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Case Report

A rare and independent association of right atrial myxoma with immune thrombocytopenic purpura

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ABSTRACT

Primary tumours of the heart are rare and the most common benign ones are myxomas. The clinical features are varied and include a myriad of presenting symptoms like embolic, constitutional, cardiac and also symptoms due to obstruction. Right atrial myxomas are very rarely seen when compared with left atrium. Such myxomas independently co existing with Immune Thrombocytopenic Purpura (ITP) is even rarer and we now present one such case in with right atrial myxoma was associated with ITP and was managed surgically. This case emphasises the fact that ITP can co-exist with myxoma and should be borne in mind when treating such patients surgically.

Keywords: Immune, Myxoma, Right atrium, Thrombocytopenia

INTRODUCTION

Intra cardiac tumours are very rare and myxomas are one such benign common tumours of the heart.[1] Usually left sided they are and right sided myxomas are even more rare occurring in the ratio of 1:4. The mechanical hemolytic effects of myxoma usually result in thrombocytopenia and sometimes there can be independent ITP existing in a myxoma patient.[2] We describe one such case wherein the thrombocytopenia was not due to the myxoma per se, but rather separate entities of myxoma and ITP existing independently in a single patient.

CASE REPORT

A forty-year-old normotensive euglycemic male, was diagnosed with immune thrombocytopenic purpura (after excluding all the causes for thrombocytopenia with connective tissue disorder work up being negative) fifteen years before and he underwent splenectomy and was on regular follow up with haematologist for the same. At that time there was no history of myxoma as an echo cardiography done fifteen year before mentions normal right atrial chamber with no intra cardiac tumours. Three months before he presented with dyspnoea on exertion New York Heart Association class II-III and chest pain for which he was referred to our centre for further evaluation. On further evaluation, Echocardiogram revealed a 5x3 cm mass (Figure 1). attached to the inter atrial septum in the right atrium protruding into the right ventricle through the tricuspid valve with features suggestive of myxoma (incidental diagnosis).

There was no tricuspid regurgitation with good biventricular function. His platelet count was 2 lakhs/cubic millimetre on admission. He was not on any immunosuppressive therapy or steroids as his haematological investigations were within normal limits.

It was decided to transfuse platelets in case of excessive bleeding and if the platelet count is <1 lakh/ cu mm post operatively.

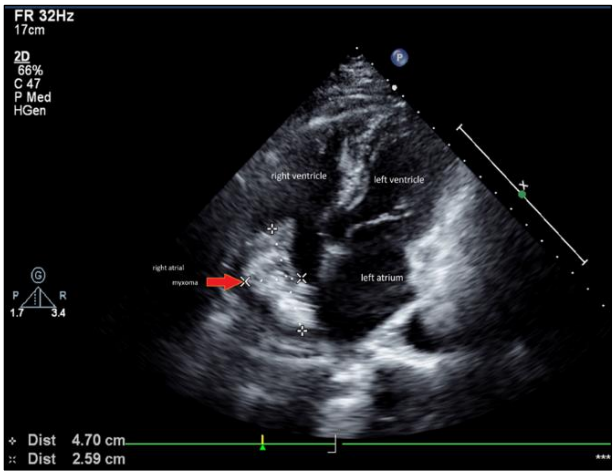

Figure 1: Echo with apical four chamber view right atrial myxoma.

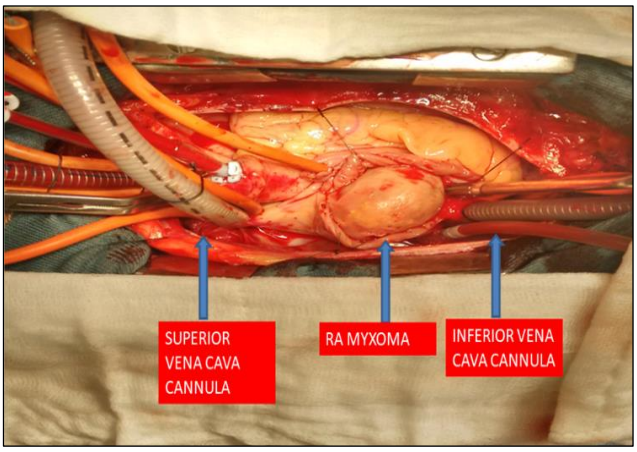

Figure 2: Myxoma mass inside the right atrium with cannula in situ.

Figure 3: Myxoma mass round surface.

Coronary angiogram showed normal epicardial coronaries. He was planned for a myxoma excision after obtaining consent from haematologist under injection meropenem and injection teicoplanin coverage. After a median sternotomy and aortic bicaval cannulation (Figure 2) right atrium was opened and the mass was removed (Figure 3) and was sent for histology and the defect in the inter atrial septum was closed with a pericardial patch.

