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CASE STUDY

PRIMARY CARDIAC CARCINOSARCOMA: A RARE ENTITY

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ABSTRACT

A case of primary cardiac carcinosarcoma of left atrium is presented because of extreme rarity and poor prognosis of the tumor. The patient was a 70 year old man admitted with sudden cardiac arrest and dyspnea. He was resuscticated and operated for a large left atrial tumor.

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INTRODUCTION

The malignant tumors of heart are divided in to cardiac sarcoma, lymphoma and tumor metastatic to the heart[1]. The cardiac sarcoma are very rare entity with angiosarcoma[1] being the most common sarcoma of the heart. The primary cardiac carcinoma is a very aggressive rare tumor of heart with only two case reports were found in the existing literature which were associated with very poor prognosis.

Case Report

A 70 year old male presented with short history of rapidly worsening dyspnea. By the time the patient reached the hospital he had a cardiac arrest. Patient was resuscitated and his condition was stabilized. The CT scan of the heart suggested a left atrial blood clot. The echocardiography showed a left atrial mass arising from the atrial wall with no septal attachment. The patient was operated for a left atrial tumor. Postoperatively he was recovering when he had a massive cardiac arrest from which he could not be revived. A clinical impression of cardiac sarcoma was made based on clinical and preoperative finding. Grossly the tumor measured 1x2.5x12 cms and grey white in appearance. On sectioning the tumor appeared fleshy and grey white with focal yellow is h areas. (Figure 1) No mucoid or gelatinous area could be identified.

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On microscopic examination a malignant tumor consisting primarily of high grade mesenchymal element. Tumor with a minor glandular carcinomatous component was appreciated (Fig. 2). The sarcomatous elements was homologous, the mesenchymal component consists of rhabdomyosarcoma element. The malignant rhabdomyosarcomatous cells consisted of highly pleomorhic bizarre to spindle shaped cells with numerous abnormal mitotic Figure.

No heterologous elements was identified. The malignant epithelial element was glandular. The glandular component was consist of cuboidal lining epithelium with nuclear stratification, pleomorphism, hyperchromasia and abnormal mitotic Figure in the glandular lining epithelium (Fig. 2). The alcian blue stain positively the glandular component staining the acidic mucin with in the cells confirming the glandular nature of the tumor (Fig. 2).

Immunoprofile

The glandular component was immunoreactive for epithelial membrane antigen antibodies and the mesenchymal elements was weakly positive for vimentin. The malignant sarcomatous component also showed strong desmin positivity (Fig. 2 & 3). The insert showing alcian blue positive mucin in the glandular elements. The alcian blue stain showing positivity for mucin in glandular component of the tumor (insert).

Fig. 1. The gross photomicrograph showing grey white soft tissue mass with cut surface showing grey white fleshy appearance with focal yellowish areas seen

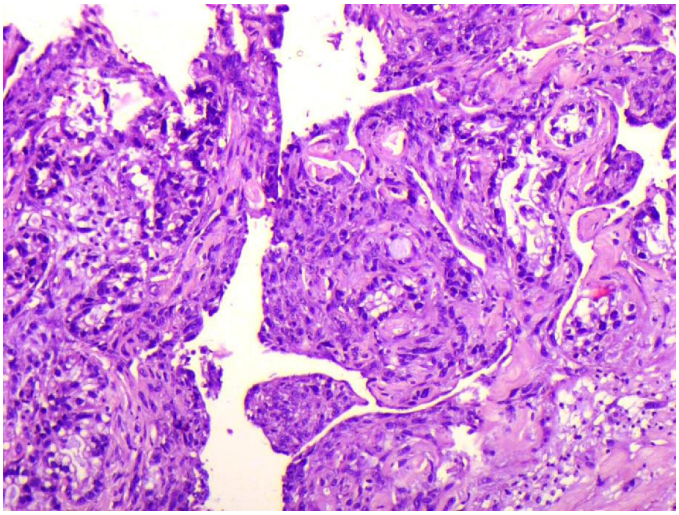

Fig. 2. H and E section shows severly pleomorphic spindle cells with epithelial component presenting as glandular element

