located on the ankle.

Because of the tumor low radiosensitivity, as well as low chemotherapeutic sensitivity, the surgery was the only therapeutic option (8).

Nine months after the surgery the patient was feeling well. Couple of months later, her physical condition deteriorated. She developed a heart failure and was treated in the local hospital. The patient died in cardiogenic shock sixteen months after the heart surgery. The autopsy revealed a large tumor mass infiltrating the whole heart. Myocardium and pericardium were infiltrated with tumor tissue but there was no evidence of metastatic pulmonary embolism. The autopsy did not reveal local or remote metastatic involvement of any other organ except the heart.

Discussion

We present an extraordinary case of an extremely rare type of tumor – extraskeletal myxoid chondrosarcoma, which produced a solitary heart metastasis. There have been no reports published about a solitary cardiac metastasis of this very rare extraskeletal (soft tissue) myxoid chondrosarcoma. There were some reports of metastatic heart chondrosarcoma (4,5), but almost all metastatic chondrosarcomas reported in the literature had their origin in the bone tissue. There was also a report on primary cardiac chondromyxosarcoma (3). In our patient, the primary tumor had its origin in the soft tissue. Most heart metastases are usually confirmed at autopsy, but in our case the diagnosis was histologically confirmed in living patient. All the clinically based assumptions and dilemmas during the medical treatment were confirmed at and corresponded to the autopsy findings.

Myxoid chondrosarcoma is a slow growing tumor, prone to local recurrences and metastases, sometimes years after initial diagnosis (7,9,10). In our case, the metastasis in the heart showed more aggressive and invasive behavior than the primary tumor on the leg. There is some evidence (11) that the remote dissemination rate is higher in patients with primary local recurrences than in patients in whom the tumor does not reappear after the extirpation of the primary process (86% vs 19%, respectively).

The conclusions about growth potential in the heart can be based on the echocardiographic findings. During the first pulmonary embolic event that occurred one year before the admission to our hospital, echocardiographic findings were normal. Only a year later, echocardiography revealed a great mass in the right ventricle. We can conclude that it took one year for the metastatic tumor to reach such great dimensions and invade the most of the right ventricle. This is in contrast with its previously described slow growing potential of this type of tumor (7,9,10).

The question is weather it was really a solitary metastasis in the heart and were the pulmonary embolic events metastatic in their origin or not? If it had been metastatic pulmonary embolism, it would not have re-pressed after only heparin treatment applied during the first pulmonary embolic event. Also, in respect to proliferative nature of the heart metastasis, whose growth was extremely fast and invasive, it would be reasonable to expect the same or even worse clinical course of the pulmonary embolism if it were metastatic. Pulmonary embolism was the first symptom at the time when transthoracic echocardiography finding was normal. It occurred even one year before the heart tumor appeared. We believe that thrombotic source was tumor surface in the right ventricle or right leg, although no thrombotic lesion was discovered on duplex scan of the lower extremities. This clinical conclusion was confirmed on autopsy, as no metastasis was found in the lung.

The heart biopsy failed because the tumor was covered with a gelatinous cap and the samples could not be taken from the deeper part of the tumor.

The more precise diagnosis could have been made before the surgery if magnetic resonance imaging (MRI) of the heart had been done. If malignant characteristics of the tumor were revealed on MRI, the clinical decision could have been more conservative because of great susceptibility for metastatic dissemination during the extra-corpooreal bypass (12). Oncological counseling recommended surgical treatment, because of the poor radio- and chemo-sensitivity of the tumor (8). Although the tumor could not be removed completely, the patient had the benefit of therapy. She recovered immediately after the operation and had a good quality of life for about a year after the surgery.

References

6. Colucci WS, Schoen FJ, Braunwald E. Primary tumors of the heart. In: Braunwald E, editor. Heart disease: a text...


Received: November 3, 2000
Accepted: February 3, 2001

Correspondence to:
Ljiljana Banfić
Department of Cardiovascular Diseases
Zagreb University Hospital Center
Kišpatićeva 12
10000 Zagreb, Croatia
banfic@yahoo.com