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HYPERTROPHIC CARDIOMYOPATHY ASSOCIATED WITH LEFT ATRIAL MYXOMA

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Abstract :
In this paper we describe hypertrophic cardiomyopathy associated with left atrial myxoma. A 63 yr old patient presented with chest pain and dyspnea. There was no history of hypertension or syncope. Examination revealed a mid diastolic murmur at the apex which varied with patient position. His blood pressure was within normal limit. ECG revealed sinus rhytm and heart rate of 62 bpm. There was left ventricular hypertrophy by voltage criteria with negative T waves in the left precordial leads. Echocardiogram showed a mass of size 3.27 by 1.95 cm attached to the atrial septum by a narrow stalk which protruded through the mitral valve during diastole. Asymmetrical septal hypertrophy was noted. Interventricular septal thickness was 2.06 cm. LV posterior wall thickness was 1.23 cm. Systolic anterior motion of mitral valve was present. No significant LVOT gradient was noted. CT coronary angiogram showed normal epicardial coronary arteries. CT showed Left atrial myxoma as a filling defect in Left atrium. Banana shaped Left ventricular cavity was also made out suggesting HCM. The patient underwent surgery to remove left atrial myxoma. Histology of the biopsy specimen confirmed myxoma. Biopsy specimen of the LV wall showed hypertrophy and disarray of myocardial muscle cells. HCM has been associated with lentiginosis. Association of lentiginosis and left atrial myxoma has been described. We could find only one literature describing an association of hypertrophic cardiomyopathy with left atrial myxoma. This is the second case description of a patient with hypertrophic cardiomyopathy associated with a left atrial myxoma. The coexistence may be more than a chance occurrence so that the association of left atrial myxoma should be sought for when one sees a patient with a hypertrophic cardiomyopathy.

Keyword :
Hypertrophic cardiomyopathy, Left atrial myxoma

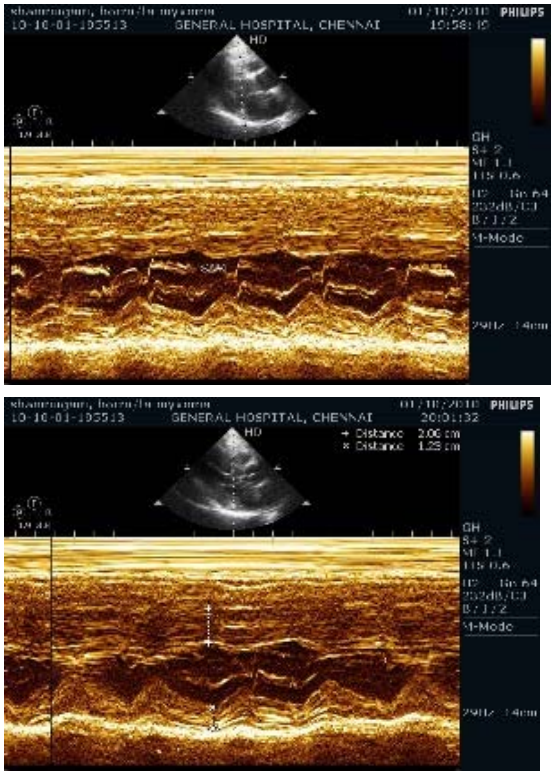
CASE REPORT: HYPERTROPHIC CARDIOMYOPATHY ASSOCIATED WITH LEFT ATRIAL MYXOMA

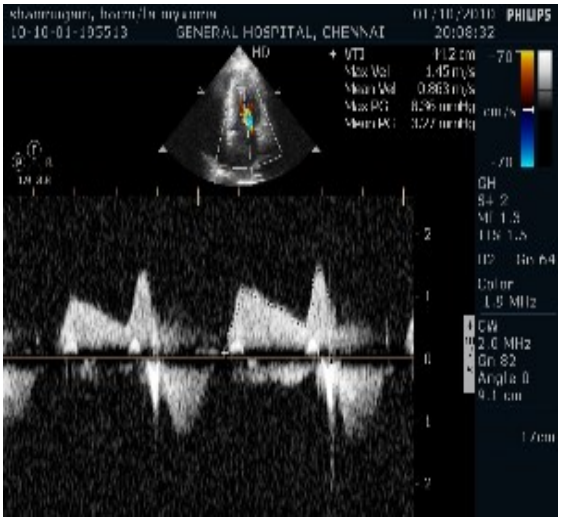
Tumours of the heart are uncommon. With the advent of echocardiography, intracardiac tumors are being found more frequently. Myxomas are the most frequent benign tumors. To our knowledge, only one medical literature has described an association of hypertrophic cardiomyopathy with left atrial myxoma.

