

Case Report

Angiosarcoma of the pericardium: a case report

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Abstract: Background: Primary malignant tumors of the pericardium are rare, and most primary malignant pericardium tumors are mesotheliomas. Primary pericardial angiosarcoma is extremely rare, and it is associated with a poor prognosis. Case presentation: We report of a 47-year-old woman who complained of activity-related chest tightness and shortness of breath. Computed tomography, magnetic resonance imaging, and transthoracic echocardiography revealed an enlarged pericardium with hematic and solid components. An exploratory pericardiectomy was performed, and the results of the histological examination were suggestive of spindle cell hemangioendothelioma. She survived for 9 months after surgery without chemotherapy and radiotherapy, and she had a relatively good quality of life. Conclusion: Primary pericardial angiosarcoma is difficult to diagnose, and it has a poor prognosis. Pericardiectomy, radiation therapy, and chemotherapy were associated with a prolongation of survival.

Keywords: Primary pericardial angiosarcoma, therapy, prognosis

Background

Primary malignant tumors of the pericardium are rare, and most primary malignant pericardial tumors are mesotheliomas. Secondary involvement of the pericardium is more common, and it is often caused by direct invasion of cardiac and lung neoplasms or by metastatic spread. Angiosarcoma is the most frequent primary cardiac malignant tumor. However, primary pericardial angiosarcoma is extremely rare, and it has a poor prognosis. The Cleveland Clinic reported nine cases of cardiac angiosarcoma, three of which included pericardial involvement, whereas one case was a primary pericardial angiosarcoma [1]. The authors of the study concluded that multimodality therapy (surgery, radiation therapy, and chemotherapy) was associated with improved survival.

Case presentation

A 47-year-old Chinese woman who worked as a farmer complained of activity-related chest tightness and shortness of breath in April 2011. The symptoms recurred the next day, and she experienced nausea, followed by a

sudden loss of consciousness with twitching of her extremities and urinary incontinence. She was admitted to the emergency room of a hospital. Her systolic blood pressure was 70 mmHg, but her diastolic blood pressure could not be detected. Transthoracic echocardiography (TTE) revealed massive pericardial effusion. Pericardiocentesis and drainage were performed, and 200 mL of bloody pericardial effusion was drained. Results of cytological and bacteriological examinations of the fluid were negative, and routine findings were recorded as follows: white blood cell count, 220/mm³; MONO, 15%; and NEUT, 85%. Additionally, the following data were obtained for biochemical indices: Pro, 55.7 g/L; Glu, 5.68 mmol/L; PPD (-); ESR, 6 mm/h; LAM-Ab (-); and ADA, 9.9 U/L. The patient was treated for suspected tuberculous pericarditis. Her condition improved, and she was discharged several days later. The same symptoms recurred in July, and she was admitted to the emergency room of our hospital. Echocardiography revealed moderate pericardial effusion, and enhanced computed tomography (CT) of the chest showed heart shadow enlargement, pericardial effusion, uneven pericardial enhancement, and left pleu-

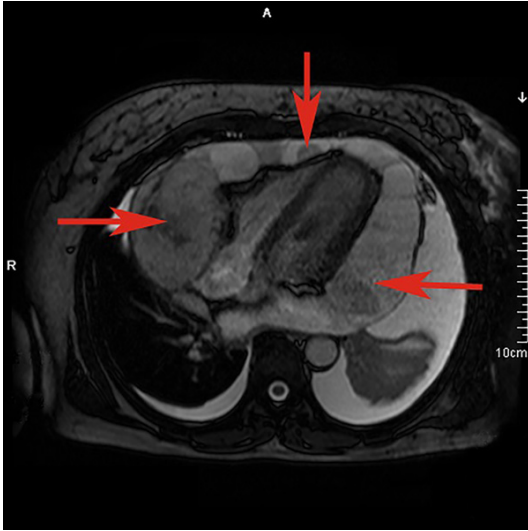


Figure 1. A cardiac magnetic resonance imaging scan showing an enlarged pericardium with hematic and solid components, as well as moderate pericardial effusion.

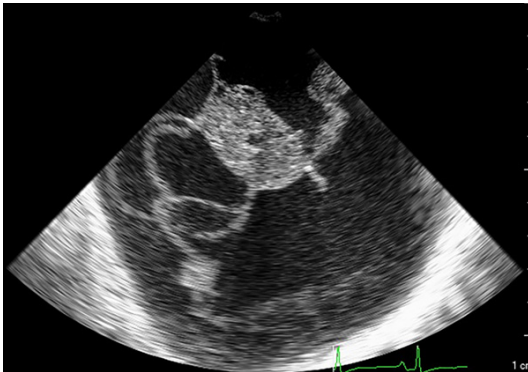


Figure 2. Transesophageal echocardiography showing several masses surrounded the heart, and a large spider-like mass located on the left lateral wall.

ral effusion. Cardiac magnetic resonance imaging (MRI) revealed an enlarged pericardium with hematic and solid components, as well as moderate pericardial effusion that caused constriction (**Figure 1**). She was admitted to the department of cardiac surgery in Peking Union Medical College Hospital, where she underwent sub-emergency exploratory pericardiectomy. The operation was performed on August 4, 2011. After anesthesia, transesophageal echocardiography revealed that several masses surrounded the heart, and a large spider-like mass was located on the left lateral wall (**Figure 2**). After opening the pericardium, we observed several masses of various shapes and sizes in the sac (**Figure 3**) and small-mound like mass-

