Incidental Finding Of Adult Cardiac Lymphangioma: Report Of A Rare Case And Current Management Strategies

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Citation

Abstract
To report the rare occurrence of cardiac cystic mass detected intra-operatively despite the best of surgical work up preoperatively. The incidental finding of cardiac lymphangioma during routine cardiac bypass graft in an adult is rare but this needed to be dealt with expediently to avoid latter complications or need for operation in later date.

INTRODUCTION
Lymphangioma is a rare benign abnormality usually confined to head and neck and is a well-recognised occurrence in children . Less than 2 % of lymphangioma is located in thoracic area and diagnosis of cardiac lymphangioma is extremely rare . To date, less than 100 cases worldwide have been reported on this rare condition.

CASE REPORT
A 72 years old gentleman presented with acute coronary syndrome typified by ST depression and T wave inversion on the lateral chest leads with no troponin elevation. Subsequent angiogram study revealed diffuse vessels disease involving distal LMCA, ostial LAD, ostial circumflex and RCA.

The patient's other co-morbidities includes atrial fibrillation, chronic obstructive pulmonary disease, previous heavy smoker, hypercholesterolemia, hypertension, open abdominal aortic aneurysm repair and prostate carcinoma.

The patient had an echocardiogram done in 1997 showing mildly dilated left ventricle with ejection fraction of 67%. The right ventricle was normal and there were no abnormal contractility or mass seen.

An elective 4 vessels coronary arterial bypass graft using LIMA and LSV grafts was scheduled. However when the pericardium was opened, a hemorrhagic cystic mass was found situated on the atroventricular groove of the right ventricle and was slightly compressing the tricuspid valve. Post completion of coronary bypass graft, the cystic mass was excised with some margin of normal right ventricle muscle and the defect was repaired with 4/0 prolene sutures and Teflon pledgets. The patient was easily weaned off cardiopulmonary bypass machine and made an uneventful recovery.

The histologic examination of the excised mass showed an ovoid cystic structure measuring 40x30x15mm with thick trabecular structures protruding into the lumen of the cyst. There were occasional bundles of smooth muscle with aggregates of lymphoid cells infiltration within the cyst wall (Figures 1 and 2). These features were consistent with the diagnosis of cardiac lymphangioma.
DISCUSSION

Lymphatic system is an extensive uni-directional system of blunt ending vessel that transport excess fluid into the venous system as well as act as a site of absorption of protein and lipid from the entero-hepatic circulation. Lymphangioma is therefore a hamartomatous malformation of the lymphatic vessels. The current literature categorised lymphangioma into 4 groups: (1) lymphangioma simplex or circumscriptum; (2) cavernous lymphangioma; (3) cystic lymphangioma or cystic hygroma and (4) diffuse lymphangiectasia. Most lymphangiomas are usually detected in children under 2 years old. It appears commonly as a cervical mass posterior to sternocleidomastoid muscle (75%) or as axillary or shoulder masses (20%). Lymphangioma extending into the mediastinum is not very common. Primary cardiac tumors are rare with an incidence of 0.002-0.28% and cardiac lymphangioma is one of the most rare heart tumors. The detection of cardiac lymphangioma in adult is less common than in paediatric age group.

The presentation of cardiac lymphangioma is usually variable. The symptoms can range from arrhythmias, palpitation, syncope, pericardial effusion to sudden death. Majority of symptoms are attributed to their size and critical locations. However, incidental finding of these tumors have been reported as well.

There are several investigations to diagnose cardiac lymphangioma. Echocardiogram shows an ill defined cystic or echogenic mass that corresponds to cluster of abnormal lymphatic channels. CT, MRI or EBCT may help to define the location and delineate the extent of involvement as well as exclude other differential diagnoses such as hemangioma, cystic teratoma or intramural thrombus.

As spontaneous increase in size of cardiac tumor can induce severe symptoms and the treatment is usually interventional. Adjuvant treatments such as radiotherapy or injection of sclerosant agents have been proposed and used but these are controversial due to risk of inflammation and swelling that may leads to heart failure as well as local recurrences such as fistula formation or infection. Complete and extensive surgical excision is the accepted modality of treatment as cardiac lymphangioma has a potential risk of recurrence. Surgical consideration in excising this tumor should take into account preserving adequate myocardium, maintaining proper atrioventricular valve function and preserving myocardium conduction system to avoid replacement of atrioventricular valve or implantation of pacemaker. In our case, we elected to resect the tumor to avoid potential risks of tumor fragment dislodgement that could embolise peripherally or micro metastasis.
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Dispersement leading to peripheral seedling. Resection of tumor also allows adequate inspection of other cardiac chambers for additional lesions. Provided that the tumor is completely resected, a good prognosis can be expected. Postoperative follow-up should be carried out using echocardiography to evaluate myocardial wall as well as recurrence of tumor.

The findings of this study are worth reporting due to the rareness and incidental finding of the cardiac lymphangioma during routine cardiac bypass graft in an adult. This case illustrates that unexpected finding of cardiac tumor mass should be dealt with expeditiously to avoid later complications or need for operation in later date.

References
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