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Clinical Cardiology Congress 2019: Left atrial myxoma with severe pulmonary hypertension in young girl

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Abstract

The association between disorder and depression is well- 17 year old girl presented to hospital with history of high fever and worsening of shortness of breath which she had for 3 months. On examination she had a pan systolic murmur best heard at apex, elevated JVP and a mild tender hepatomegaly was found. ECG was normal and CXR showed biatrial dilatation and pulmonary congestion. transthoracic 2D echocardium followed by TOE performed and showed EF> 50% dilated LA with large LA myxoma with a central hypoechoic area measuring 32mm x 60 mm. it is attached to IAS and highly mobile. There was grade II MR. There is significant TR with TRPG of 94mmHg with severe pulmonary hypertension. RA,RV mildly dilated and TAPSE 15. Patient underwent surgical excision of left atrial myxoma next day. on day 2 post surgery TRPG was 28mmHg.The cause of pulmonary hypertension in left atrial myxoma patients is due to mitral stenosis like pathology of the myxoma. Pulmonary arterial hypertension in these patients were also explained by rise in IL-6 in patient with atrial myxoma. It has shown that IL-6 is a valuable biomarker. Right sided myxomas are rare to develop pulmonary hypertension but it has been described. The cause of pulmonary hypertension in these patients are mostly due to chronic micro thromboembolic phenomenon of the tumour and associated with cytokines such as IL-6 directly entering the pulmonary circulation. Because the pulmonary hypertension in right heart myxoma caused by a chronic change in the lung vasculature, pulmonary hypertension may not resolve after resection of tumour but usually it does in left atrial myxomas.

Myxoma is the most common primary cardiac tumour, found in the left atrium in 75% of cases.1,2 Clinical features range from being asymptomatic to symptoms of mitral stenosis, embolisation and systemic illness.3-5 Pulmonary complications, including pulmonary hypertension,4,6,7 pulmonary infarction8 and lymphadenopathy,9 though uncommon, have been reported. Our patient presented with a pulmonary triad of all the complications mentioned above and all of them resolved immediately following successful excision of the tumour.

We hope to report this case for educational and clinical purposes, due to the unusual combination of pulmonary complications, as well as atrial arrhythmias associated with a left atrial myxoma.

Discussion:

Primary cardiac tumours are rare, with an incidence reported to be below 0.03%.1 By comparison, metastatic involvement of the heart is significantly more frequent and has been reported in up to one in five patients who died of cancer on autopsy.1 Of all the first cardiac tumours, myxoma is that the commonest (70–80%).9 Myxomas tend to be more frequently seen between the third and sixth decades of life, with a female-to-male ratio of 2:1 to 3:1.1,2,10

About 75% of all myxomas occur within the LA and typically originate from the interatrial septum within the fossa ovale region; but a fifth are found within the right atrium, and 5–8% within the ventricles.5 Other locations are reported, though much rarer.1

The size of a myxoma can vary from a couple of millimetres to fifteen cm, with an averaged maximal dimension of three .7 to 5.6 cm.10 Therefore, the myxoma found in our patient was far larger than most reported and, thus, are often classified as giant or colossal.

The clinical features of a cardiac myxoma are associated with its location, size and mobility.

The tumour can present with one or more symptoms of the three categories: outflow obstruction of the mitral valve, embolism and constitutional or systemic symptoms.4,5,12 In our patient, embolism manifested as pulmonary infarction.

The most common symptoms of los angeles myxomas are associated with bicuspid valve obstruction (>60%): dizziness or syncope, palpitations, dyspnoea, cough, pulmonary oedema, or congestive coronary failure . Embolic manifestations are less common (16%). A similar percentage of patients (15%) may present with systemic or constitutional symptoms such as myalgia, muscle weakness, arthralgia, fever, weight loss, fatigue, and even Raynaud syndrome. he detection of it has become more frequent, likely due to the easy access and wide availability of echocardiography.10 The fact that our patient presented to us rather late, and the large myxoma size, suggested an extended period of growth. Hence, he had developed a good range of symptoms along side the pulmonary complications.

Atrial fibrillation has been described as a complication of a LA myxoma,9,10,12 and it are often a consequence of either the myxoma itself or the surgery . In the former case, fibrillation is resolved after a successful operation, but it remains difficult to determine a transparent association between the 2 , although such association is well known between rheumatic mitral stenosis and atrial fibrillation. It would not be illogical to state that the large myxoma in our patient caused significant mitral valve obstruction and lead to the initial atrial fibrillation, and even subsequent atrial flutter after surgery.

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