Liver fibrosis in primary intestinal lymphangiectasia: An undervalued topic

Raffaele Licinio, Mariabeatrice Principi, Enzo Ierardi, Alfredo Di Leo

Abstract
The relationship between primary intestinal lymphangiectasia (PIL) and liver fibrosis is an emerging topic with many obscure aspects due to the rarity of the disorder. A recent paper reported that a six-month low-fat diet improved liver fibrosis. We report the case of a 17-year-old girl affected by PIL whose hepatic fibrosis progressively worsened within one year, despite dietetic support. This and the previous case report describe extraordinary events, which do not allow clear-cut clinical aspects to be established. Nevertheless, both cases suggest that in patients with PIL, it is necessary to closely monitor liver morphology with in-depth investigations including not only ultrasonography, but also elastography.

© 2014 Baishideng Publishing Group Inc. All rights reserved.

Key words: Hepatic transient elastography; Liver fibrosis; Low-fat diet; Primary intestinal lymphangiectasia

Core tip: The relationship between primary intestinal lymphangiectasia and liver fibrosis is an emerging topic with many obscure aspects due to the rarity of the disorder. The fibrosis outcome after a low-fat diet in the patient described in this report is in contrast with other literature reports. We emphasize the need for systematic monitoring of liver fibrosis in primary intestinal lymphangiectasia.

INTRODUCTION
Primary intestinal lymphangiectasia (PIL), featuring a dilatation of intestinal lymphatic vessels and malabsorption, is a rare condition often requiring nutritional enteral/parenteral support[1]. Enteral nutrition is based on a hyperproteic, low-fat diet with vitamin and medium-chain triglyceride supplementation[2]. The association between PIL and primary liver fibrosis is uncommon[3], however, Milazzo et al[4] recently reported a case of associated PIL and liver fibrosis characterized by high stiffness at elastography. The authors reported that a six-month low-fat diet combined with medium-chain triglyceride supplementation improved liver alterations by reducing fibrosis. The authors attributed the fibrosis onset to lymphatic stasis, as occurs in cardiac congestive liver. However, since fibrosis reversibility has not been previously described in this condition, the hypothesis may be purely speculative. In this scenario, we believe our case of PIL featuring progressively worsening hepatic fibrosis, despite dietetic support, may be of interest.

CASE REPORT
A 17-year-old female patient was admitted to our unit for peripheral and facial edema, ascites and intestinal malab-
The rarity of PIL and the extraordinary events surrounding its uncommon association with liver fibrosis are exhibited by the present case, thus preventing the establishment of clear-cut clinical characteristics. Indeed, this report demonstrates that liver fibrosis may not improve after nutritional therapy. Nevertheless, this and a previous case\cite{1} suggest that in patients with PIL, it is necessary to closely monitor liver function, with in-depth investigations including not only ultrasonography, but also elastography\cite{2}. Early detection of liver involvement in PIL is important in order to promote regression and prevent progression towards portal hypertension and recurrent cholangitis.

**REFERENCES**

1. Vignes S, Bellanger J. Primary intestinal lymphangiectasia (Waldmann’s disease). Orphanet J Rare Dis 2008; 3: 5 [PMID: 18294365 DOI: 10.1186/1750-1172-3-5]
2. Desai AP, Guvenc BH, Carachi R. Evidence for medium chain triglycerides in the treatment of primary intestinal lymphangiectasia. Eur J Pediatr Surg 2009; 19: 241-245 [PMID: 19440223]
3 Chagnon JP, Burge J, Hay JM, Devars du Mayne JF, Ricahrd JP, Hardouin JP. Congenital hepatic fibrosis, multiple renal cysts and primary intestinal lymphangiectasia (author’s transl). *Gastroenterol Clin Biol* 1982; 6: 326-332 [PMID: 7084582]

4 Milazzo L, Peri AM, Lodi L, Gulbertini G, Ridolfo AL, Antonori S. Intestinal lymphangiectasia and reversible high liver stiffness. *Hepatology* 2014; 60: 759-761 [PMID: 24449480 DOI: 10.1002/hep.27025]

5 Colli A, Fraquelli M, Casazza G, Conte D, Nikolova D, Duca P, Thorlund K, Gluud C. The architecture of diagnostic research: from bench to bedside--research guidelines using liver stiffness as an example. *Hepatology* 2014; 60: 408-418 [PMID: 24277656 DOI: 10.1002/hep.26948]
