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Case Report

Primary Angiosarcoma of Pericardium with Cardiac Tamponade: A Case Report☆

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SUMMARY

Primary tumors of the heart and major vessels are rare. Angiosarcoma is the most frequent malignant tumor of the heart and usually involves the right atrium. Angiosarcoma originating from the pericardium is extremely rare and only several cases have been reported to date. The current study presents a case of primary angiosarcoma of pericardium with cardiac tamponade. After surgical treatment to relieve symptoms, the patient refused further therapies and died 3 months after diagnosis.

1. Introduction

Primary malignant tumors of the heart and major vessels are rare. Amongst the malignant tumors of the heart, angiosarcoma is the most common and accounts for more than two thirds of these malignancies[1,2]. Angiosarcoma frequently involve the cardiac chambers, especially the right atrium. However, detection of angiosarcoma of the pericardium is very rare[3]. The clinical presentation of pericardial angiosarcoma depends on the location of involvement rather than histological type. The prognosis for a patient with angiosarcoma is poor with an overall survival duration of between 6 months and 12 months even under multi-modality treatments[1]. Patient outcome is improved if chemotherapy is applied after surgical resection. There are several reports of patients' surviving up to 40 months.

2. Case report

An 83-year-old male patient presented with a history of atrial fibrillation, chronic ischemic heart disease, and congestive heart failure, for several years. He received regular medical control at out-patient clinics. One week before admission, progressive dyspnea on exertion, a dry cough, and a worsened low leg edema were present. He was brought to the out-patients department for first aid. A chest X-ray revealed a huge, bottle-shaped heart shadow (Fig. 1). Electrocardiogram revealed atrial fibrillation with diffuse low voltage amplitude. Routine laboratory studies were normal. Radiological findings indicated pericardial tamponade. An emergency transthoracic echocardiogram was arranged and showed a massive pericardial effusion with cardiac tamponade. Pericardiocentesis and drainage were performed and 500 mL hemorrhagic pericardial fluid was drained. The patient's symptoms were rapidly relieved. Cytological and bacterial examinations of the pericardial effusions were all negative. Chest computerized tomography (CT) was arranged on suspicion of a malignant tumor, and showed a 4.7 cm × 2.5 cm soft tissue nodule present at the pericardial space along the left atrioventricular groove. A 3 cm × 1.5 cm lesion next to the lateral wall of the left ventricular wall associated with mild pericardial effusion, and multiple mediastinal lymphadenopathy was also detected (Fig. 2). A positron emission tomography (PET) scan further confirmed a pericardial malignancy with mediastinal lymph node metastasis (Fig. 3). Surgical resection of the pericardial tumor was later performed. The pathology indicated multiple nodular proliferations on the pericardium. Tumor cells were strongly positive for CD31; weakly positive for WT-1; and negative for cytokeratin (AE1/AE3), HMB-45, and S-100 (Fig. 4). All these findings are consistent with angiosarcoma. Adjuvant chemotherapy and radiation therapies were recommended but the patient refused all treatments. He died following tumor recurrence 3 months later.

☆ Conflicts of interest: All contributing authors declare that they have no conflicts of interest.

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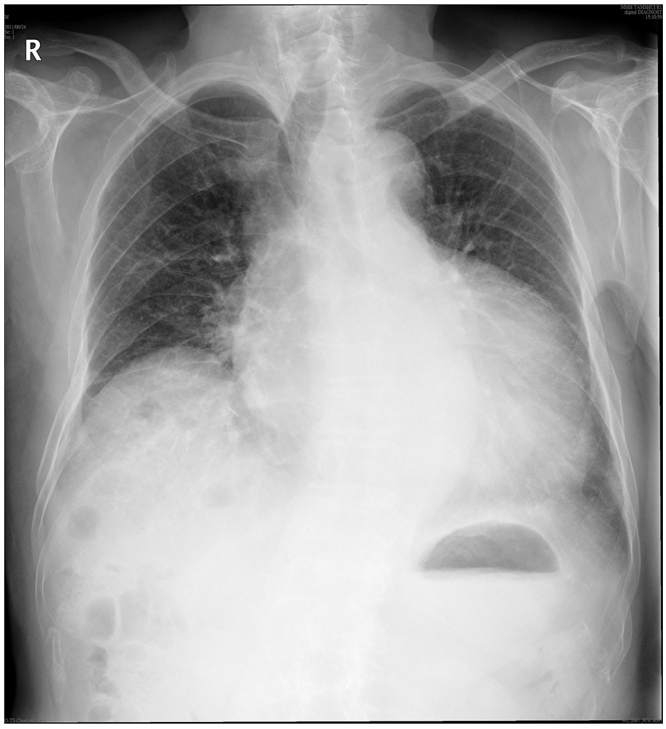

Fig. 1. Chest X-ray reveals bottle-shaped and enlarged heart size.

3. Discussion

Primary cardiac tumors are rare; their incidence was estimated to be 0.0017–0.003% in a study of routine autopsy[1]. The majority of cardiac tumors are benign with myxoma being the most common benign tumor of the heart (50%), followed by lipoma (~25%)[4].