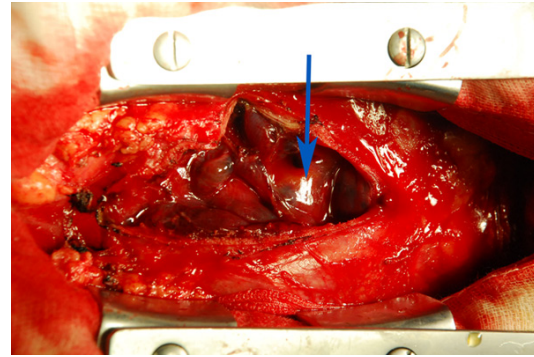


Figure 3. Intraoperative photograph of the angiosarcoma.

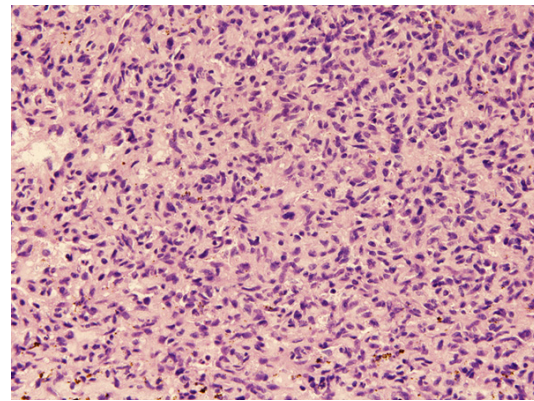


Figure 4. Histologic analysis of the biopsy specimen demonstrating spindle cell hemangioendothelioma.

es on the surface of the heart, the texture of which resembled jelly or grapes. There was a small amount of pericardial effusion. We removed two small masses measuring 0.5 and 1.0 cm, respectively, for fast frozen section examinations. The results of the histological examination were suggestive of spindle cell hemangioendothelioma (**Figure 4**). Although we ligatured the base before we removed the tissue, bleeding continued, but it was stopped after performing pressing hemostasis for 30 min. It was obvious that removal of the entire tumor was extremely dangerous, as we could not control the bleeding and there was a risk of injury to the heart. Thus, we decided not to remove the tumor, and the pericardium to the atrioventricular groove was excised to relieve constriction. We opened the left pleural cavity, and 800 mL of a yellowish transparent liquid was aspirated. There was no neoplasm in the pleural. Then, we closed the chest. After the operation, the patient's condition improved. The final outcome of the pathologic examina-

Angiosarcoma of the pericardium

tion was spindle cell hemangioendothelioma. We consulted the hospital oncologist and reviewed the literature of this disease, finding that the prognosis is poor even treated with chemotherapy or radiotherapy. We informed the patient and her family about the prognosis, and they decided against chemotherapy or radiotherapy, instead electing for conservative therapy and support. The patient was discharged 2 weeks later. She died of cardiac tamponade and multiple organ failure 9 months later.

Discussion

Angiosarcoma is the most common type of primary cardiac sarcoma [1], but primary pericardial angiosarcoma is extremely rare. Exposure to vinyl chloride is believed to cause angiosarcoma. However, as the patient was a farmer, she has no history of employment in the plastics industry, but we do not know whether the environment where she lived was polluted. Angiosarcoma is most likely to occur in the 3rd to 5th decades of life, and it is more common in men [2]. The tumor progresses without causing symptoms for a long period, during which patients appear healthy before presenting with non-specific symptoms such as chest pain, fever, shortness of breath, and pericardial and pleural effusions. In our patient, the effusion culture for bacteria, acid-fast bacilli, and viruses was always negative, as was the examination for malignant cells.

TTE is helpful for detecting tumors, but it is not sufficiently efficient. Enhanced CT can help to delineate the outline and blood supply of the tumor and MRI has excellent diagnostic advantages regarding tumor delineation and clarification of the tumor location, its local spread, and the involvement of adjacent structures. In our case, the findings of MRI were consistent with the results of exploratory thoracotomy. However, MRI, CT, or TTE cannot replace biopsy, and the final diagnosis depends on biopsy.

On immunohistochemical analysis, the tumor is positive for CD31, CD34, and Factor VIII. In our case, it was positive for CD31, CD34, and SMA and negative for desmin [3]. Angiosarcoma grows rapidly with local invasion and distant metastasis. Constriction of the pericardium is caused by both the tumor itself and the hemorrhagic pericardial fluid. Tumor growth around the heart can also lead to constriction of the pericardium.

The prognosis of patients with angiosarcoma is extremely poor. The mean survival is 6-14 months, and few patients survive beyond 14 months [1, 4]. This tumor responds poorly to chemotherapy and radiotherapy. The tumor can be resected by cardiac surgeons only when it grows as a localized mass, but even when chemotherapy is applied, survival is short [5]. Considering the adverse effects of chemotherapy and the poor prognosis, we recommend conservative therapy and support. Our patient survived for 9 months after surgery without chemotherapy and radiotherapy and had a relatively good quality of life.

Primary pericardial angiosarcoma is difficult to diagnose, and it has a poor prognosis. Pericardiotomy, radiation therapy, and chemotherapy are associated with prolonged survival.

Disclosure of conflict of interest

None.

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