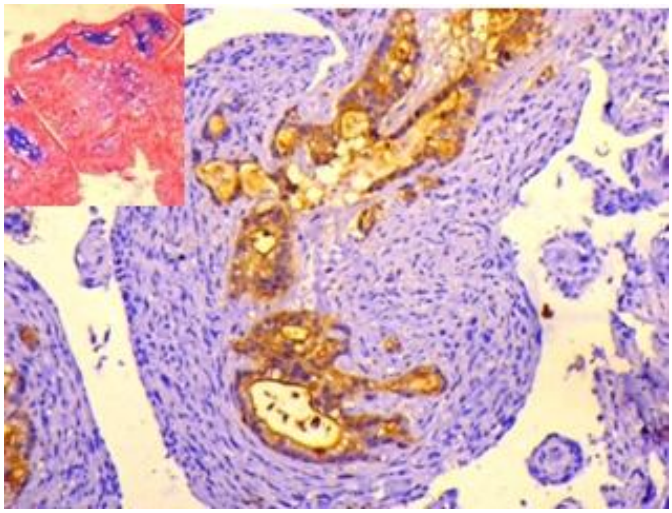

Fig. 3. The glandular epithelial component shows EMA positivity

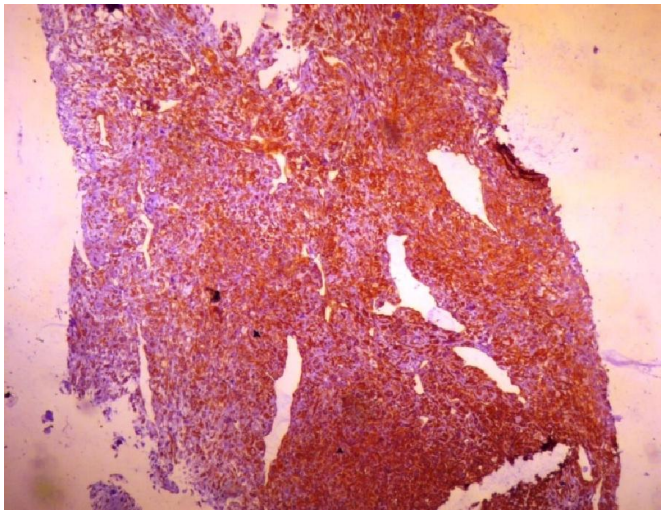

Fig. 4. The spindle cells of sarcomatous components shows strong positivity for desmin

DISCUSSION

The primary cardiac neoplasm are very rare tumor with estimated frequency of 0.0017- 0.019% with about 20% of neoplasm being primary cardiac sarcoma (Kim *et al*., 2011). The sarcomas commonly are angiosarcoma, epithelioid hemangioendothelioma undifferentiated pleomorphic sarcoma, fibrosarcoma and myxosarcoma, rhabdomyosarcoma, leiomyosarcoma and synovial sarcoma (Burke *et al*., 2002). The primary cardiac carcinosarcoma are extremely rare with only two previous cases reported by Ian Ramnarine, (2001), and Chen (1984). The present case was clinically diagnosed as myxoma. The sarcomatous component came out to be rhabdomyosarcoma with carcinomatous component consisting of adenocarcinoma. The grade of tumor was made as per criteria followed for grading of soft tissue sarcomas (Burke *et al*., 2002). The overall prognosis of primary cardiac tumors depends on the multiple of factor like location, size, friability, rate of growth and invasiveness of the wall and various structure invading the heart (Ibrahim *et al*., 2013). The average survival of patient being 6 months (Burke *et al*., 1992). Chen in 1984 presented the first case report of primary carcinosarcoma heart which was a left atrial tumor and presented clinically with features mimicking left atrial myxoma. Histologicaly the features defined were that of malignant counterpart of cardiac myxoma. Ramnarine *et al* in 2001 presented the second case report of a primary cardiac sarcoma in a 64 year old male presenting with dyspnea, chest pain, ankle swelling and weight loss. They found a invasive tumor in atria invading the walls and interatrial septum. The incompletely resected tumor showed sarcomatous component consisting of rhabdomyoblast, spindle cells and carcinomatous component consisting of tubuloacinar pattern. Radiological investigations like TTE, TEE, CT and MRT is of utmost importance in preoperative assessment of cardiac tumors (Auger, 2011). CT scan beside providing the morphology, location, and extent of a cardiac mass, also provide assessment of extra-cardiac disease or metastases (Dawson *et al*., 1990 and Hoey *et al*., 2012). MRI allows wide field of view, high contrast and spatial resolution, and

multiplanar imaging capabilities leading to precise demonstration and location of a mass, its anatomic relationship to cardiac chambers and extent of involvement and spread (Freedberg *et al*., 1988 and Randhawa *et al*., 2011). The cardiac sarcomas are of poor prognosis because it is not resectable often due to their large size, anatomic location and infiltrative growth pattern. Therapeutic approach involves debulking in most cases (Park *et al*., 2011). Thus the primary aim of surgery is palliative treatment in most cases. Although no long term study is available regarding prognosis of this tumor, studies involving cardiac sarcomas showed extremely poor survival of 11 months with median of 6 months (Burke *et al*., 1992). The primary carcinosarcoma showed high tumor grade with Zhang *et al* showed tumor grade to have a potential prognostic significance in cardiac sarcoma[14]. Treatment modalities like cardiac transplantation might provide better survival to these patients. In conclusion, primary carcinosarcoma heart clinical presentation is nonspecific and early diagnosis is often not reached. The investigations like CT and MRI were informative in establishing the tumor nature of mass but histological confirmation along with immune his to chemistry is only definitive way of establishing the tumor as primary cardiac sarcoma.

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