A 63 year old gentlman was referred to outpatient department of this hospital for further evaluation of retrosternal chest pain of 4 months duration.Chest pain was brought on by exertion and relieved by rest (CCS class 2). Chest pain was not associated with increased sweating. He had shortness of breath on exertion for past 2 months (NYHA class 2). There was no history of paroxysmal nocturnal dyspnea. There was no history of palpitation or syncope. He was diagnosed to have type 2 diabetes mellitus 6 years ago and was on oral hypoglycemic agent. There was no history of hypertension. No family history of sudden death, syncope, cutaneous pigmentation or endocrine disorder. He was not a smoker.Clinical examination revealed only a soft, short, rumbling middiastolic murmur at the apex in the left lateral decubitus position. The loudness of the murmur varied with position of the patient and sometimes it was not audible.The blood pressure was 132/74 mm Hg. The heart rate was 62 beats per minute and regular.Results of the remainder of the clinical examination were normal. Blood investigations were unremarkable.The electrocardiogram revealed sinus rhythm and left ventricular hypertrophy by voltage criteria with negative T Waves in the left precordial leads.

Two-dimensional echocardiography showed a 3.27 by 1.95 cm mass in the left atrium attached to the atrial septum by a narrow stalk. The tumour protruded through the mitral orifice during diastole. Mean gradient across mitral valve was 3.27 mm Hg. Mitral valve area calculated by pressure half time method was 1.44 cm^2. Asymmetrical septal hypertrophy was noted. Interventricular septum thickness was 2.06 cm. Left ventricular posterior wall thickness was 1.23 cm. Systolic anterior movement of the anterior mitral leaflet was noted. LVOT maximum gradient was 9.4 mm Hg and mean gradient was 4.4 mm Hg. The left ventricular diastolic dimension was 3.29 cm and systolic dimension was 1.94 cm. Ejection fraction was 73%. The walls were