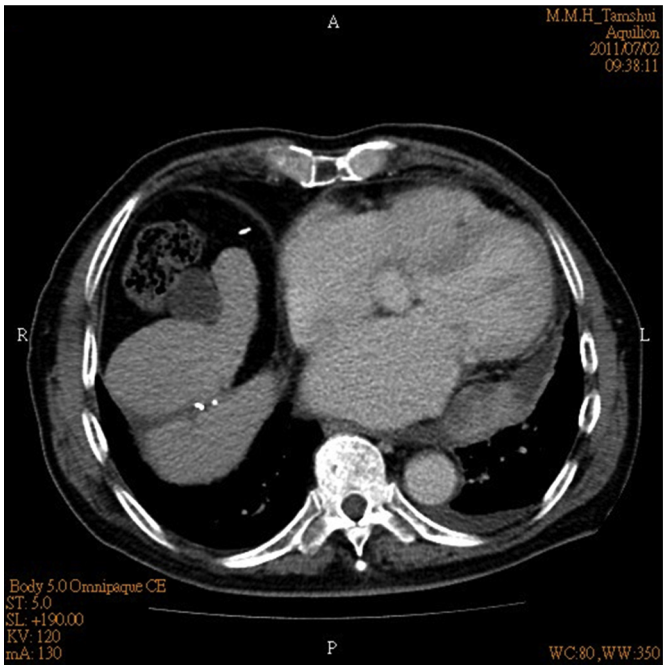

Fig. 2. A computed tomography scan reveals a 4.7 cm × 2.5 cm soft tissue nodule present in the pericardial space along the left atrioventricular groove with mild pericardial effusion.

Malignant cardiac tumors are estimated to account for 25% of all cardiac tumors[5]. Angiosarcoma is the most frequent malignant tumor of the heart. Malignant cardiac tumors frequently tend to involve the pericardium and right heart chambers, especially the posterior wall of the right atrium, as was seen in a report of 119 cases by Meng et al[4]. The prevalence does not differ by sex[1], however; a greater tendency was reported in male patients[6].

Angiosarcoma is the most common malignant tumor of the heart and its incidence is estimated to be about 31% of all malignant tumors. Angiosarcoma tends to occur in people aged 30–50 years. The initial diagnosis of angiosarcoma is usually delayed because of its nonspecific characteristics and detection depends on the surgical biopsy of tumors. Symptoms of angiosarcoma include dyspnea, chest discomfort, heart failure, palpitations, and general weakness. The most common site of involvement is the right atrium, followed by left atrium, right ventricle, and left ventricle[7]. Metastasis is about 80% when angiosarcoma is diagnosed. The lungs, mediastinal lymph nodes, and vertebrae are the most common metastatic sites.

A chest radiograph may disclose cardiomegaly, pulmonary congestion, or pericardial effusion. Transthoracic echocardiography is also an important tool used to detect cardiac tumors. Transesophageal echocardiography might have better ability to delineate the tumor location and the severity of local infiltration. Chest CT is useful to identify metastasis. Cytological analysis of pericardial effusion is also helpful; with the positive rate ranging from 75% to 87%.A transvenous endocardial biopsy procedure was reported in one study[8]. Despite all the above methods, surgical biopsy is always necessary to yield an adequate sample for diagnosis. Immunohistochemical stains are positive for CD31, CD34, and Factor VIII, which are characteristic features of vascular origin.

The overall average survival time is between 6 months and 12 months. Rare cases may survive up to 2–4 years. Tumor staging of cardiac angiosarcoma is currently not available, which may be due to its rarity. There is also a lack of large studies to compare the effects of different therapies in combination. Surgical resection remains important in order to examine the tissue and for rapid relief of symptoms. Even though total surgical resection is the treatment of choice, recurrence is common[6]. Angiosarcoma generally responds poorly to chemotherapy. The most frequently used chemotherapy drugs include Adriamycin, ifosfamide, cyclophosphamide, vincristine, and dacarbazine. There are not many studies comparing different chemotherapeutic regimens because of the rarity of cardiac angiosarcoma. Adriamycin-based regimens are the most commonly used regimens by oncologists. Pegylated liposomal doxorubicin has been used for the treatment of Kaposi's sarcoma in AIDS, its use was also reported for the treatment of angiosarcoma in one study[9]. Both Kaposi's sarcoma and angiosarcoma are tumors of the endothelial cells and may share some biological properties[10]. Recently, paclitaxel combined with liposomal doxorubicin has also been reported to demonstrate favorable results in the treatment of angiosarcoma[11]. Even though the overall response is poor with chemotherapy, patients treated with chemotherapy had better survival outcomes than did those without chemotherapy[12]. Adjuvant radiotherapy has a limited role of therapy due to its cardiotoxicity. It may play a role in patients with brain metastasis or superior vena cava syndromes[13]. Patients who avail of multidisciplinary therapies survived longer than those who were treated with a single therapy[6,12]. Overall survival may be up to 19 months in patients who undergo surgical resection and chemotherapy. Orthotopic cardiac transplantation has been tried in rare cases of angiosarcoma but patient survival did not differ from those without transplantation[14].

In conclusion, pericardial tamponade is an emergent medical condition which could be caused by a number of systemic diseases