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Right atrial primary cardiac angiosarcoma extending into the right ventricle

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A B S T R A C T

Primary cardiac tumors are quite rare and about 75% of all primary cardiac tumors are benign in nature. We report a 34-year-old mentally retarded male who was diagnosed to have primary right atrial angiosarcoma which was extending into the right ventricle through the tricuspid valve. The tumor was totally resected at both sides including the interatrial septal attachment and the free wall of the right atrium with the resection margin being extended to the superior and inferior cavo-atrial junctions. Right atrium was reconstructed using glutaraldehyde-treated autologous pericardium. Light microscopy showed tumor formation with increased vascularity, scattered papillary invaginations and focal areas of necrosis with tumor cells having marked pleomorphic, hyperchromatic nuclei. Neoplastic cells were CD31, CD34 and vimentin-positive in immunohistochemical staining. After the surgery, the patient was scheduled for chemotherapy and was placed on the national waiting list for heart transplantation.

Introduction

Primary cardiac tumors are quite rare; the incidence was reported to be ranging from 0.001-0.03% in autopsy studies. About 75% of all primary cardiac tumors are benign in nature and right atrial myxomas comprise nearly half of all benign cardiac tumors (Reynen, 1995) while angiosarcoma is the most common malignant cardiac tumor (30%) followed by rhabdomyosarcoma (20%), with the 5-year survival rates for benign and malignant tumors being reported as 83% and 30% respectively (Hoffmeier et al., 2014). Therapeutic approach to the malignant cardiac tumors is challenging and requires implementation of a multimodal care since survival after resection of the tumors is dependent to various factors including non-specific and delayed presentation of the disease, spreading of the tumor cells via blood stream, necessity of a successful radical resection and uncertainty of the benefit of adjuvant therapy

(Ostrowski et al., 2014). In this paper, we report a case of primary right atrial angiosarcoma which was extending into the right ventricle through the tricuspid valve.

Case report

A 34-year-old mentally retarded male presented to our emergency department with hemoptysis, chest pain, shortness of breath and fever. The patient had no previous history of cardiac or pulmonary disease. On arrival, he had a heart rate of about 140 bpm, blood pressure of 90/60 mmHg and body temperature of 39°C. Physical examination was normal except for right parasternal 3/6 systolic murmur and mild leg edema. On chest x-ray, cardio-thoracic ratio was greater than 50%. Transthoracic echocardiography revealed a mobile intra-cardiac mass originating from right atrial side of the interatrial septum and extending into the right ventricle through tricuspid valve (Figure 1A). The mass was occupying most of the right atrial chamber. Since the patient was totally uncooperative and restless, transesophageal echocardiography was not attempted. Whole body computed tomography showed no tumor and no lymphadenopathy elsewhere. The mass lesion was suspected to be intracardiac thrombus or vegetation of infective endocarditis. The patient was initiated on intravenous broad-spectrum antibiotics and low molecular weight heparin. One week after the hospitalization, TTE was repeated and the lesion showed no regression in size. The patient was scheduled for surgery.

The operation was performed under general anesthesia. A midline sternotomy was performed, the pericardium was incised and pericardial fluid was evacuated which was hemorrhagic and approximately 300 ml in volume. The fluid was sampled for histopathological and microbiologic analysis. The surface of the heart was covered by a dense fibrous tissue and abnormal vascular formations. The whole right atrium and the upper part of right ventricle were occupied by the tumor. Left femoral artery and vein were prepared for in case of emergency cardiopulmonary bypass. After cannulating the ascending aorta, superior and inferior vena cavae were directly cannulated and CPB was initiated. Right atrium was opened via an oblique incision. The tumor mass was 45x25x5 mm in size, non-encapsulated and attached to the right atrial side of the interatrial septum. It was totally resected at both sides including the interatrial septal attachment and the free wall of the right atrium with the resection margin being extended to the superior and inferior cavo-atrial junctions (Figure 1B). Right atrium was reconstructed using glutaraldehyde-treated autologous pericardium. After removal of cross clamp, the heart restarted spontaneously in sinus rhythm and the patient was separated from cardiopulmonary bypass after complete rewarming. No other complications occurred.

