Propose a Case of Cardiac Osteosarcoma Primary-Results and Review of Literature

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Abstract

Overview: Primary cardiac osteosarcoma, a bone connective tissue cancer, is a rare form of heart cancer. In the literature, there are few reports of primary osteosarcoma. Herein, we report a rare clinical case of primary cardiac osteosarcoma. A 71-year-old male patient was admitted to the hospital with symptoms of dyspnea, echocardiography revealed a tumor in the left atrium verified as an osteosarcoma following biopsy. After surgery, the patient self-treated with traditional medicine but relapsed and passed after 20 months.

Research Methods: Based on a clinical case at Viet Duc University Hospital, we reviewed the literature to describe the pathology and discuss treatment methods.

Conclusion: Primary cardiac sarcoma is rare, treated with surgical resection and adjuvant chemotherapy, has high recurrence rate, and continue to maintain a poor survival prognosis.

Keywords: Cardiac tumor primary, Osteosarcoma.

Introduction

Primary cardiac tumors are rare and, according to Butany et al., the incidence is 0.001 - 0.030 % 1. About 75% of primary cardiac tumors are benign, including myxoma, lipoma, papillary fibroelastoma and rhabdomyoma. Myxomas are predominant. Malignant heart tumors are usually derived from metastases elsewhere in the body. Among primary malignancies, sarcomas are the predominant cancer. Currently, only a few hundred cases of cardiac sarcomas have been reported in the literature. Angiosarcoma and myxofibrosarcoma are the most common types of cardiac tumors with osteosarcomas being rare. Primary myocardial sarcoma may occur at any age but is more common during middle age. The clinical presentation of patients with cardiac sarcoma depends on the size and location of the tumor. Patients often have dyspnea alongside other symptoms due to embolism, pericardial effusion, and rarely metastases to other sites. Depending on the location of the tumor in the heart, complete removal is often difficult for a single operation. Therefore, a combination of chemotherapy and adjuvant radiation therapy remains popular in the treatment of cardiac sarcoma. If the tumor is localized to the heart, treatment with heart transplantation has been shown to provide long survival in some patients with little evidence of recurrence observed despite immunosuppression. However, the prognosis of primary cardiac sarcoma is generally poor. Median survival in patients with cardiac sarcoma has been reported to be approximately 11 months on average. In patients with primary cardiac sarcoma, complete resection of the tumor is recognized as the most effective and mainstay treatment to prolong survival. In this study, we report a case wherein after surgical treatment of the primary cardiac osteosarcoma, the patient relapsed and died 20 months after surgery. In this study, we review the literature from which to make recommendations on management and treatment of cardiac osteosarcomas.
Research Methods

Clinical case description at Viet Duc University Hospital in August 2021 with follow-up after 20 months in April 2023 and retrospective literature review on pathological descriptions and treatment of cardiac osteosarcomas.

Clinical Case

We describe a case of a 71-year-old male patient with no previous history of cardiovascular disease or chronic disease. The patient's primary occupation is that of a rural farmer. A week before admission, the patient appeared to have dyspnea and chest tightness. He was referred to Viet Duc University Hospital wherein a large tumor was discovered in the left atrium. Clinical examination revealed NYHA class II dyspnea, regular heart rate, and no abnormal murmurs. Echocardiography: detected in the left atrium a mass of 3.16 x 4.16 cm in size attached to the atrial septum causing moderate stenosis of the mitral valve (Figure 1). Cardiac multi-slice computed tomography showed a mass occupying most of the left atrium and part of the left ventricle measuring 80x39 mm with lack of enhancement after injection (Figure 2). Electrocardiogram: sinus rhythm 100 beats per minute, regular, normal axis.

After diagnosing left atrial tumor with mitral valve obstruction, we decided to pursue surgical removal of the tumor. The operation consisted of a midsternal chest cavity exposure and extracorporeal bypass circulation with 2 IV cannulas. We chose the incisions through the right atrium and the atrial septum to access the tumor. The tumor filled the left atrium and the atrial wall showed scattered infiltrates in biopsy. The tumor was completely resected via cauterization of the tumor’s stem. Moreover, we found the mitral valve to be hypertrophied with anterior prolapse in the A2 region (according to Carpentier), resulting in mitral regurgitation and a deformed annulus via a nodule on the anterior leaflet. We remove the nodule and placed a mitral valve ring. Biopsy confirmed the osteosarcoma (Figure 3). Extracorporeal circulation was 150 minutes. Aortic clamp time was 100 minutes. The patient was extubated after 5 hours.

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Figure 1: Echocardiography.

Figure 2: Computer Tomography tumor in the left atrial.

