An unusual presentation of right atrial myxoma: a case report

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ABSTRACT: A right atrial myxoma extending to inferior vena cava (IVC) with associated right-to-left shunting leading to systemic desaturation is an exceedingly rare clinical entity. The number of such cases reported in literature till date is not more than five. This case study presents a 45-year lady who was referred to our Centre with symptoms of breathlessness, easy fatigability, generalized weakness and *central cyanosis*. On routine blood investigation polycythemia was detected. On preoperative 2D-Transthoracic Echocardiography (TTE), the diagnosis of right atrial mass was confirmed but reason of cyanosis was not revealed. Tumor thrombus in a branch of right lower pulmonary artery with small locoregional pulmonary infarct along with right atrial myxoma was reported in CT angiography of heart and great vessels. On-table Transesophageal Echocardiography (TEE) showed patent foramen ovale (PFO) with right-to-left shunting because of raised right atrial pressure and dynamic obstruction of the tricuspid valve by the mass. The myxoma was resected completely via right atrial approach along with primary closure of PFO. Post-operative period was uneventful, and she was discharged on day 7. The Patient is doing well during the 3-month follow-up.

KEYWORDS: atrial, mass, myxoma, cyanosis, shunting

INTRODUCTION: Right atrial myxoma extending to IVC with right-to-left shunting resulting in systemic desaturation is rare. Here we will present a right atrial myxoma with IVC extension with *cyanosis* in a 45-year female after proper informed and written consent obtained from the patient.

CASE REPORT: A 45-year female was referred to our Centre with symptoms of shortness of breath, easy fatigability, generalized weakness and bluish discoloration of lips, fingers and skin for about four months. On general examination patient was malnourished and thin built; central cyanosis was present. On auscultation grade 3/6 diastolic murmur was audible at tricuspid region. Other systems were normal.

Polycythemia was detected in routine blood investigation. Arterial blood gas (ABG) analysis showed pO2 of 32mm Hg and SpO2 of 68% at room air and 72% on oxygen (nasal prong/face mask). Chest X-ray revealed no abnormality. 2D-Transthoracic Echocardiography (TTE), the diagnosis of right atrial mass was

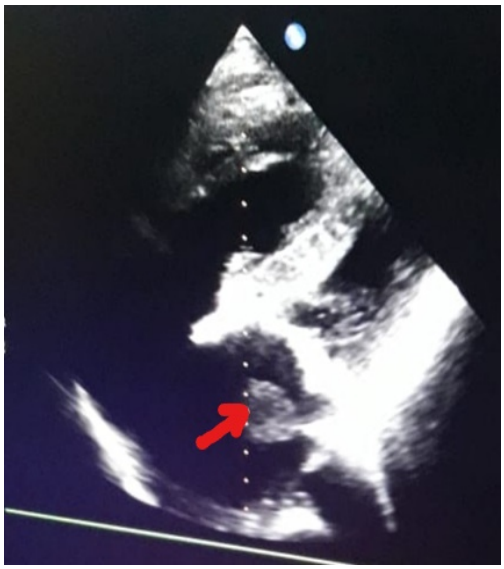

Figure 1 *Transthoracic Echocardiography showing right atrial mass (red arrow)*

confirmed [Figure 1] but reason of cyanosis could not be explained. Tumor thrombus in a branch of right lower pulmonary artery with small locoregional pulmonary infarct along with right atrial myxoma was reported in CT angiography of heart and great vessels and CT pulmonary angiography (done outside). Cardiac catheterization study was not possible due to ongoing COVID pandemic and institutional COVID-19 testing protocol. We still had diagnostic dilemma between right atrial myxoma with right-to-left shunting and pulmonary thromboembolism.

