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Case report

# Combined orthotopic heart transplantation followed by autologous stem cell transplantation in a patient with light chain amyloidosis and isolated cardiac involvement

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ABSTRACT

We present a case of amyloidosis AL with isolated myocardial involvement. Because of a refractory heart failure picture, patient underwent orthotopic heart transplant (OHT). The replaced heart showed an important midwall infiltration. Ten months after he underwent autologous stem-cell transplantation (ASCT) with a favorable outcome. The case demonstrates that OHT followed by ASCT in highly selected patients with light chain amyloidosis is a life-saving procedure.



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## The case

A 44 years-old male was admitted to emergency department because of sudden onset of exertional dyspnea, hand numbness and skin purple.

The EKG demonstrates sinus rhythm and low voltages on the precordial leads (Fig. 1).

He underwent transthoracic echocardiography demonstrating high left ventricle concentric hypertrophy (maximum thickness 28 mm) and ground glass texture of the left ventricle walls with preserved ejection fraction (EF) (left ventricle-EF: 65%) (Fig. 2).

The diagnosis of cardiac amyloidosis was performed by endo-myocardial biopsy specimens that showed: myocellular texture with loss of myofibrillar material, nuclear hypertrophy and apoptotic nuclei, important thickening of intramural arterioles and Congo-Red staining positive; with polarized light the amyloid has the typical ''apple-green'' birefringence (Fig. 4 – panels A and B). Proteinuria with lambda chain confirmed the diagnosis: *amyloidosis AL with isolated myocardial involvement* given that all remaining soft tissues were infiltration-free.

Bone marrow aspirate showed 17% plasma cell infiltration.

Thalidomide and dexamethasone were soon started but failed even after bortezomib addition.

A new re-admission, due to NYHA class impairment, occurred. EKG showed atrial fibrillation treated with electrical cardioversion.

Because of a severe (persistent NYHA class IV) and refractory heart failure picture, he underwent orthotopic heart transplant (OHT). The replaced heart showed an important midwall infiltration (Fig. 3).

After high dose melphalan therapy (melphalan 0.22 mg/kg once daily for 4 days per week with prednisone 40 mg daily for 4 days per week), ten months after he underwent autologous stem-cell transplantation (ASCT) (Fig. 4).

Nowadays, seven years after the double transplantation, monoclonal components are not present neither in serum nor in urine. There are no transplant rejection signs.

He is hemodynamically compensated with normal troponin and NT-proBNP levels [1] and there is a complete hematological remission. He is back to a normal life with a persistent NYHA class I.

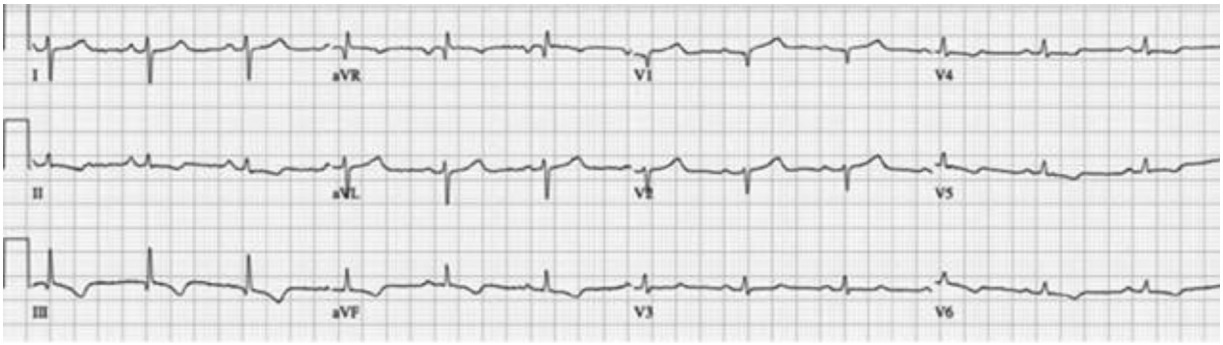


**Fig. 1 – The EKG shows low voltages.**

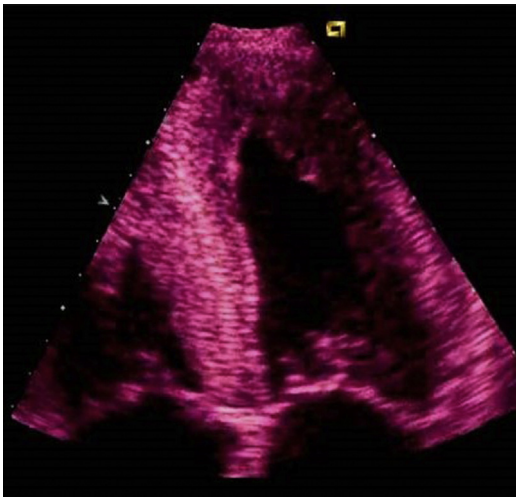

**Fig. 2 – The transthoracic echocardiogram four chamber view shows the ground glass texture of the left ventricle walls and the thickness of the mitral valve.**

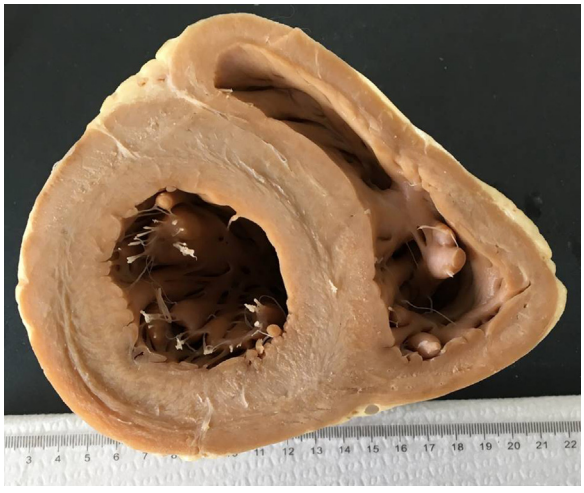

**Fig. 3 – The short axis section of the replaced heart shows severe biventricular concentric hypertrophy with midwall infiltration by amyloidosis.**

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