Case Report

DOI: 10.7860/JCDR/2015/12477.6563

Pathology Section

An Extemly Rare Case of Left Atrium and Right Pulmonary Vein Leiomyosarcoma

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ABSTRACT

We report a case of a 43-year-old female, presented with complaints of breathlessness for 1 month and had three episodes of syncopal attacks in the past two months. On clinical evaluation, bilateral pedal oedema was noticed. Echo cardiogram revealed large left atrial blood clot measuring 5.7x4.3x4.3 cm. Ultrasound whole abdomen was normal. We received an excised mass with right pulmonary vein end arterectomy speciemen. Histopathology of the mass revealed characteristic features of leiomyosarcoma with grade III, according to FNCCS grading system. The tumour was consistent with substantial amount of poorly differentiated fasicles of pleomorphic spindle cells and brisk atypical mitosis, with marked necrosis. Immunohistochemistry revealed the tumour cells in strong diffuse cytoplasmic positive for smooth muscle actin and Ki-67 showed 15-20% of tumour cells postivity. The prognosis depends on the individual tumour origin for individual site, size of tumour and depth of tumour than histological features. Pulmonary venous leiomyosarcomas were assumed to be misinterpretation of left atrial leiomyosarcomas with growth of the tumour into pulmonary vein lumen. We report this case in view of its extreme rarity.

Keywords: Primary cardiac tumour, Soft tissue tumour, Tumours of vascular origin

CASE REPORT

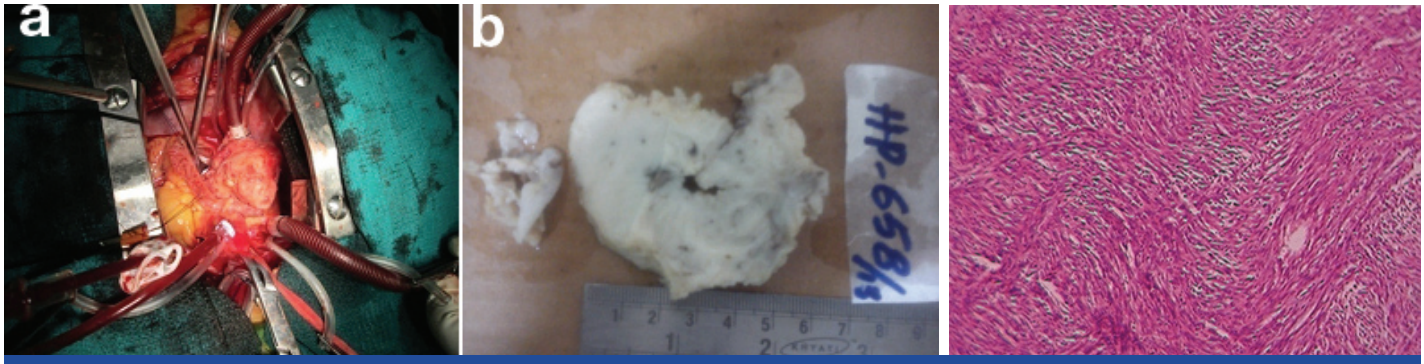
A 43-year-old female came to pulmonology department in GVP Institute of Health Care and Medical Technology, presented with dyspnoea on exertion since one month. She had a history of three episodes of syncopal attacks, in the past two months. For the past ten months, she is using anti-hypertensives. The patient did not have family or personal history of cardiac disease. On clinical examination, lungs were clear. S1 and S2 sounds were normal. Bilateral pedal oedema present. When she came to the hospital initially, Echocardiogram revealed grade II diastolic dysfunction. After two months, patient came with similar complaints to the hospital and we repeated Echo. It revealed large left atrial blood clot measuring about 5.7x4x4 cm. It extended into interatrial septum and right pulmonary vein. The mass was also obstructing mitral flow and mitral valve. Mild AR and PR, moderate TR with PAH and RV dysfunction was noted. Ultrasonography of the whole abdomen was normal. No mass lesions in the abdomen found. Chest X-ray showed cardiomegaly. Epicardial vessels were normal, as shown by coronary angiogram. Laboratory investigations were within normal limits. Serology was negative for HIV, HbsAg and HCV. A clinical diagnosis of left atrial myxoma or blood clot was made. Subsequently, surgical excision of Left Atrial mass with right pulmonary vein endarterectomy done.

Gross examination revealed a single, gray-white, firm to white mass, measuring 5.2x4x4 cm. A piecemeal specimen of Right Pulmonary vein endarterectomy was received. The cardiac muscle and pulmonary vein junction is obliterated by tumour. Hence, the junction cannot be demarcated.

