A 23-month-old boy presented with a 3-month history of progressive abdominal distension and anorexia without vomiting or pain. There was no past medical history.

A painless mass occupying the entire right upper quadrant was felt on physical examination. No associated fever, jaundice or lower limb edema were noted. Blood and liver tests as well as α-fetoprotein levels were normal. Hepatitis B and C serology were negative. Abdominal ultrasonography and MRI revealed a heterogenic intra-hepatic multi-septa cystic formation, measuring 12.6 cm in its largest diameter and involving the right liver (fig. 1A, B). No extra-hepatic lesions were found. A right hepatectomy was performed to ensure complete resection of the mass (fig. 2). The post-operative course was uneventful. Histopathology examination of the resected specimen revealed an intra-hepatic encapsulated tumor composed of numerous cysts lined by endothelium and containing a serous acellular fluid. The diagnosis of hepatic lymphangioma was established. There was no recurrence two years after surgery.

Fig. 1. A, B MRI showing a huge cystic lesion (12.6 × 9.4 × 11.5 cm) with multiple septa involving the right liver. There is no invasion of the main hepatic vessels and no dilation of the intra- or extra-hepatic ducts.
Hepatic lymphangioma is an extremely rare benign disease (with only 1 reported case in infants) [1]. It is usually associated with lymphangiomas of other viscera. It could result from a developmental defect in the lymphatic vessels or in its communication with the venous system [2, 3]. Surgical excision is recommended [3]. No recurrence has been reported after complete excision.

References