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Mistaken identity: A case report of simultaneous right atrial myxoma and bilateral pulmonary artery inflammatory myofibroblastic tumor

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Introduction: Inflammatory myofibroblastic tumor (IMT), previously known as pseudo-tumor 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 pseudo-tumor. 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 Report: A 46-year old female presented with constitutional symptoms (fever and weight loss) with intermittent dyspnea, was diagnosed with a right atrial (RA) mass (3.4×2.4×5.9 cm) with a stalk attached to the inter-atrial septum and irregularly shaped echogenic mass attached to the pulmonary artery (PA) wall measuring 1.5×3.1 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 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 behaviors 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.

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