There were multiple hemorrhagic tissue fragments on gross pathological examination. Frozen section revealed gray-crimson colored hemorrhagic tissue fragments which form papillary invaginations resembling malignant sarcoma. Light microscopy showed tumor formation with increased vascularity, scattered papillary invaginations and focal areas of necrosis with tumor cells having marked pleomorphic, hyperchromatic nuclei. Neoplastic cells were CD31, CD34 and vimentin-positive in immunohistochemical staining (Figure 2A-B). The patient was diagnosed with grade 3 angiosarcoma and scheduled for adjunctive chemotherapy. Since there were no distant metastases, he was considered eligible for a

heart transplant and was placed on the national waiting list.

Cardiac malignant tumors are mostly metastatic lesions that can originate from other organs. Primary cardiac malignant tumors are very rare and almost all malignant primary cardiac tumors are sarcoma. Angiosarcoma are the most common type of cardiac sarcoma, being about 75% of all primary cardiac malignant tumors followed by rhabdomyosarcoma (Sanoudos and Reed, 1972). Angiosarcomas are most commonly localized in right atrium followed by left atrium, right ventricle, interatrial septum and interventricular septum. Patients with cardiac angiosarcoma may present with various manifestations including, chest pain, hemoptysis, systolic and/or diastolic murmurs, finger clubbing, anemia, cardiomegaly, systemic and/or pulmonary embolism, atrioventricular conduction disturbances, cardiac tamponade and symptoms related with metastatic involvement. Functional mitral and tricuspid valve stenosis, superior and inferior vena cavae obstruction are clinical scenarios which may occur due to atrial angiosarcoma (Larriev et al., 1982).

There have been a number of case reports in the literature regarding the approach and treatment of cardiac angiosarcomas. Kugai et al. (2014) reported a 56-year-old man who presented with cardiac tamponade and right atrial angiosarcoma. The authors reported that despite successful excision of the tumor and adjunctive chemotherapy-radiotherapy, the patient died of multiple metastases 14 month after the operation. Chen et al. (2012) reported a 33-year-old man who presented with acute tamponade and then underwent emergency operation. The patient received one cycle of chemotherapy with a combination of docetaxel and gemcitabine and he was still alive 14 months after the operation despite having brain and lung metastases. Nakamura-Horigome et al. (2008) reported another 49-year-old man with right atrial angiosarcoma who responded well to a combination of docetaxel and radiotherapy. Suderman et al. (2011) reported that a 58-year-old patient with inoperable primary cardiac angiosarcoma survived 16 months after completion of radiotheraphy although he showed partial response the combination of docetaxel and radiotheraphy. These reports draw attention to the potential role of docetaxel and radiotheraphy combination in treatment of primary cardiac angiosarcomas.

Surgical resection of the primary angiosarcomas is still the standard treatment for primary cardiac angiosarcomas. However, benefit of adjunctive chemo-radiotherapy seems affected by the underlying cardiac decompensation as the patients with angiosarcomas present with different clinical scenarios ranging from effort dyspnea to acute life-threatening cardiac tamponade (Patel et al., 2014). Li et al. (2014) conducted a prospective survival analysis on 29 patients undergoing surgical resection of primary cardiac sarcomas within a period of 17 years. The authors postulated that aggressive surgical treatment or radiotheraphy prolonged the survival whereas the role of adjuvant chemotherapy was unclear.

This case was interesting given being diagnosed in his 3rd decade. Also there was no tumor invasion within both vena cavae although the right atrium was almost completely occupied by the tumor tissue. Differential diagnosis included right atrial thrombus or vegetation of infective endocarditis. Cardiac murmur was an important finding since initial symptoms include fever and dyspnea which might have

been misdiagnosed as pneumonia. Transthoracic echocardiographic evaluation helped differentiating between these diagnoses. Even though transthoracic echocardiography is of crucial importance in the diagnosis, detailed information including localization and size of the lesion could be precisely obtained by transesophageal echocardiography (Edwards and Louise, 1994) which was not feasible in our patient. Computed tomography and magnetic resonance imaging can also be used to evaluate mural invasion, local extension and distant metastases of malignant cardiac tumors. In our patient, whole-body CT showed no distant metastases.

