Dynamic left ventricular outflow tract obstruction in senile cardiac amyloidosis

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Abstract Dynamic left ventricular outflow tract (LVOT) obstruction is classically seen in hypertrophic obstructive cardiomyopathy (HOCM). This can also be seen in cardiac amyloidosis. We describe a rare case of senile systemic amyloidosis with dynamic LVOT obstruction and concomitant three vessel coronary artery disease presenting with clinical and echocardiographic findings similar to those seen in HOCM. We also highlight the importance of distinguishing the sub-types of amyloidosis so that the appropriate therapy can be offered to patients with cardiac involvement, including coronary artery bypass grafting and septal myotomy/myectomy to relieve LVOT obstruction in the more benign forms of cardiac amyloidosis.

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Case report

A 73-year-old Caucasian gentleman presents with a three-year history of presyncope and chest discomfort. He was considered New York Heart Association (NYHA) Class III with an effort tolerance of 50 yards.

Six months prior, an extensive work-up including chest X-ray (CXR), electrocardiogram (ECG), laboratory data, 24-h ambulatory Holter recording, and physical examination was completed. Subsequent ambulatory blood pressure monitoring, exercise stress test with nuclear tomographic imaging, and CT scan of the head were all normal. Coronary angiography revealed only moderate coronary atherosclerotic disease at multiple sites. None of the stenoses were considered hemodynamically significant and ventriculography revealed a normal ejection fraction with no regional wall motion abnormalities.

He subsequently underwent electrophysiologic assessment with poorly tolerated polymorphic ventricular tachycardia resulting in hemodynamic collapse and subsequent restoration of hemodynamic stability with urgent cardioversion. This finding was considered non-specific by the electrophysiologist.

On physical examination, he appeared younger than his stated age. Heart rate was 70 beats per minute and blood pressure was 160/80 mm Hg, neck veins were not distended; the precordial examination was unremarkable except for a forceful well localised apical impulse. A soft ejection systolic murmur at the upper left sternal border, grade 1/6 was appreciated; carotid upstroke was considered normal; no added cardiac sounds were noted; remainder of the physical examination was normal with no postural blood pressure drop.

Basic laboratory data were within normal limits. Twelve-lead ECG showed normal sinus rhythm with
occasional premature supraventricular beats, left axis deviation, and minor non-specific ST–T changes with normal voltages. The CXR showed mild calcification of the aortic knuckle with normal heart size and clear lung fields. Adenosine stress test revealed reversible defects in the anterior and posterior circulation consistent with multivessel coronary artery disease. Two-dimensional echocardiogram revealed thickened left and right ventricular walls (the interventricular septum measured 18 mm) with evidence of dynamic outflow tract obstruction considered mild in the right ventricular outflow tract (RVOT) (20 mm Hg gradient) and a gradient across the left ventricular outflow tract (LVOT) of 46 mm Hg at rest and 81 mm Hg post-Valsalva. Systolic anterior motion of the mitral leaflet was also present (Figs. 1–5). LA was moderately enlarged (55 mm). Degenerative changes of the aortic valve were noted. The diastolic parameters were consistent with a marked relaxation abnormality. There was no evidence of pericardial effusion.

Coronary angiography confirmed severe three vessel coronary artery disease and septal biopsy confirmed cardiac amyloidosis with focal nodular and interstitial amyloid on sulfated LC and ALCIA in blue stain. Immunohistochemical stains were positive for serum amyloid protein and prealbumin. Stains for kappa and lambda light chain, serum amyloid associated protein, beta II microglobulin and albumin were negative. Findings were consistent with senile familial amyloidosis.

The patient was referred for coronary artery bypass grafting (CABG) which was completed along with an extended left ventricular septal myectomy. The resting gradient in the LVOT was 80 mm Hg confirmed on intraoperative transesophageal echocardiography as well as by direct intracardiac manometry. Pressure in the aorta and LV were measured as 80/40 mm Hg and 167/20 mm Hg, respectively. An extended left ventricular septal myectomy was completed with no residual LVOT gradient post-procedure.

The patient had an uncomplicated postoperative course, and at 5 years postoperative was doing well with complete resolution of his symptoms and was NYHA Class I. On a 6 year postoperative examination the patient was NYHA Class II with new onset atrial fibrillation, moderate evolving mitral regurgitation and moderate diastolic dysfunction.