Figure 3: Pathology of the Osteosarcoma, A. Macroscopic imaging of tumor organization shows heterogeneous colorization and density with pinkish-white regions with unclear borders for myocardial organization. B. HE x 100. Two interstitial components consisting of spindle cells, and neoplastic cells alternates within the hyaline and chondrocyte matrix. C. HE x 400. Tumor infiltration into the bone matrix with calcium deposition. Neoplastic cells are polymorphic and mitotic. D. HE x 400. Large necrotic neoplastic area alongside tumor infiltrate of the bone matrix.
Right hilar lymph nodes and nonspecific parenchymal opacities were found in the right lower lobe (Figure 6). Based on our evaluations and the patient's wishes, we did not pursue further surgical intervention. The patient was discharged from the hospital, receiving palliative care and at-home monitoring. The patient passed 20 months after surgery, due to progressive dyspnea and organ failure.

**Discussion**

In the literature, primary cardiac tumors are rare. According to Butany et al., the incidence based from biopsied specimens is 0.001 - 0.030 % [1] with metastatic tumors being 20 to 40 times higher in prevalence [2]. To note, 75% of all cardiac tumors are benign. Within the malignant group of cardiac tumors, sarcomas are common, with at least a 95% occurrence rate in the malignant group [3]. Wang et al. reports 53 cases in a pooled report based on their own literature review [4]. 80% of cardiac tumors arise in the atria and blood vessels. of the lungs, in which angiosarcomas (vascular sarcoma) and myxofibrosarcomas (mucous fibrosarcoma) are common types. Vascular sarcomas are the most common, accounting for about 50%. Cardiac osteosarcomas are rare, making up less than 10% of cardiac sarcomas. Burke et al. only recorded 9 cases of osteosarcoma out of 81 cases of cardiac sarcomas within a timespan of 12 years [5]. Extra-skeletal osteosarcomas are most common in the soft tissues of the lower extremities. In the WHO 4th edition classifications released in 2015, osteosarcomas were listed as a separate tumor type via ICD code 9180/3 [6]. Most cardiac sarcomas have several features in common: these tumors appear at any age, but the earlier in age they appear, the more malignant they are likely to be. There is no gender preference among those with cardiac sarcomas but there is commonly atrial involvement in up to 75% of cases. In particular, cardiac angiosarcoma (the most common type of cardiac sarcoma), is most often found in the right atrium or right ventricle whereas osteosarcomas are commonly found in the left atrium [4,5,7]. Compared to other types of noncardiac sarcomas, the prognosis for cardiac sarcomas is very poor [3-8]. Bakaeen et al. tracked the median survival time after cardiac sarcoma surgery to be 23.5 months. Therefore, a thorough differential diagnosis of benign heart tumor from sarcoma is crucial to producing the best treatment options for each patient.

