A Left Ventricular Myxoma arising from the Anterior Papillary Muscle in an 11 Years Old Male Child: A Case Report

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Abstract:
Primary cardiac tumors are mostly myxomas but are very rare. Among them, left ventricular myxomas are extremely rare and account for 2.5% of all cardiac myxoma cases. A left ventricular myxoma was diagnosed by Transthoracic two-dimensional echocardiographic imaging in an 11-year-old male. It was found originating from the anterior papillary muscle of left ventricle and extending towards the left ventricular outflow tract (LVOT). The tumor was successfully resected through the mitral valve by trans-atrial septal approach. Histopathology confirmed the diagnosis of myxoma. We report this case for being a rare entity of myxoma arising from the anterior papillary muscle of the left ventricle and to our knowledge this is the only reported case of its kind in the world.

Keywords: Myxoma, Interventricular Septum, Left Ventricle

Introduction:
Myxoma is a rare cardiac tumor. Approximately 80% of myxomas are localized in the left atrium, 75% of which involve the interatrial septum. Between 7% and 20% are found in the right atrium; the rest are either biatrial, in the right ventricle, or in the left ventricle.

The first description of a primary intra cardiac tumor was in 1559, located in the left ventricle. In a review of literature, in 1982, Mazer and Harrigan by 2D echocardiography reported the first case of LV myxoma¹. Up to 1996, only 37 cases of left ventricular myxomas have been reported and 72 cases up to 2014.

It is thought that myxoma arises from the embryonic residues after the in-utero septation of the heart². Thus, cardiac myxomas may originate from anywhere within the cardiac chambers. Majority of cardiac myxomas occur in the atria with only 3% - 10% arise in either the left or right ventricle. These tumors usually project from the endocardium into the cardiac chambers.

Patients with myxomas can have a wide range of symptoms e.g. palpitation, dyspnea, fever or stroke. Embolic accidents have been reported frequently, which can cause disability or sudden death³.

A left ventricular myxoma originating from the anterior papillary muscle of the left ventricle and extending towards the left ventricular outflow tract is very uncommon and so this case had been reported.

Case report:
We are presenting a case of an 11-year-old South East Asian male patient who presented with the complaints of fever, palpitation and extreme fatigability after moderate exertion. No history of headache, convulsion or faintness were given. A systolic murmur was present in the third and fourth intercostal spaces along the left sternal edge, and in the second right intercostal space.

Transthoracic echocardiography demonstrated a large pedunculated pear-shaped mass (26 x 17 mm in diameter) in the left ventricular cavity, attached near the apical portion of the interventricular septum, extending into the outflow tract (Figure-1). LV function was normal. The other cardiac valves and cavities were free of lesions. Cardiac CT revealed a pear shaped mass in the LV, attached near the interventricular septum and extending towards the LVOT (Figure-2). The patient was at an increased risk of sudden death and systemic embolization of the mass through the aortic valve. So, an urgent resection of the intra cardiac mass was required.

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The patient underwent median sternotomy under general anesthesia and standard cardiopulmonary bypass was established. After right atriotomy and atrial septostomy, we visualized the gelatinous fragile mass on the interventricular septum and partially attached to the papillary muscle. Extraction of the myxoma through the mitral valve and small part of papillary muscle was accomplished (Figure-3,4). Peroperative saline test and postoperative TEE revealed no mitral regurgitation with no residual mass. Histopathologic examination confirmed the diagnosis of myxoma (Figure-5).

**Figure-1:** 2D echocardiography reveals LV myxoma arising from LV cavity

**Figure-2:** Cardiac CT reveals the myxoma arising from left ventricular side of interventricular septum

**Figure-3:** Peroperative view of the myxoma

**Figure-4:** Myxoma after excision

**Figure-5:** Histopathology confirms the diagnosis of LV myxoma

**Discussion:**
Cardiac myxomas are found to be associated with embolism, obstruction to the outflow tract, arrhythmia and other constitutional symptoms. Embolism occurs in 30% to 40% of all cases and the risk of embolization mainly depends on the morphology of myxoma rather than its size. Cardiac myxomas are of two types: (1) friable polypoid type and (2) smooth-surface rounded type. The polypoidal type is friable in consistency and intracavitary in location so tends to result in embolism4. Cardiac myxomas recur more often in young men and up to 40% of patients who have a family history of the tumor5. The hospital mortality rate of solitary myxomas after surgical resection is very low with excellent prognosis. Late recurrences have been reported to be 0.4–5% of surgically treated patients after 3 months to 22 years of operation6. The clinical presentation of patients with myxoma is commonly nonspecific and early diagnosis of the myxoma may be a challenge. The classical triad of obstruction, embolic and systemic manifestations are rarely present. Nonspecific symptoms such as recurrent fever, anaemia, arthralgia, weight loss mostly arise
from the elevation of IL-6 and disappear after removal of the tumor. This patient presented with vomiting episodes, a non-productive cough, mild fever and fatigability. When cardiac conducting system is involved, arrhythmia, mostly atrial fibrillation is present which is associated with heart failure. None such condition was observed in our patient.

**Conclusion:**
To our knowledge this is the first case of LV myxoma originating from the anterior papillary muscle of the left ventricle not only at the department of cardiac surgery, BSMMU but also in the world. Transthoracic echocardiography is sufficient for making a diagnosis. Surgical resection is the treatment of choice, having relatively low-risk with excellent postoperative prognosis for most patients.

**Conflict of interest:**
The authors declare that there is no conflict of interests regarding the publication of this paper.

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