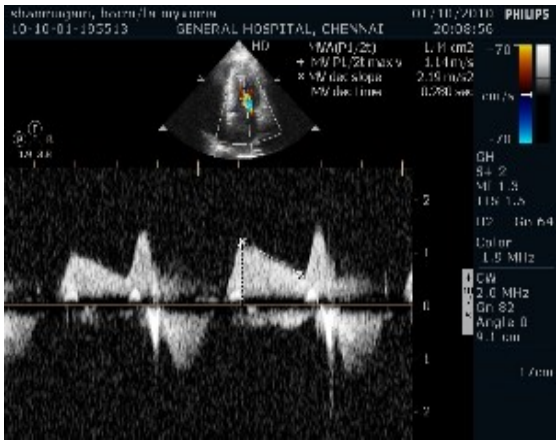
SAM



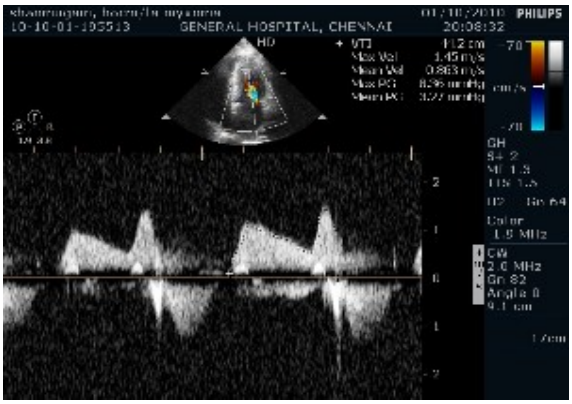


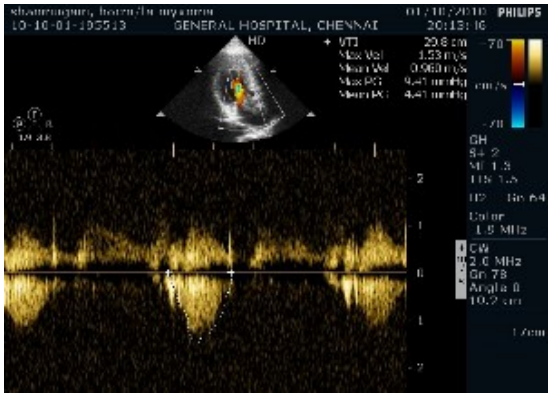
Mitral valve area LA Myxoma A4C-

Colour Doppler

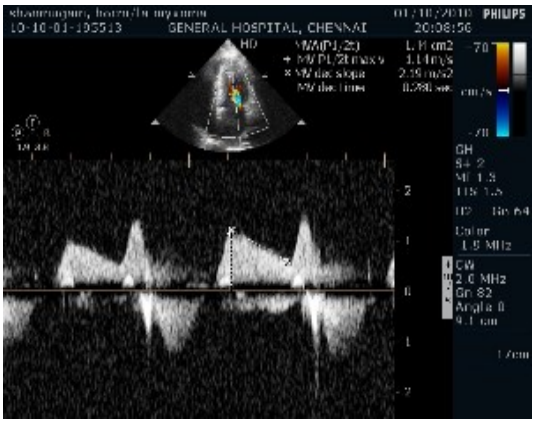


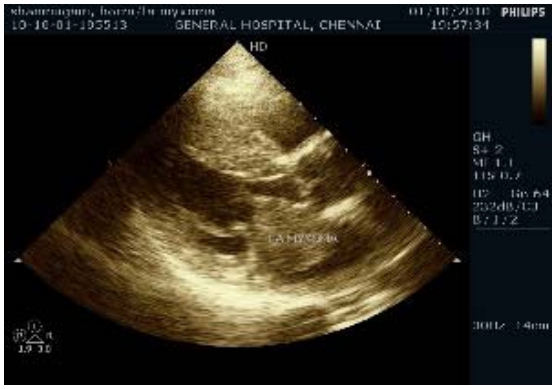
LV Dimensions LA myxoma dimensions

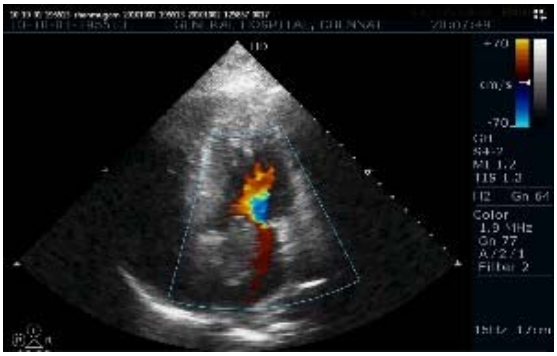


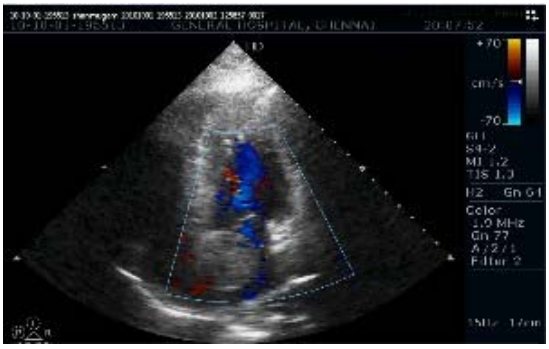


Mitral valve gradient

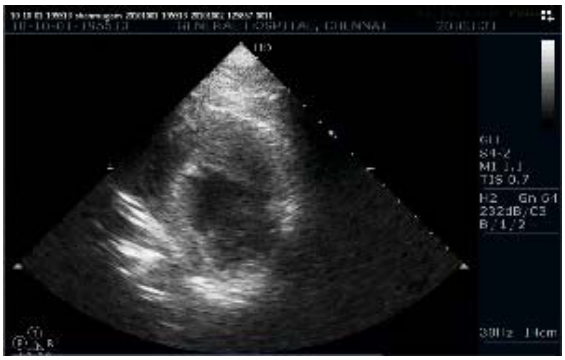


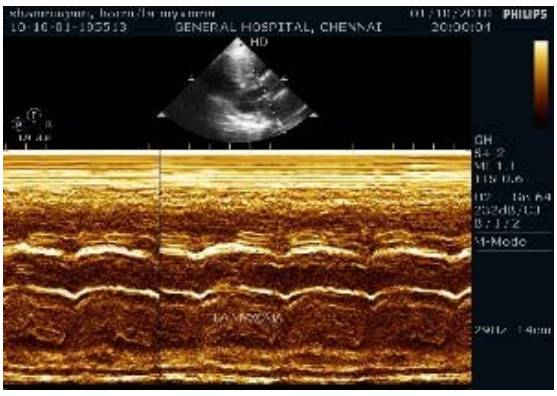




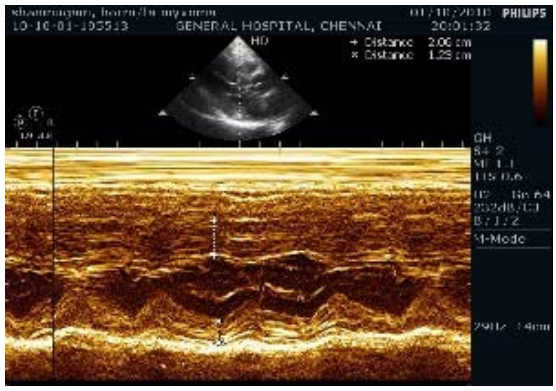


A4C-colour doppler

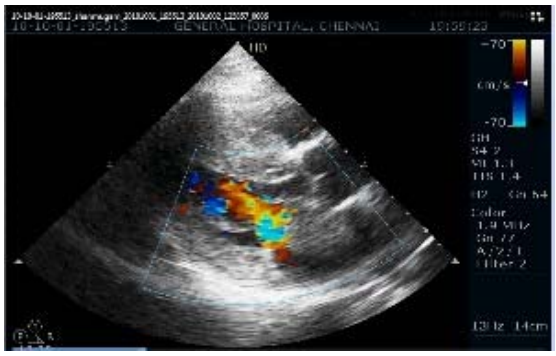




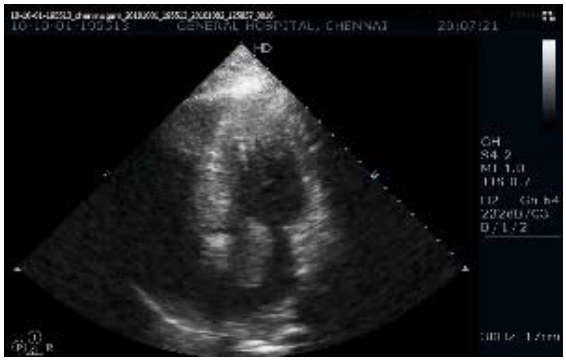
PSAX-ASH



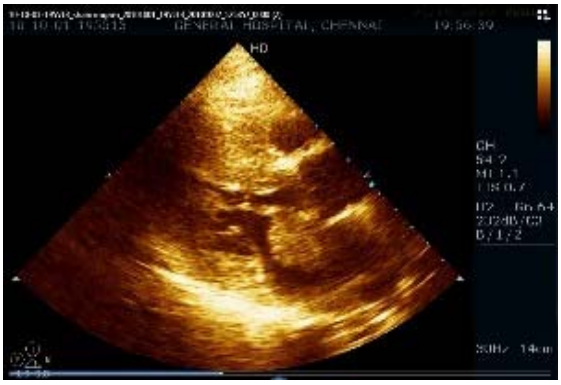
M mode ASH



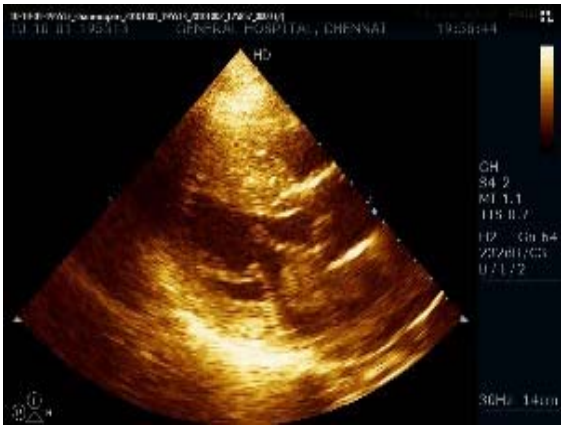
PLAX-colour doppler



LA MYXOMA PROLAPSING INTO MV



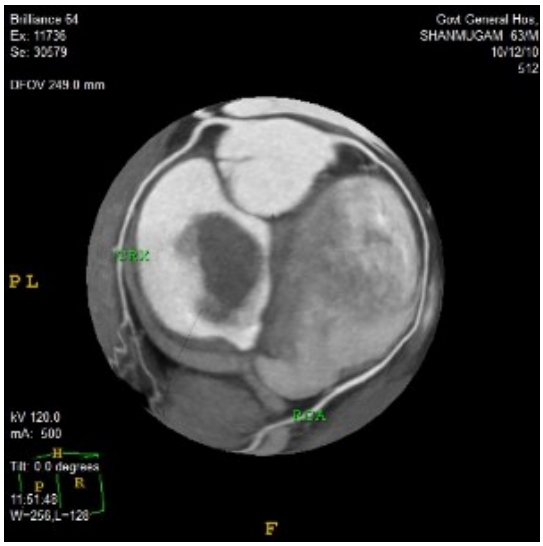
Lt atrial myxoma

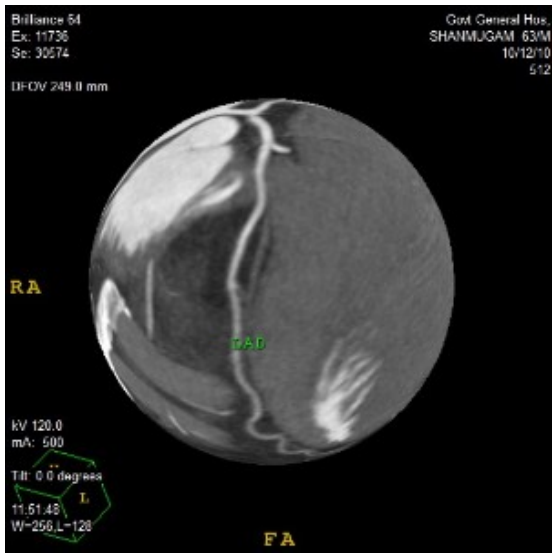


PLAX-Lt atrial myxoma