**Clinical symptoms and diagnosis**

The clinical presentation of cardiac sarcomas depends on tumor size and anatomical location. Growth rate, friability, and invasiveness are also important determinants of clinical features. Most cardiac tumors are usually asymptomatic and may be identified only via imaging or autopsy. Clinical symptoms manifest when tumor size impairs of the functionality of the heart...
or other proximal structures. Symptoms from excessive cardiac tumor size are manifest similarly valve obstruction, arrhythmias, or heart failure. Vascular occlusion due to tumor emboli and metastases of adjacent structures may be observed. Mass effect, compression, impeding cardiac function are the most suggestive symptoms for a malignant heart tumor [4,5,9]. Although non-specific, our report also covers the common symptoms of cardiac sarcoma. The most prevalent symptoms are associated with congestive heart failure - such as dyspnea, angina, edema, cough, tachycardia, palpitations, arrhythmia, and hemoptyis, in addition to symptoms. CNS symptoms, e.g., syncope, headache, dizziness, and hemiparesis are also associated. Gastrointestinal symptoms are prevalent and include nausea, vomiting, anorexia, and epigastric discomfort. Systemic symptoms may be observed which include fatigue, fever, and weight loss, are all prognostic for a malignancy. Moreover, an ejection murmur may be heard due to valvular obstruction. Timing of symptoms may arise suddenly or gradually. It is important to note that there may be no clinical symptoms observable. On laboratory testing, elevations of BNP and ANP are common, as well as elevations of hepatic markers AST, ALT, and LDH. In addition, there is increased creatinine, blood urea, ESR, CRP, PT/PTT, and elevated WBC [4]. Echocardiography is the most commonly used first-line modality for diagnosis of cardiac tumours. Typically, osteosarcoma tumors appear heterogeneous with areas of high echogenicity indicating calcification. However, calcification may not always be detected [4]. Other imaging studies such as computed tomography (CT) and magnetic resonance imaging (MRI) have a role in staging of the tumor, its adjacent structures, and the assessment of its metastasis. On CT, the tumor is often heterogeneous and rarely has obvious calcifications. Bone scintigraphy is important in patient follow-up and metastasis assessment for osteosarcomas [2]. Electrocardiogram shows no significant markers. On CXR, cardiomegaly with or without interstitial opacities is the most common finding, and pleural effusion may also be observed. However, biopsy is the definitive diagnosis for all cardiac tumors [4,9]. In this case, we encountered a rare case of cardiac osteosarcoma, which had multiple nodules in the atrial wall as well as the anterior mitral leaflet indicating a high prognosis for recurrence. Because of the tumor mass effect compressing the mitral valve, surgery was performed with the initial diagnosis of a left atrial mucinous tumor. After surgery, and based on pathology, we confirmed diagnosis of a cardiac osteosarcoma. Histologically, the tumor may appear as clusters, either mushroom-shaped or circular. The outer surface can be smooth or rough. Invasion into surrounding structures is often observed. Grossly, it is often yellow-gray, and hemorrhagic-appearing with regions of necrosis. All three common cell types of osteosarcomas that normally arise in bone (osteoblasts, trophoblastics, and fibroblasts) can be observed in a primary cardiac osteosarcoma. The cellular origin of primary cardiac osteosarcoma remains unknown [4,5]. According to previous reports, nearly all types of soft tissue sarcomas can occur in the heart. For most patients, primary cardiac sarcoma presents with no family history. These findings suggest that most types of sarcomas (excluding hemanginomas, synovial sarcomas, and mesothelioma, as these tumors are uncommon in the left atrium) may originate from a similar origin, the mesenchymal root of the left atrium [4]. In summary, it is difficult to accurately diagnose cardiac osteosarcomas using imaging alone, as most cardiac tumors may present similarly.2. Routine laboratory blood tests are often nonspecific. Of all cardiac malignancies, the most common abnormality is elevation of myocardial enzymes such as BNP/ANP, possibly due to severe myocardial damage. The gold standard for definitive diagnosis is made via biopsy.

Treatment and follow-up

Compared with other types of noncardiac sarcomas, the prognosis for cardiac sarcomas is very poor [3-8]. Currently, the authors agree on a multi-therapeutic approach for patients with malignant heart tumors. This approach involves a combination of intervention, surgery, medical resuscitation treatment, chemotherapy, and radiation. Surgery is often performed before chemotherapy because most cases are diagnosed as atrial mucinous tumor before surgery [3,5]. Surgical removal of the tumor is important for the treatment of cardiac osteosarcomas. All studies recommend that complete tumor resection by surgery is necessary to increase positive prognoses for patients. However, the difficulty of complete resection of the tumor due to the invasion of surrounding structures and high recurrence limits the benefit of surgery. The mortality rate from surgery-related causes for cardiac sarcoma is 8.3%, according to Zhang et al [9]. All reports show a high rate of local recurrence in the absence of adjuvant therapy. In addition to surgery, postoperative chemotherapy is also one of the most effective treatment strategies. Documented chemotherapeutic agents include doxorubicin, cisplatin, cyclophosphamide (or ifosfamide), etoposide, mitomycin, methotrexate, and gemcitabine. It is recommended to treat cardiac sarcomas with radiation therapy and, if possible, definitively via heart transplantation if the malignancy is isolated to cardiac tissue. Radiation therapy is less recommended due to damage to healthy myocardial tissue [3]. Heart transplantation has recently been used to treat cardiac sarcoma but its role is controversial and not widely accepted as a popular treatment. The main means for treatment remains surgical for cardiac sarcomas [3,4,9]. According to our findings, there was no correlation between tumor size and location with patient survival. Histological classification seems to have prognostic significance as none of the patients with low-grade sarcoma died of the disease while half of the patients with high-grade classification died [4,9]. The general physical condition of the patient to tolerate cardiac surgery, improvements in cardiac surgical equipment and technology, and improved postoperative
care may be factors that contribute towards reduction in mortality rates post-operatively. Surgery remains the treatment of choice for cardiac sarcoma. Thus, in addition to the grading of the tumor, survival of the initial surgery is an important prognostic indicator for patients with high-grade cardiac sarcomas. Despite advances in surgery and aggressive chemotherapy, the prognosis of osteosarcoma and cardiac sarcoma remains poor. Over the years, no significant improvement in the treatment of the disease have been reported. Most studies express high rates of postoperative recurrence and distant metastases with the lungs, brain, and bone being the most common sites.

**Conclusion**

Osteosarcoma and cardiac sarcoma are rare diseases with poor prognoses. Currently, the most effective strategy involves a multimodal approach combining surgery and adjunctive chemotherapy.

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**References**