



Cardiac angiosarcoma: a case report and short review of diagnostic modalities and therapy possibilities

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Primary cardiac neoplasms are rare, and angiosarcoma is the most common malignant cardiac tumor, which is very invasive and has a poor prognosis. We report a 54-year-old man with the huge mass in the right atrium. The patient underwent median thoracotomy and tumor was resected, which microscopic section showed angiosarcoma.

Keywords: cardiac tumor, angiosarcoma, immunohistochemistry.

Introduction

Primary cardiac neoplasms are rare, with an incidence of 0.0001–0.030% at autopsy [1]. Angiosarcoma is the most common malignant cardiac tumor, is very invasive and has a poor prognosis [2]. Typical localization is the right atrium [3]. Clinical symptoms are non-specific, usually presenting as cardiac tamponade or right-sided cardiac failure [4]. Diagnosis is often made after the disease has progressed. One of the most sensitive diagnostic tools is echocardiography. CT and MRT can be used to detect sites of metastasis. Histologically, angiosarcomas present in three patterns: a vascular area with anastomosing channels, a solid high-grade epithelioid area, and a spindle cell Kaposi-like area [4]. Immunohistochemical staining can be used as an adjunctive diagnostic tool. Surgical resection is the therapy of choice. The therapeutic effect of chemotherapy and radiotherapy is poor and multimodal therapy is under investigation.

Case report

A 54-year-old man with dyspnea, abdominal pain and echocardiographic diagnosis of pericardial effusion and a huge mass in the right atrium was referred to our institution. Physical examination revealed hepatomegaly and cardiac murmur over the 4th ICS at the left sternal border. X-ray

showed cardiomegaly. Angiography showed no pathological findings in the coronary arteries. Transthoracic echocardiogram demonstrated a large mass (3.5×10 cm) (Fig. 1) in the right atrium which prolapsed into and almost filled the entire right ventricle. 1 cm pericardial effusion was also present.

The patient underwent emergency surgery. Median thoracotomy was performed. Sanguineous pleural fluid was evacuated. Via right atriotomy the right atrial mass was well visualized. Part of the right ventricle was disguised by the tumor. The tumor (23×4×4cm) with a part of the right atrium was resected (Fig. 2). The part which invaded right ventricle was impossible to resect. The patient tolerated the procedure well. Microscopic section showed angiosarcoma. Immunohistochemistry showed strong positive CD31, CD34, CD 99, ERG-TLE1 markers and Ki-67 proliferation index was 20-50% (Fig. 3). He was transferred to medical oncology for adjuvant therapy on the 7th post-operative day.

Discussion

Angiosarcoma is the most common malignant cardiac tumor and has a poor prognosis [4]. It is a very invasive endothelial neoplasm which has high incidence of systemic metastases [3]. Symptoms are non-specific and diagnosis is mostly delayed. Echocardiography is a sensitive di-

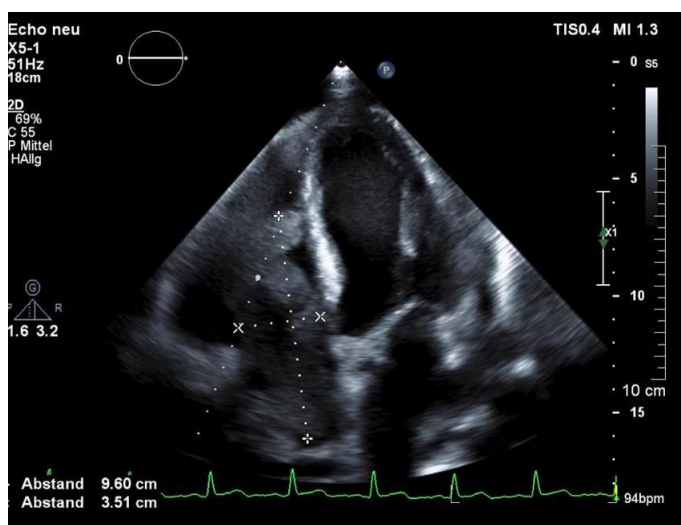


Figure 1.
Echocardiography four- chamber view. Huge mass in the right atrium expanding to the right ventricle.



Figure 2.
Huge angiosarcoma after surgical extracting from right atrium.

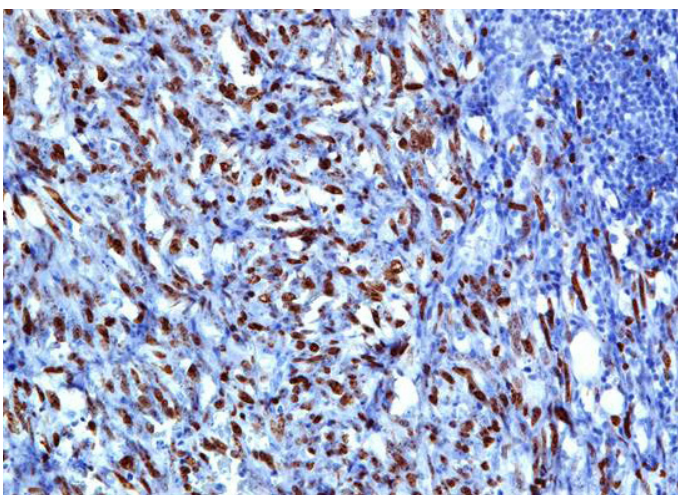


Figure 3.
Immunohistochemical analysis using Ki-67.

agnostic tool and can easily be used for initial diagnosis. CD31, CD34 and FLI-1 are the markers most commonly used to detect tumors of endothelial origin. Ki67 is a marker which used as a prognostic factor. Values of more than 10% are correlated with poor outcome. Angiosarcomas are often resistant to radiation and chemotherapy. Although possibilities are limited, the treatment of choice is surgical resection and multimodal therapy is currently under investigation [4].

Conclusion

From this case it is evident that angiosarcomas can present as a huge cardiac mass and complete resection remains challenging since tumors can encase essential cardiac structures. So, it is essential pay attention to leading non-specific symptoms in young patients and to diagnose disease before its progression which makes adequate surgical therapy impossible.

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