Iatrogenic transthyretin cardiac amyloidosis after sequential liver transplantation

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Introduction: Sequential liver transplantation (SLT) uses livers excised from patients with hereditary transthyretin-related amyloidosis during liver transplantation as grafts to other patients with severe hepatic pathologies and a reserved prognosis. We intended to investigate the development of cardiac manifestations consistent with iatrogenic transthyretin amyloidosis (iATTR).

Methods: We retrospectively analyzed the medical records of 72 consecutive patients submitted to SLT between 2007 and 2010, who received livers with V30M mutation.

Results: Our sample had 79% male patients and a mean age at transplantation of 55±6 years. Median follow-up time was 80 months, were 44% of the patients died. One-year mortality rate after SLT was 7%. Clinical manifestations of iATTR occurred in 29% of individuals, on average 6 years after SLT, and amyloid was seen in 76% of those who underwent a biopsy. Left ventricular hypertrophy (LVH) was identified in 42 (58%) patients at baseline. Considering 39 patients that had an echocardiography at baseline and during follow-up, 22 (61%) presented de novo LVH or basal LVH worsening during follow-up, with a significant increase of wall thickness (11±1 to 13±3 mm; $p<0.001$). They had similar age at presentation (55±5 vs 58±5, p=0.249) and incidence of hypertension (52% vs 64%, p=0.365) but higher incidence of chronic kidney disease (CKD; 68% vs 29%, p=0.023). Mortality during follow-up was higher in patients with de novo LVH or worsening LVH but not significantly, probably due to the sample size (23% vs 7%, p=0.221, log rank test p=0.262). Considering the global sample, significant conduction changes were rarely seen (1 patient); however, there was a trend towards an increase in PR interval and atrial fibrillation was reported in 8% of cases.

Conclusions: In our sample, probable iATTR was often seen within a decade after SLT. Further investigation of LVH needs to be made in these patients, as it can represent amyloid cardiomyopathy, but other contributing factors such as hypertension, CKD and age need to be taken into consideration. In our sample, development of a possible infiltrative pattern was relatively more common and conduction disorders were rarer than one would extrapolate from hereditary early onset ATTR V30M patients. Further studies may help us clarify if indeed these patients behave like late onset ATTR V30M. Our data suggests that these patients should probably undergo periodic cardiac imaging during follow-up.