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Zarish Umar et al., Int J Cardiovas Res 2017, 6:5
DOI: 10.4172/2324-8602-C1-003

Unusual presentation of syndrome of left atrial myxoma, anterior communicating artery aneurysm, severe dextroscoliosis and macular hypopigmented skin lesions

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Introduction: Atrial myxomas tend to present clinically in myriad forms, associations and syndromes requiring diligence in diagnosing the condition.

Case Report: A 59 year female with history of hypertension, headaches and coil embolization of right anterior communicating artery aneurysm presented with a month of intermittent non-exertional substernal left chest fullness associated with nausea and vomiting. Physical examination was remarkable only for bilateral arm macular hypopigmented patches. No JVD was observed with chest clear to auscultation. Cardiovascular examination revealed normal S1 and S2 with no murmurs, rubs or gallops. Labs: unremarkable. ECG: sinus rhythm and left axis deviation. Telemetry: no arrhythmias. Chest X-ray: severe dextroscoliosis. TTE: LVEF 0.60, grade 1 diastolic filling of LV and a large mobile mass 5 x 2 cm in the left atrium, attached to the interatrial septum with protrusion through the mitral valve in diastole. Coronary angiogram showed left dominant circulation, normal coronary arteries and contrast blush of the left atrial mass. CT head: coils in the left anterior communicating artery aneurysm. An open heart surgery was conducted with excision of gelatinous left atrial mass. Histopathology of the mass confirmed the myxoma.

Discussion: The symptoms in this patient with left atrial myxoma, angiographically normal coronary arteries were unusual in that there was nonexertional intermittent substernal left chest fullness with nausea and an episode of sweating with vomiting. A plausible cause of the aforementioned symptoms could be the repeated protrusion of the left atrial myxoma into the mitral valve in every diastole causing transient falls in left ventricular filling, low cardiac output, coronary hypoperfusion and myocardial ischemia. Cerebral arterial aneurysms are known to be associated with myxomas either through genetics or overproduction of interleukin-6 by myxomas. Association of myxoma and scoliosis is anecdotal. There is only anecdotal evidence of atrial myxomas associated macular hypopigmented skin lesions as an atypical presentation in a patient with Carney complex which essentially comprises cardiac, cutaneous or mucosal myxomas, hyperpigmented skin lesions, neural and endocrine tumors due to PRKAR1A gene mutation.

Conclusion: Atrial myxoma is a great mimicker with unusual symptoms requiring a high index of suspicion. It's myriad associations with other organ systems have both genetic as well as cause and effect relationships.

Biography

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