# Primary Left Ventricular Leiomyosarcoma - A Case Report

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**Abstract**

Cardiac leiomyoscarcomas is a rare subset of the already infrequent primary malignant cardiac neoplastic spectrum. The most common site for a primary leiomyosarcoma is the right ventricle with fewer than five globally reported cases in the left ventricle. Most present with non-specific symptoms but attention is usually sought after the appearance of compressive symptoms or arrhythmias. We present the case of a left ventricular leiomyosarcoma that had a delayed diagnosis and its subsequent surgical management.

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## Abstract

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## Background

A primary cardiac neoplasm is a rare diagnosis with a reported prevalence of between 0.001 to 0.03% in autopsy series. The most frequent primary tumors are angiosarcomas with the rarest being leiomyosarcomas.[1]

We present a case of an extremely rare primary cardiac leiomyosarcoma arising from the left ventricle which was surgically managed in the first instance.

## Case report

A 50 year old female patient with a background of systemic lupus erythrematosus presented to various hospitals with chest pain and shortness of breath over the last three years. The history was also significant for sweats, low grade temperatures and weight loss of seven kilograms. Unfortunately, despite multiple imaging attempts, a diagnosis was not made until recently when a trans-thoracic echocardiogram showed the presence of a mass either arising from the pericardium or ventricle. A left video assisted thoracoscopic (VAT) pericardial window and biopsy of the mass was done which did not show any evidence of malignancy.

A re-presentation with heart failure prompted a trans-oesophageal echocardiogram which demonstrated a mass compressing the left ventricle. A coronary angiogram was normal apart from feeding vessels into the tumour. Due to the multiple hospital admissions and new heart failure, a decision was made to excise the tumour.

A median sternotomy was performed and the patient placed on aorto-atrial bypass. A large mass arising from the left ventricle was mobilised (Figure 1) Cardioplegic arrest allowed a full thickness ventriculotomy and subsequent removal of the tumour. The defect was repaired with a continuous 2-0 Prolene stitch followed by overlocking 4-0 Prolene with a Teflon buttress.

The phrenic nerves were spared bilaterally. The patient was easily weaned off cardiopulmonary bypass but had to have the chest packed due to generalised coagulopathy. The chest was unpacked and closed primarily the following day after which she made an uneventful post-operative recovery.

Docetaxel and Gemcitabine chemotherapy was started by the treating oncology team after discharge from hospital.

## Pathology Report

Gross examination revealed a 65x45x32mm nodular mass attached to a portion of cardiac muscle (Fig 2). The cut surface showed an infiltrative solid white lesion containing areas of necrosis and haemorrhage (Fig 3). Histologic examination revealed a heterogeneous sarcomatoid malignancy with lobular architecture, widely invading the myocardium (Fig 4, 5 and 6). The constituent tumour cells were spindled and epithelioid, arranged in diffuse sheets and interlacing fascicles within a background of variably myxoid stroma. A spectrum of nuclear atypia was seen, including moderately pleomorphic blunt nuclei with vesicular chromatin transitioning to areas with marked cytologic atypia. Frequent mitotic figures were identified, numbering up to 32 per 10 high power fields (Figure 7). Broad zones of necrosis were present and lymphovascular invasion was identified.

Immunohistochemistry revealed the tumour cells to be positive for smooth muscle actin and desmin (Fig 8 and 9), with only focal positive staining for pan-cytokeratin and AE1/AE3. The tumour cells were negative for CK7, CK20, EMA, myogenin, MYO D1, STAT 6, calretinin, CD31, CD34, ERG, SOX 10, S100 and p40.

The morphologic features and immunohistochemical profile were in keeping with a high grade cardiac leiomyosarcoma.

## Discussion

Primary cardiac tumours are exceedingly rare neoplasms comprising less than 1% of the 0.001-0.28% of all reported cardiac malignancies. The left atrium is the most common site reported in literature with the left ventricle being the rarest cardiac site[2,3]. We note that our report may be the fifth in the global literature in any form describing a primary left ventricular leiomyosarcoma.

Diagnosis has been reported to be straightforward with echocardiography, CT and MRI having very high sensitivity.[3]Unfortunately as is highlighted by our case, vague symptoms with the differential diagnosis cofounded by the presence of chronic disease can lead the clinician into ordering imaging that does not focus on the heart. In a particularly unfortunate turn of events in this case, a biopsy of the mass via VAT returned inflammatory tissue. Regardless, we recommend the use of echocardiography and CT to aid in the diagnosis if vague cardiac symptoms are reported.

Wang et al. reported that surgery was the most common therapy provided followed by adjuvant chemotherapy. Incomplete resection was a feature of many of these cases due to the invasive nature of the tumour.

Prognosis has been universally reported as poor with Wang et al noting that the five year survival rate was 25.4%. Adjuvant chemotherapy was also noted to improve outcomes in many studies.[2,3]

## Conflicts of Interest

The authors report no conflicts of interest.

## Contributions

VB and AJ were responsible for conception, drafting and critical revision of the article. VY and TB were responsible for conception and critical revision of the article.

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