The aortic cross clamp time was 21 minutes and cardiopulmonary bypass time was 85 minutes. There was no drop in the platelet count or any major bleeding needing significant blood or blood products transfusion. His post-operative course was uneventful and was discharged in a stable condition. The platelet count was 1.5 lakh/cu mm on discharge. The histological features were consistent of myxoma (Figure 4). On last follow up which is two years post-surgery, the patient is doing well with echo cardiography showing no recurrence of myxoma.

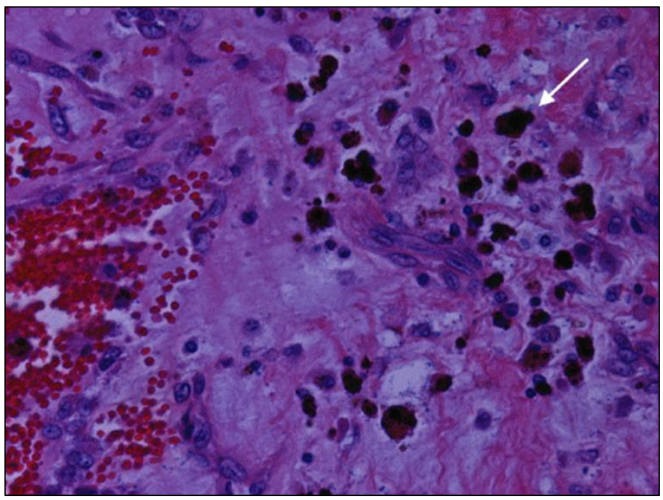

Figure 4: Hematoxylin eosin stain with 40X magnification spindle shaped myxoma cells in white arrow.

DISCUSSION

Myxomas are commonest primary benign intra cavitatory tumours with the incidence of 0.5 per million populations.[1] They account for 0.3% of all cardiac surgeries performed and commonly arise from the left atrium and only 25% of it arises from the right atrium or ventricle.[2] Clinically, they are characterized by triad of embolization, blood flow obstruction, and systemic symptoms (Goodwin's triad).[3] Most of the myxoma patients present as embolic event, with cerebral embolization occurs in upto 45%, and this commonly occurs in the middle cerebral artery territory[3]. Obstruction to blood flow can result in cardiac failure or syncope in 41-79% of cases. Left ventricular outflow tract obstruction because of the mass can mimic mitral stenosis and can cause pulmonary hypertension and even congestive heart failure.[3] Right sided myxoma can also be associated with obstruction to the right heart and can present as cardiovascular collapse during induction.

Thrombocytopenia has been reported to be associated with cardiac tumours Large cardiac tumours often result in intracardiac blood flow obstruction, eventually leading to right heart failure.[4] Cardiac tumour-associated thrombocytopenia is often associated with other hematologic disorders such as anaemia or erythrocytosis as well. The mechanism by which intracardiac tumour leads to thrombocytopenia remains unclear, although it has been postulated that abnormal mechanical shear stress, caused by tumour-induced flow obstruction, may be responsible for the platelet break down.[4] Fever, malaise, weight loss, fatigue, anaemia, and raised erythrocyte sedimentation rate are common constitutional symptoms which occur in around 90% patients with myxomas. These features resolve immediately after surgery and are due to release of inflammatory mediators from tumour cells which can include cytokines. [4]

Echocardiography is the investigation of choice as it is non-invasive and allows preoperative diagnosis with fair degree of accuracy. Transthoracic echocardiography can determine the location, size, shape, attachment and mobility of a tumour. The transoesophageal approach can give a clear idea about the site of attachment and morphological features of left atrial and ventricular myxomas. Surgical management is the treatment of choice for myxomas. The recurrence of myxoma has been reported to be less than 2% on most series.[4] The debate still continues as to the choice of the most appropriate surgical approach to achieve complete excision of intracardiac myxoma with techniques including Left, right or bi atrial approach.[4]

Structurally, myxomas are of two types, one with round, non-mobile surface, as in our case, (explaining the absence of embolic symptoms) and another polypoid type with irregular shape, mobile surface and this latter type has the higher incidence of embolism and this is the commonest type to prolapse into the ventricles.[5] Cardiac myxomas form a very small percentage of the cardiac tumours. Instant surgical treatment is essential in all patients. A close follow-up of all the patients is mandatory because the reappearance of myxomas has been documented at various intervals.

This case leads to further impression that though thrombocytopenic effects of the cardiac mass are to be kept in mind especially when treating the tumour, the two entities(myxoma and thrombocytopenia) can co-exist separately and should be taken into account which can help in the accurate treatment of both the conditions. Careful and prudent peri operative management in terms of platelet transfusion as well as haematological disturbances are to be expected which should be treated accordingly.

CONCLUSION

One must understand that myxoma and ITP can co-exist separately and treatment should be tailor made in such situations addressing both the conditions. Not necessary that thrombocytopenia is always due to the hemolytic mechanical effects of myxoma and both these conditions can co-exist independently too. The treatment varies and all the options should be kept in mind before deciding on the exact treatment.

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