Mistaken Identity:

A Case Report of Simultaneous Right Atrial Myxoma and Bilateral Pulmonary Artery Inflammatory Myofibroblastic Tumor

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Abstract

INTRODUCTION: Inflammatory Myofibroblastic Tumor (IMT), previously known as pseudotumor, is currently classified by WHO as an intermediate spindle cell type, with recent reports of presence of local recurrences, and regional metastases, challenging its previous concept of being a benign reactive lesion. The pathogenesis is unclear and true incidence and anatomical distribution are difficult to establish because of the many vague terms used to previously described IMT such as pseudotumo. The lungs, liver and gastrointestinal tract are the most common sites, but has never been reported in the pulmonary artery. We present the first case of IMT of the pulmonary artery occurring simultaneously with a right atrial Myxoma.

CASE: A 46-year old female presented with constitutional symptoms (fever and weight loss) with intermittent dyspnea, was diagnosed with a right atrial (RA) mass (3.4 x 2.4 x 5.9 cm) with a stalk attached to the interatrial septum, and irregularly shaped echogenic mass attached to the pulmonary artery (PA) wall measuring 1.5 x 3.1 cm by echocardiography. Contrast-enhanced CT scan of the chest showed filling defects in the main pulmonary artery causing occlusion of left and right pulmonary arteries. Due to the simultaneous presence of the RA mass consistent with a myxoma, differential diagnoses for the the PA mass were either tumor embolization, a multi-centric myxoma, or thromboembolism, all of which were reported in literature. Endarterectomy with complete excision was done and patient's symptoms resolved. Histopathology and immunohistochemistry revealed myxoma of the RA, and IMT of the PA.

CONCLUSION: Recent opinions suggested that immunohistochemistry does not play a role in diagnosis but may play role in predicting aggressive behaviours of IMT. Due to the age of this patient, ALK negativity and moderate Ki67 proliferation marker (10-30%), and the rarity of the location of this IMT, malignant potential and possible recurrence for this tumor is possible, as suggested by some studies. Nevertheless, this case report is the first intravascular IMT (of the pulmonary artery) reported and highlights the importance of including IMT as a differential diagnosis in patients with PA mass. A close follow-up is important for surveillance of possible recurrence for both myxoma and IMT in this case.

KEY WORDS: inflammatory myofibroblastic tumor, pseudotumor, myxoma

Image

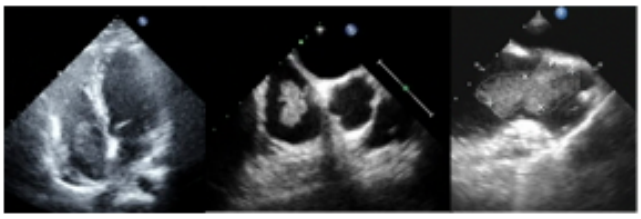


Figure 1: from left to right: (leftmost) Apical 4-chamber view of the transthoracic echocardiogram showing the right atrial mass; (middle picture) short-axis view of the great vessels at the mid-esophageal level on transesophageal echocardiogram showing the stalk attached to the interatrial septum; (rightmost) Bicaval view at the mid-esophageal level on TEE showing the attachment to the SVC rim of the interatrial septum (IAS).

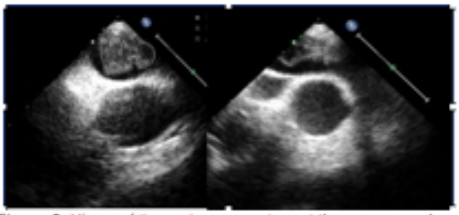


Figure 2: Views of the pulmonary artery at the upper esophageal level on TEE showing the irregularly-shaped echogenic mass attached to the main pulmonary artery wall and extending to the left and right PA branches.

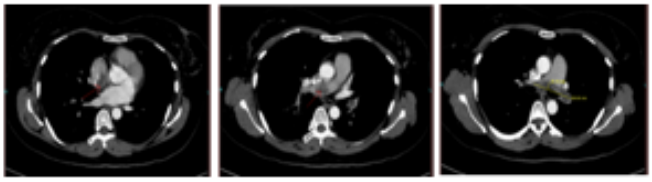


Figure 3. Contrast-enhanced CT scan of the chest showing large filling defect straddled within the distal main pulmonary artery (MPA)(arrow) extending to the right and left and pulmonary arteries measuring 7.3 cm in length with widest diameter of 1.3 cm in the right and 1.5 cm in the left pulmonary artery.

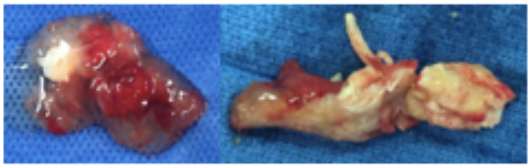


Figure 4. Gross appearance of the right atrial mass (left picture), and mass from pulmonary artery (right picture).

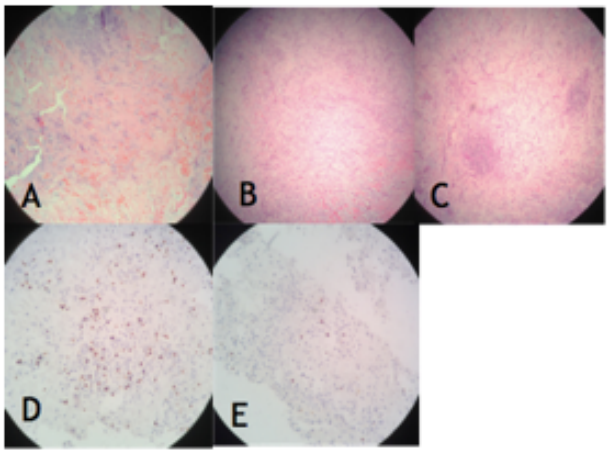


Figure 5: Histopathology of ***A***: right atrial mass showing plump to spindle neoplastic cells in cords and vague nests in myxoid stroma consistent with myxoma; ***B and C***: variable areas showing spindled to stellate cells haphazardly arranged and set in loose myxoid matrix, densely collagenized storm and occasional compact cellular areas with prominent inflammatory cells (composed of lymphoplasmacytes and neutrophils) permeating the lesion, ***D and E***: Ki-67 proliferation of 10-30% in right and 5% left pulmonary mass, respectively. (Similarly the Ki-67 of the right atrial mass is <5%; p53 is <1% for both atrial masses and negative for ALK (anaplastic lymphoma kinase 1 protein) - not shown.

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Biography

Jhoanna Marcelo is currently in her third year of fellowship training in Adult Cardiology at the Philippine Heart Center, and the Chief-Fellow of the department. She plans to pursue further training in Interventional Cardiology after she graduates. Her ceaseless enthusiasm for research is equally matched by her assiduousness in treating her patients. A promising clinician, she is keen on identifying great clinical cases for learning, and is looking for opportunities to get involved in researches that would have impact in her country and its healthcare policy.

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