The prognosis of cardiac sarcoma is poor; local extension and metastasis rates are as high as 80% until the diagnosis is made. Complete or partial surgical resection can be performed to remove obstruction and relieve symptoms in suitable cases. Yet, all of the treatment options can only provide palliation while role of adjunctive therapy including chemotherapy and radiotheraphy after surgical resection warrants further research.

Figure.1A Echocardiographic view of the mass extending through the tricuspid valve into the right ventricle; **B** Operative view of the tumor

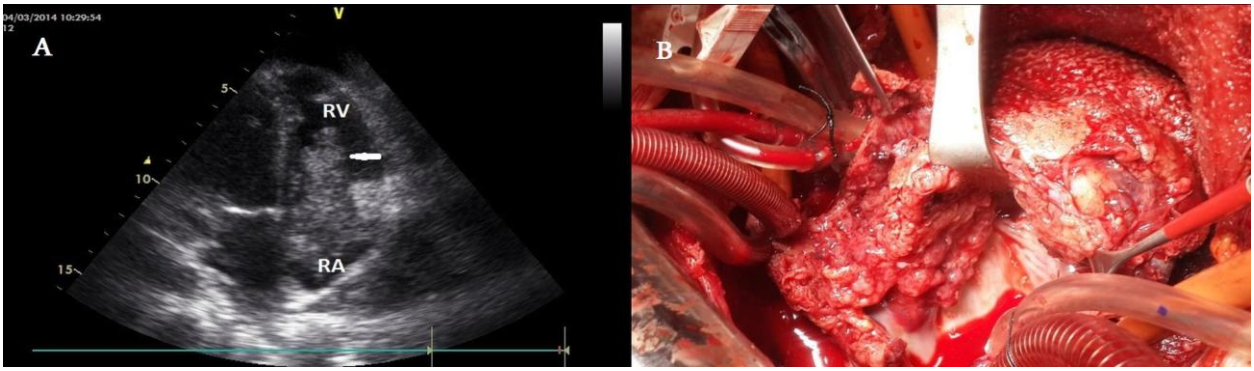
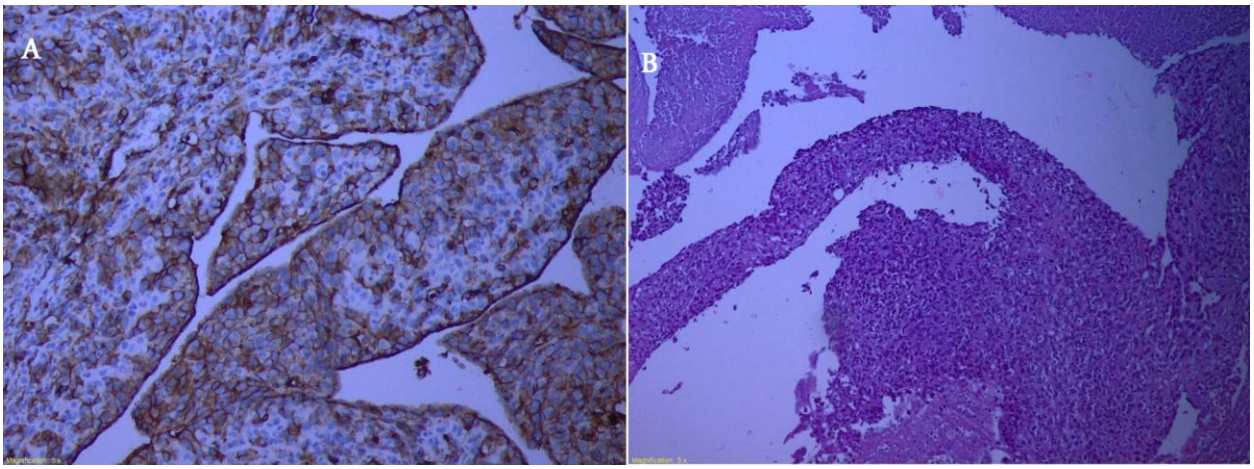


Figure.2A CD31 positive membranous and cytoplasmic immunohistochemical staining (Immunoperoxidase x 200); **B** Histopathological image (H&E x 100)



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