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LETTER TO THE EDITOR

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Successful sequential orthotopic liver transplantation in the treatment of familial amyloidotic polyneuropathy

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Sir: Familial amyloidotic polyneuropathy (FAP) is an autosomal, dominant, inherited disease that affects the peripheral and autonomous nervous system as well as the heart, the kidneys, and the intestinum. Its prognosis is lethal. Symptoms usually occur after the second decade of life and lead to death within 5–7 years.

Orthotopic liver transplantation has been described as the treatment of choice for FAP [4]. The native liver of these patients is morphologically and functionally intact except for the production of a pathologic protein that causes amyloidosis 20 years following exposure to the protein [3]. It is conceivable that such a liver could be used as a donor organ for patients not eligible for liver transplantation because of advanced pathology (e.g., malignancy).

We describe here a clinical situation in which a FAP patient received a liver from a cadaveric donor. The excised native liver of the patient was transplanted into a patient suffering from hepatocellular carcinoma and cirrhosis due to hepatitis C.

The first patient was a 34-year-old female with a family history of amyloidotic polyneuropathy type I. Neurologic symptoms with gastrointestinal paresis, bladder dysfunction, and peripheral polyneuropathy were demonstrated by electromyographic studies. Abnormal depositions of marked serum amyloid P (SAP) were found in the spleen, kidneys, and adrenal glands by scintigraphy. Transthyretin methionine 30, the pathologic protein, was detected by immunoblot testing and PCR. The patient was in stage IA of the disease with orthotopic hypotension, sensory deficit in the lower extremities, neurogenic bladder dysfunction, and constipation. She received a cadaver liver with the use of a venovenous bypass. The native liver, which was morphologically completely intact, was excised including the retrohepatic vena cava. Simultaneous intra-arterial and intraportal flushes were performed in situ with 500 ml UW solution each. The cadaver liver was implanted with an end-to-end vena cava anastomosis to the suprarenal and infradiafragmatic vena cava of the recipient and end-to-end portoportostomy, an end-to-end anastomosis of the arteria hepatica, and a choledochocholedochostomy.

In the second patient, who was a 60-year-old male with a multifocal hepatocellular carcinoma, preoperatively staged as T4N0M0 with underlying hepatitis C cirrhosis, a cava preserving hepatectomy was performed following informed consent by the patient about the origin of the donor liver and the natural history of amyloidotic polyneuropathy. The transplantation was performed using a laterolateral cavocavostomy without venovenous bypass [2].

The postoperative course in both patients was uneventful. Liver parameters normalized rapidly and both patients received conventional immunosuppression consisting of cyclosporin A, azathiopirine, and prednisone. Patient one was discharged from the hospital after 27 days and patient two 20 days following liver transplantation.

These results show that the procedure can be safely performed. Similar experiences have been reported in a recent series of endemic regions including Portugal [1]. While conservative therapy in FAP patients remains disappointing, orthotopic liver transplantation has been shown to be successful in the clinical management of the disease. Since it is known from the natural history of the disease that symtoms occur 20 years following exposure to the pathologic protein transthyretin, the morphologically intact liver of the FAP patient can be used as a donor organ in selected patients after carefully informing the patient and receiving informed consent. It seems appropriate to use these livers in aged patients for second choice indications, such as liver failure with advanced stage hepatocellular carcinoma and cirrhosis due to viral proliferation, which can recur in the transplanted liver. The procedure can contribute to a better understanding of the disease, its natural history, and the pathogenesis of FAP. A follow-up of these patients, including a quantification of the biochemical markers of the disease and neurological and electromyographic investigations, should be included.

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