

# Cystic lymphangioma of heart

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## Abstract

Primary cardiac tumours are rare and myxomas are the most frequent. Cystic lymphangiomas are most often found at cervicomediastinal locations and are exceedingly rare in the mediastinum. We report a case of cystic lymphangioma involving the right ventricle and the right atrium mimicking a mediastinal tumour. Complete resection was performed via a median sternotomy under cardiopulmonary bypass. Histopathological examination of the tumour mass was undertaken and was diagnosed as a cystic lymphangioma.

## INTRODUCTION

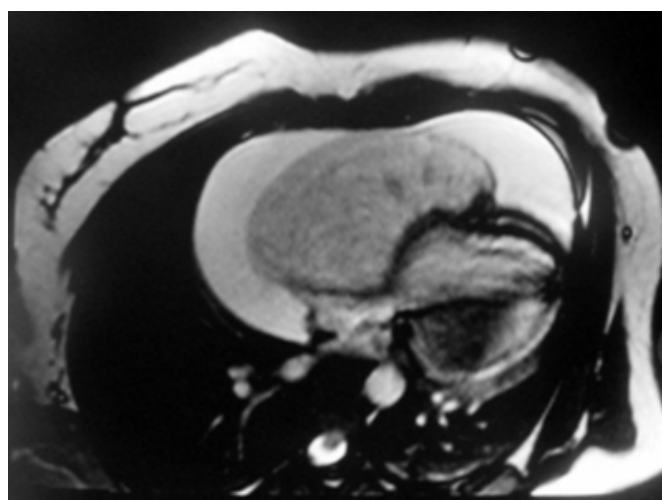
Primary cardiac tumours are rare, their incidence ranging from 0.0017% to 0.33% at autopsy.<sup>1</sup> Cystic lymphangioma (CL), which is usually confined to the head and neck, is a well recognized tumour that occurs during childhood. However, a cardiac lymphangioma is exceptionally uncommon and a particularly rare form of cardiac disease.<sup>1</sup> Here we report a case of cystic lymphangioma presenting as a cystic mass in the right atrium and the right ventricle.

## CASE REPORT

A 32 year-old female presented with breathlessness on exertion for two months associated with productive cough and heaviness in chest with a dragging sensation and discomfort radiating towards the right lateral side over the same period of time. Past medical history consisted of complains of easy fatigability and vague light headedness since 4 years. She weighed 72 Kg and physical examination was normal. Baseline biochemical and haematological tests were within normal limits. Patient was then extensively worked up with all forms of imaging including 2 dimensional Echocardiography, a CT scan (Figure-1) and an MRI. The possibility of an epicardial cyst or loculated pericardial effusion or cardiac tumour compressing right atrium and right ventricle, was entertained for which a limited right heart study was done which did not reveal any intra cavity connection. Intraoperative transesophageal echocardiogram (TEE) demonstrated a mass of variable echogenicity filling the entire pericardial cavity. No valvular or regional wall motion abnormality was noted. The tumour was exposed by primary median sternotomy under total cardiopulmonary bypass.

## Figure 1

Figure-1 CT Scan showing mediastinal pericardial cystic mass

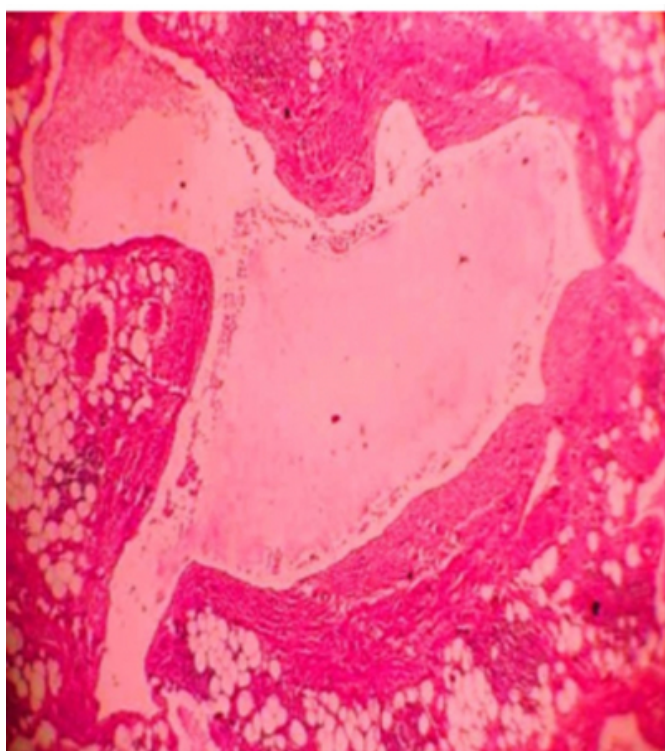


The pericardium was opened and a pinkish brown mass covering the entire right ventricle was seen. On closer look there were two components of the mass; one smaller measuring 4×2.5cm extending over the aortic root from the inner curvature of the heart and other much larger measuring at least 10×6cm in the longitudinal and transverse planes covering the sternocostal surface of the right ventricle and most of the body of right atrium leaving ascending aorta and a bit of the adjoining right atrium free. Inferiorly the mass extended up to the inferior vena cava but did not involve it. The tumour was excised as much as possible. The post operative course was uneventful. The tumour mass was sent for the histopathological examination to our department. On gross pathological examination the tumour pieces altogether measured 10×5×2cm. The tumour had variable consistency