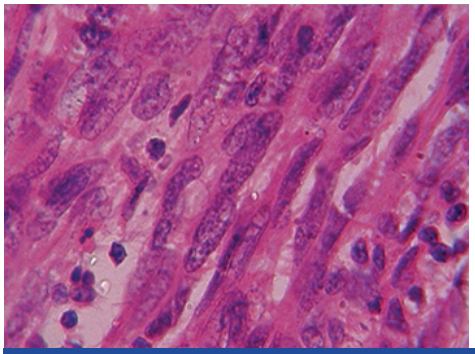
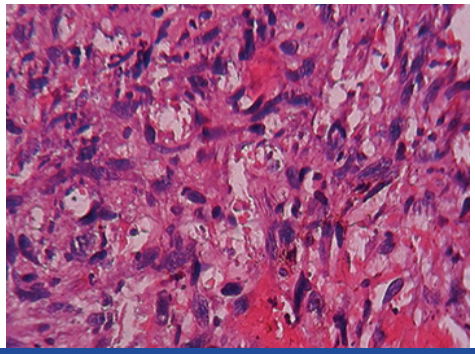
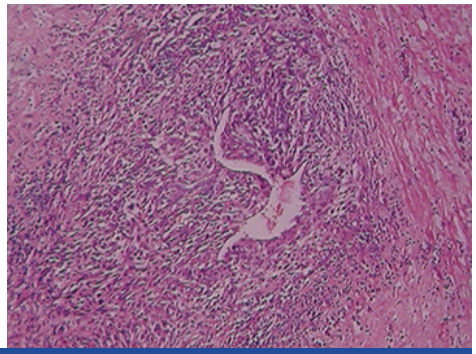
On cut-section, the borders of the mass were irregular and lobulated, whorled focal areas showed areas of haemorrhages and necrosis [Table/Fig-1a,b].

Microscopy: Multiple sections revealed moderate to haphazard arrangement of pleomorphic spindle tumour cell fascicles [Table/Fig-2]. Tumour cells showed delicate eosinophilic cytoplasm with paranuclear vacuoles [Table/Fig-3]. Greater degree of architectural disarray was noted. 'Cigar-shaped' nuclei were observed. Pleomorphic, bizarre, multinucleated giant cells with marked anaplasia noted. Brisk, atypical mitosis seen (>10 mitotic activity/10 HPF) [Table/Fig-4]. Tumour revealed Coagulative necrosis. (<50% in the examined area). The tumour cells are infiltrating into pulmonary vein endarterectomy specimen in microscopy sections [Table/Fig-5].

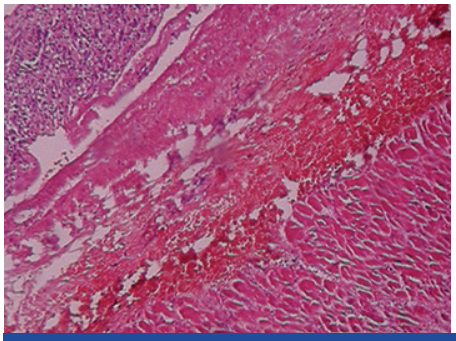
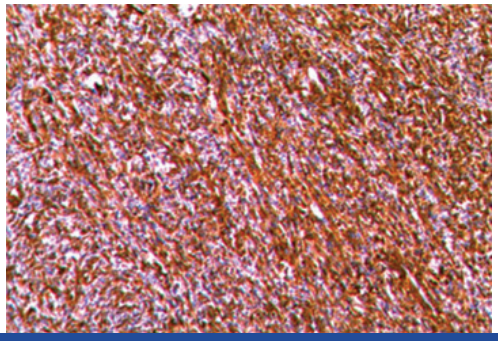
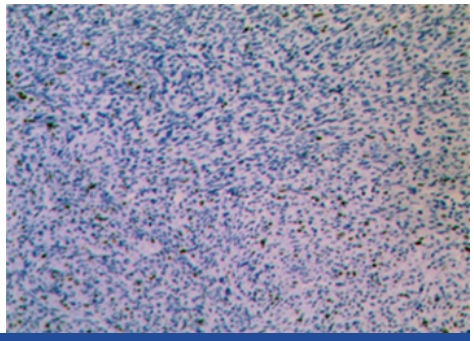
The sections of tumour mass peripheries showed tumour cells ingiltraion in to cardiac muscle [Table/Fig-6]. According to FNCCL grading, Tumour score was 6 and was under Grade III. Immunohistochemistry was carried out for tumour sections. CD34,



[Table/Fig-1a,b]: Intraoperative findings **[Table/Fig-2]:** Haphazard fascicles – 100x, H&E

[Table/Fig-3]: Blunt-ended nuclei - 100x,H&E **[Table/Fig-4]:** Marked anaplasia with atypical mitosis -400x,H&E **[Table/Fig-5]:** Tumour with pulmonary vein-100x,H&E

[Table/Fig-6]: Tumour with cardiacmuscle - 400x,H&E **[Table/Fig-7]:** SMA_100X, IHC **[Table/Fig-8]:** KI 67_100X, IHC

S100, Desmin were negative for reaction. Diffuse strong cytoplasmic positivity was shown by Smooth Muscle Actin [Table/Fig-7]. A total of 15-20% positivity was shown by tumour cells for Ki-67 [Table/Fig-8].