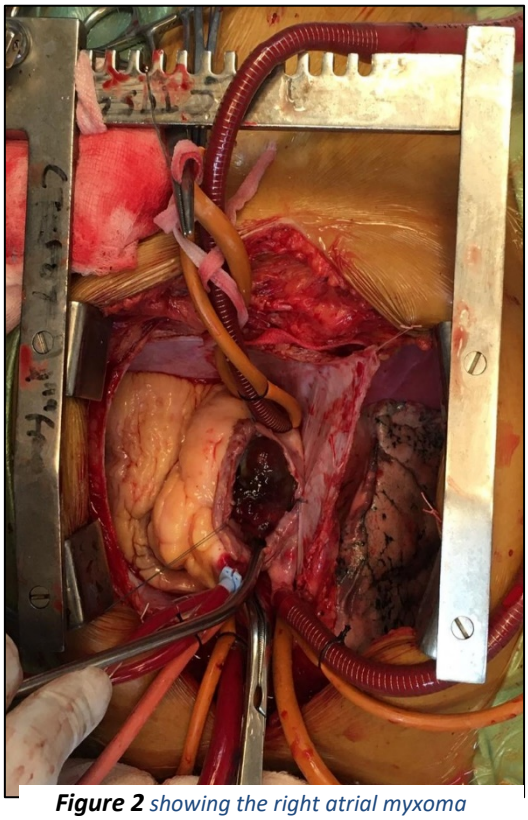

Figure 2 *showing the right atrial myxoma*

We planned surgical resection of the myxoma on earlier basis. On-table intra-operative Transesophageal Echocardiography (TEE) done under anesthesia showed patent foramen ovale (PFO) with right-to-left shunting because of raised right atrial pressure and dynamic obstruction of the tricuspid valve by the right atrial myxoma. Our approach was via median sternotomy. Standard aorto-bicaval cannulation was done. Cardiopulmonary bypass was established. The myxoma was pedunculated and it was attached to anteromedial aspect of RA-IVC junction with extension to IVC [Figure 2]. The myxoma was resected via trans-RA (right atrial) approach. IVC extension of the tumor was removed by going into total circulatory arrest (TCA). The PFO was closed primarily [Figure 3]. The surgical specimen was sent for histopathological examination and came out to be a myxoma.

Post-operative period was uneventful and the saturation at room air of the patient was 100% and she was discharged on day 7. The Patient is doing well during the 3-month follow-up period.

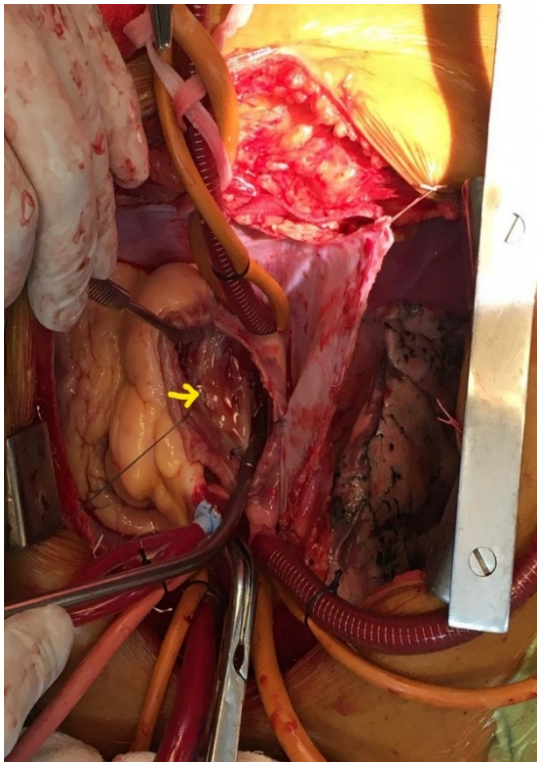

***Figure 3**: The yellow arrow showing the PFO*

DISCUSSION: Myxomas are the most encountered primary cardiac tumor in adults; the left atrium is the most common site. Right atrial myxomas are more sessile and solid than left atrial myxomas, with wider attachment to the septum or atrial wall. Left atrial myxomas produce symptoms of dyspnea and hemoptysis resembling features of mitral stenosis, commonly of short duration, episodic, and with syncope, rapidly progressive ended up with heart failure. Right atrial myxomas may present with episodic symptoms and may progress rapidly. Abdominal distension from hepatomegaly and ascites and peripheral edema are often presenting complaints. Embolic manifestations neurologic deficits, coldness and pain in an extremity, angina(coronary), and dyspnea(pulmonary). Constitutional symptoms are often subtle or even absent also when the tumor bulk is small, but occasionally become the only presenting symptom complex [1]. Newman et al. reviewed 312 cases of right and left atrial myxomas, out of which only two were associated with atrial septal defect [2]. Natarajan et al. has reported one case [3]. Right atrial myxoma is rare and the usual signs are right-sided heart failure and pulmonary embolism [4]. The association between a myxoma and significant right-to-left shunting at atrial level in adults is exceedingly rare [5]. Our patient was cyanotic at the time of presentation because of the right-to-left shunting at the level of atria via PFO because of raised right atrial pressure in the background of dynamic obstruction of the tricuspid valve by the right atrial myxoma.

DECLARATIONS:

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Informed consent: Written informed consent was obtained from the patient for her anonymized information to be published in this article.

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