being soft and spongy at places and firm and fibrous at others giving a honeycombed appearance. The cysts were small and were filled with clear straw coloured fluid. The cyst wall was smooth and no hard or nodular area was identified. Microscopy revealed interconnecting channels lined by flat endothelial cells with smooth muscle bundles and lymphoid infiltrate underneath [Figures 2 & 3]. The absence of red blood cells in the cyst contents ruled out hemangioma .

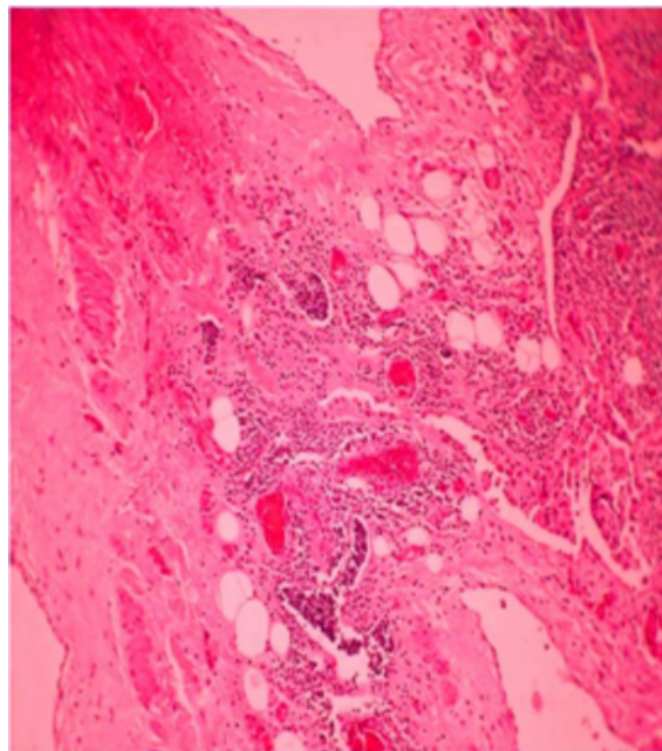
**Figure 2**

Fig-2. Cystic lymphatic space with surrounding adipose tissue and lymphoid follicles.(H&EX100;)



**Figure 3**

Figure-3. Tiny lymphatic spaces filled with lymphocytes. Myocardial tissue is seen on the left (H&E X400)



## DISCUSSION

Cardiac lymphangioma is a very rare tumor of the heart, first reported in 1911 by Armstrong and Monckeberg. Only nine cases of cardiac lymphangioma have been reported in the medical literature. A review of reported cases of CL shows locations in the neck in 75% of cases, in axilla and shoulder joint in 15%, and in other sites (cervicomedial location, retroperitoneal area, in the spleen or the colon, esophagus and chest wall) in 10%. In the thorax they represent 0.7–4.5% of all mediastinal tumours in the adult population. Mediastinal lymphangiomas are most often asymptomatic masses incidentally discovered on chest X-ray. However, cardiac lymphangiomas are commonly revealed during congestive heart failure, syncopal or embolic pathology, arrhythmias, palpitations, or cardiac tamponade<sup>2,3</sup>. CL is typically a multiloculated lesion with cystic cavities divided by septa of variable thickness. On gross pathological examination, cardiac lymphangiomas may be either soft and spongy or firm and fibrous. The size of these tumours varies, and the largest one reported was 9 cm in diameter. The tumours most commonly occur in the pericardial space, but other unusual primary sites include the myocardium, the posterior wall of the left atrium, and the AV node regions.<sup>4</sup> Although the head, neck and axilla are sites that account for

one half to three fourths of all lymphangiomas, these masses may affect almost any part of the body including the heart, as in the current case.<sup>5,6,7</sup> Histopathology shows interconnecting channels lined by flat endothelial cells beneath which are bundles of smooth muscle and lymphoid nodules. The absence of red blood cells in the cyst contents eliminates hemangioma or lymphangiohemangioma.<sup>8</sup>

### CONCLUSION

Cardiac lymphangiomas can be a rare presentation and must be in mind of a pathologist, especially a cystic lesion in this area. Lymphangioma although a benign lesion but can abut the cardiac functionality due to space occupation and pressure atrophy of the heart.

### References

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