DISCUSSION

Leiomyosarcomas account for 5-10% of soft tissue sarcomas [1]. Mean age for patients with leiomyosarcoma is 45 years. The incidence is twice common in females [2]. Histologically, soft tissue leiomyosarcomas arising in different locations are similar. Based on the site of tumour origin, prognosis and treatment modalities differ. For this reason, soft tissue leiomyosarcomas are divided into several site related sub groups (based on the significant clinical and biological differences). Soft tissue leiomyosarcomas are less common when compared with leiomyosarcoma of uterine and GIT origin. Histologically, the typical cell of leiomyosarcoma is elongated and has abundant cytoplasm. The nucleus usually centrally located and blunt- ended nuclei or cigar–shaped nuclei. Smooth muscle cells show vacuole at one end of nuclei. Poorly differentiated tumour shows haphazard fasicles and marked anaplasia. Multinucleated tumour giant cells are common. Hyalinization is relatively common [3]. Myxoid pattern also noted in some tumour [4]. Criteria of malignancy in smooth muscle cell leiomyosarcoma include significant nuclear atypia and coagulative necrosis. Mitotic activity varies considerably. However, even low levels of mitotic activity (<1/10hpf) in the face of significant atypia, is sufficient evidence of malignancy. Histologically differential diagnosis of leomyosarcoma of smooth muscle cell tumours are spindle cell neoplasm, which includes leiomyoma, malignant fibrous histiocytoma, malignant peripheral nerve sheath tumour, fibrosarcoma and monophasic synovial sarcoma, although low-power appearance of all these tumours can be similar. Hence, cytological features in high power view and immunohistochemistry play a major role in differential diagnosis [5].

Localization of muscle antigens by means of immunohistochemistry has assumed more importance in diagnosis than electron microscopy over the past several years. Smooth muscle actin is detected in almost all leiomyosarcoma [6]. Leiomyosarcoma of vascular origin comprises an extremly rare group of soft tissue leiomyosarcoma [7]. Primary cardiac neoplasms are found to be very rare, its incidence is about 0.02% [8]. About 25% of cardiac tumours are malignant. Most of them are sarcomas. Primary cardiac leiomyosarcoma constitute <0.25%, and is the rarest [7]. To date, there have been only a few hundred published case reports of leiomyosarcomas of vascular origin. Predominant location is in inferior vena cava and right heart chambers, and less frequently in the pulmonary artery. An extremely rare type of this tumour is Leiomysarcoma of the pulmonary veins or left atrium [8]. Only 17 cases of leiomyosarcoma of the pulmonary veins and 20 cases of leiomysarcoma of the left atrium have been reported [9]. Pulmonary venous leiomyosarcomas were assumed to be misinterpretation of left atrial leiomyosarcomas with growth of the tumour into pulmonary vein lumen. This may be due to: 1) Difficulty intra operatively to determine site of tumour origin; 2) Special features of left atrial leiomyosarcoma is diffusion of tumour into the pulmonary vein lumen [10]. Leiomyosarcomas of the pulmonary veins, the pulmonary vein-left atrial junction and the left atrium is a uniform disease with the origin of the tumour in the smooth muscle cells located in the subendocardial/subendothelial layer of the left atrium and pulmonary veins [9]. Patient came for review check up after 3 months and repeated 2D echo, we noticed recurrence of tumour.

CONCLUSION

Leiomyosarcomas of the soft tissues include those found within the deep soft-tissue of the retroperitoneum, skin and subcutaneous tissue, blood vessels. An important concept to appreciate is that the microscopically similar smooth muscle neoplasms arising at different sites, do not always behave in the same manner. The establishment of criteria for malignancy has been difficult and required correlation of site-specific pathological findings with the patient outcome. Criteria for malignancy for individual soft tissue sites include tumour location, size, and mitotic rate.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Dec 09, 2014**
Date of Peer Review: **Mar 18, 2015**
Date of Acceptance: **Jul 05, 2015**
Date of Publishing: **Sep 01, 2015**