Amyloidosis is a progressive systemic disease affecting a variety of tissues and organs, including the heart. In a subset of patients, the disease is hereditary because of a mutation in TTR, the gene encoding transthyretin, a transport protein of thyroxin and retinol-binding protein. In the case of advanced disease, liver transplantation is a treatment option to remove the source of the abnormal transthyretin. The explanted liver is often used for patients with liver insufficiency (domino transplantation). Amyloidosis after receiving a liver allograft donated by a patient with ATTR amyloidosis is an established, albeit relatively uncommon phenomenon. However, even if it does occur, it usually takes many years for overt amyloidosis to develop in the recipient, reflecting the slow development of amyloidosis in donor patients with a TTR mutation, which legitimizes the use of these livers for transplantation. Moreover, the ensuing amyloidosis is usually confined to neuropathy. In fact, we are aware of only a single report of amyloidotic cardiomyopathy, which was demonstrated at autopsy. The present case is remarkable for several reasons. First, it shows that amyloidosis may occur as early as 5 years after transplantation. Second, this is the first report of cardiac amyloidosis diagnosed ante mortem after receiving an ATTR liver. Finally, it shows that bone scintigraphy allows early diagnosis of cardiac involvement in patients with transthyretin-derived amyloidosis, even before conventional echocardiography.

Disclosures

None.

References


From Department of Cardiology (M.P.v.d.B.), Department of Nuclear Medicine and Molecular Imaging (R.H.J.A.S.), Department of Gastroenterology and Hepatology (H.B.), and Department of Rheumatology & Clinical Immunology (B.P.C.H.), University Medical Center Groningen, University of Groningen, The Netherlands.

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Correspondence to Maarten P. van den Berg, MD, PhD, Department of Cardiology, Thorax Center, University Medical Center Groningen, Groningen 9713GZ, the Netherlands. E-mail m.p.van.den.berg@umcg.nl

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Figure 1. Abdominal fat tissue specimen stained with Congo red dye (A) and red amyloid deposits in ordinary light showing green birefringence when viewed in polarized light (B).

Figure 2. Bone scintigram with 99mTc-hydroxyethylene diphosphonate showing uptake in several soft tissues as well as the heart.
Transthyretin-Derived (ATTR) Amyloidotic Cardiomyopathy After Receiving a Domino Liver Allograft
Maarten P. van den Berg, Riemer H.J.A. Slart, Hans Blokzijl and Bouke P.C. Hazenberg

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**Movie Legend**

**Movie 1.** Echocardiographic cine images 7 years after transplantation, parasternal long axis view. Best viewed with Windows Media Player.

**Movie 2.** Echocardiographic cine images 7 years after transplantation, apical 4-chamber view. Best viewed with Windows Media Player.

**Movie 3.** Echocardiographic cine images 9 years after transplantation, parasternal long axis view. Best viewed with Windows Media Player.

**Movie 4.** Echocardiographic cine images 9 years after transplantation, apical 4-chamber view. Best viewed with Windows Media Player.

**Movie 5.** Echocardiographic cine images 9 years after transplantation, apical 2 chamber view (zoom). Best viewed with Windows Media Player.