Discussion

Hypertrophic cardiomyopathy with dynamic LVOT obstruction has classic echocardiographic and clinical features. Mimickers of LVOT obstruction have
recently been described in patients with acute myocardial infarction. LVOT obstruction has been described in secondary amyloidosis with multiple myeloma and type I Gaucher’s disease. In a study of 204 surgical pathology specimens of subaortic septal myectomy associated with hypertrophic obstructive cardiomyopathy, amyloidosis was detected in three cases and was considered to be senile based on the patients’ ages. In all the three of these cases, the finding of amyloidosis was unexpected, as opposed to the case we report in which the suspicion was raised preoperatively. To our knowledge, neither a case report of senile cardiac amyloidosis causing dynamic LVOT obstruction has been described in the literature nor has there been a report of such a patient undergoing CABG and septal myotomy/myectomy successfully with excellent results at 5 year follow-up. It is known that transmitral flow indices consistent with restrictive physiology are strong predictors of cardiac death; therefore we attribute the good follow-up result to low grade diastolic dysfunction at presentation.

It may be challenging to differentiate cardiac amyloid from hypertrophic cardiomyopathy on the basis of two-dimensional echocardiography features alone. Asymmetric increases in septal wall thickness, systolic anterior motion of the mitral valve, and dynamic LVOT obstruction have been previously described in patients with cardiac amyloidosis.

It has been shown that patients with systemic senile amyloidosis (SSA) have a survival of 60 months compared to patients with primary amyloidosis (AL) of only 5.4 months. Senile cardiac amyloidosis is a heterogenous group of disorders that is not readily distinguished by routine microscopy or transmission electron microscopy. It is recommended that patients with suspected cardiac amyloid be considered for endomyocardial biopsy in the absence of evidence of amyloid deposition in other organs. In patients with negative urine and serum monoclonal electrophoretic bands, specific immunohistochemical staining for prealbumin (transthyretin) and immunoglobulin light chain should be undertaken to distinguish between SSA and immunoglobulin derived amyloidosis because of the vast differences in prognosis and hence therapies.

References

Infective endocarditis complicating hypertrophic obstructive cardiomyopathy

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Abstract Infective endocarditis is a rare complication of hypertrophic cardiomyopathy. It's estimated incidence is 1.4 per 1000 person/year in all patients and it increases to 3.8 per 1000 person/year in patients with left ventricular outflow obstruction. The most common site of vegetation is the ventricular aspect of anterior mitral valve leaflet. We report a case of a 43-year-old man who was admitted for mitral infective endocarditis resulting in severe mitral regurgitation complicating a hypertrophic obstructive cardiomyopathy. The patient underwent mitral valve replacement. Post-operative outcome was good with relieve of symptom and resolution of left ventricular outflow obstruction. Literature data are reviewed.

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Case report

A 43-year-old man was admitted in our department with dyspnoea, fever and weakness lasting for two months. He had no history of hypertension or known heart disease, no family history of sudden death was reported and he had no symptoms before.

At presentation, the patient was pale with a temperature of 38.5 °C, a tachycardia of 100 beats/min. His blood pressure was 120/70 mm Hg. The auscultation revealed a systolic murmur suggestive of mitral regurgitation. There was no sign of heart failure and all peripheral pulses were present. The dental state was bad with multiple teeth decay. A 12 lead electrocardiogram showed a sinusal tachycardia of 100 /min, a left ventricular hypertrophy and abnormal lateral repolarisation. The chest X-ray showed a cardiothoracic ratio of 0.45. Laboratory investigation revealed a hypochromic microcytic anaemia (haemoglobin, 10 g/dl), leukocytosis of $13.7\times10^9$ /l, raised inflammatory markers and normal serum creatinine. A set of three blood cultures grew viridans streptococci sensitive to ampicillin. Transthoracic echocardiography disclosed an asymmetric septal hypertrophy (18 mm), posterior wall thickness of 11 mm, hyperkinetic left ventricle, dilated left atrium (two-dimensional area of 30 cm²), systolic anterior motion of the mitral valve with obstruction.

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