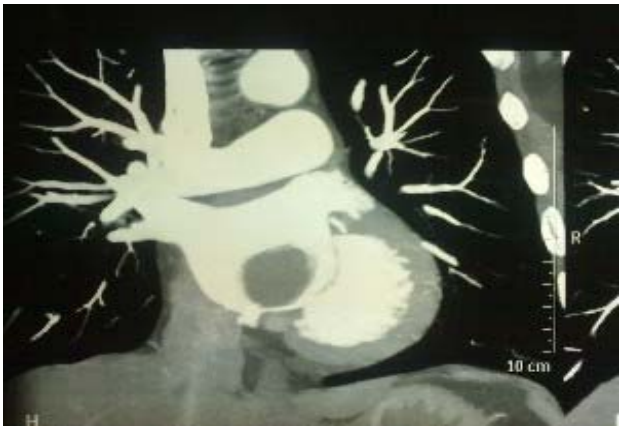
CT coronary angiogram showed normal epicardial coronary arteries. CT showed Left atrial myxoma as a filling defect in Left atrium. Banana shaped Left ventricular cavity was also made out suggesting HCM.

RCA and LCX





LAD



The patient underwent surgery to remove the left atrial tumor and a 3.3 ×2.01 cm pedunculated tumor of gelatinous consistency was found. Histologic examination confirmed the diagnosis of left atrial myxoma. The patient had an uneventful recovery. A biopsy specimen of the left ventricular wall showed hypertrophied and bizarre arrangement (disarray) of myocardial muscle cells.

DISCUSSION:

Hypertrophic cardiomyopathy (HCM) or hypertrophic obstructive cardiomyopathy (HOCM) has been described in association with various congenital cardiac anomalies such as coronary arteriovenous fistula. Familial lentiginosis has been reported to be associated with HOCM and the term “progressive cardiomyopathic lentiginosis” has been proposed for this syndrome.One case of lentiginosis associated with left atrial myxoma has been described before but that case was not associated with hypertrophic cardiomyopathy. The only link between hypertrophic cardiomyopathy and left atrial myxoma could be through lentiginosis. We could find only one literature describing an association of hypertrophic cardiomyopathy with left atrial myxoma.In this case, none of his family members or primary relatives were known to have either a myxoma, the appearance of spotty pigmentation, or any endocrine disease.

This is the second case description of a patient with hypertrophic cardiomyopathy associated with a left atrial myxoma. The coexistence may be more than a chance occurrence so that the association of left atrial myxoma should be sought for when one sees a patient with a hypertrophic cardiomyopathy

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