medications and if not alcohol septal reduction or even surgical myectomy in severe conditions.

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Ebstein malformation associated with left atrium myxoma

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A 62 year old lady, diagnosed with Ebstein malformation 12 years ago, presented to clinic with severe fatigue, breathlessness, chest pain, palpitations, and loss of weight. She had clear evidence for irregular rhythm, rales at the basal lung fields and clinical signs of severe right sided failure. A 12 lead ECG showed atrial fibrillation with wide QRS complex (130 ms) and right bundle-branch block. Echocardiography showed septal tricuspid valve leaflet displacement of 1.6 cm into the right ventricle (Figure 1&2). The right atrium was enlarged, 70 x 110 cm in diameter, with moderate tricuspid regurgitation and estimated pulmonary hypertension of 90 mmHg. In addition, there was an echo dense mass (26 x 30 mm) attached to the left atrial side of the interatrial septum, with morphologic appearance consistent with atrial myxoma. According to our knowledge, this case of such combination seems the first reported in the literature.

The Ebstein’s anomaly, first prescribed by Wilhelm Ebstein in 1866, is a rare cardiac anomaly that accounts for less than 1% of all congenital heart disease and involves both genders equally. Ebstein anomaly consists of abnormal attachment of tricuspid valve leaflets to the annulus, with a downward displacement of the tricuspid valve into the right ventricle, dysplastic valve tissue associated with tricuspid regurgitation. Most patients with this anomaly remain asymptomatic for years. The predominant symptoms are fatigue, breathlessness on exertion and cyanosis. Palpitation in the form of paroxysmal atrial arrhythmia and premature ventricular beats are common, whereas ascites and peripheral edema are present in some of these patients. Ebstein anomaly may be associated with other congenital heart malformations: ventricular septal defect, interatrial septal defect or patent foramen ovale, aortic coarctation, pulmonary atresia with an intact ventricular septum, partial atrioventricular canal, hypoplastic pulmonary arteries, patent ductus arteriosus, pulmonary stenosis, tetralogy of Fallot, cleft anterior leaflet of the mitral valve or mitral valve prolapse. Surgical treatment of Ebstein anomaly includes valve replacement or the native valve repair.

Our patient remained silent for 50 years until presented with
paroxysmal atrial fibrillation when the diagnosis of Ebstein anomaly was made. At that stage tricuspid regurgitation was just mild-moderate in severity. Now, the clinical picture has significantly changed with worsening regurgitation and development of pulmonary hypertension. While the former complication is related to the syndrome the latter might not necessarily. It seems the development of left atrial myxoma was an additional cause for this patient’s pulmonary edema and pulmonary hypertension. Such combination has not been reported before, and it urges a critical echocardiographic examination in Ebstein patients who present with unexplained pulmonary hypertension.

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Myocardial Crypts: An incidental finding in a symptomatic patient with raised troponin

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A 61 year old female of Asian origin presented with a two day history of intermittent sharp central chest pain radiating to the left shoulder, with each episode lasting a few seconds. Associated symptoms included exertional dyspnoea and limited exercise capacity to one flight of stairs. She denied any palpitations, pre-syncope or syncope. She had no orthopnoea, paroxysmal nocturnal dyspnoea or ankle swelling. She had stable pulmonary sarcoidosis, dual-chamber pacemaker insertion for symptomatic bradycardia 7 years ago, non-insulin dependent diabetes mellitus, hypertension, benign paroxysmal positional vertigo from the right ear, and hearing difficulties. Drug history included Metformin 500 mg, Ramipril 10 mg, Bendroflumethiazide 2.5 mg, Aspirin 75 mg and Simvastatin 20 mg. She had no known drug allergies.

Clinical examination revealed mild crepitations at the right lung base. She was haemodynamically stable and clinically euvoaemic. Twelve lead ECG showed paced rhythm. Chest x-ray showed satisfactory pacemaker wires position but was otherwise unremarkable. Blood tests were completely normal apart from a rise in Troponin I level to 0.10ug/L. A repeat Troponin level was 0.07ug/L (in house value <0.03 is considered normal).

In view of the patient’s risk factors for atherosclerosis and the rise in Troponin level, a decision was made to perform a coronary angiogram. This showed unobstructed arteries. Left ventriculogram showed a localised pouch at the inferior LV wall (Figure 1), which was confirmed on a transthoracic echocardiogram (Figure 2), and again with a cavity contrast echo for LV opacification (LVO) (Figure 3).

Figure 1: Left Ventriculogram showing a recess or pouch (arrow) in the inferior wall of the Left Ventricle.