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Clinical profile, response to therapy and outcome of primary intestinal lymphangiectasia in children

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Objective: Intestinal lymphangiectasia (IL, primary or secondary) is an important cause of protein-losing enteropathy. We evaluated the clinico-laboratory profile, response to therapy, complications and outcome of children with primary IL (PIL).

Methods: Consecutive children (≤18 years) diagnosed with PIL (clinical setting, typical small bowel histopathology and exclusion of secondary causes) from 2007 to 2017 were evaluated.

Results: 28 children with PIL (16 boys, age at symptom onset-12 months and at diagnosis 8 years) were studied. Pedal edema (93%), chronic diarrhea (78.6%) and recurrent anasarca (64%) were the common presentations. Ascites, pleural and pericardial effusion was seen in 64% (n-18; chylous-5, non-chylous-13), 18% and 18% cases respectively. Hypoproteinemia, hypoalbuminemia, hypocalcaemia and lymphopenia were present in 82%, 82%, 75% and 39% cases respectively. Duodenal biopsy established the diagnosis in 86% cases, while 14% required distal small bowel biopsies. Dietary therapy was given in all and 6 cases required additional therapy (octreotide-6, tranexamic acid-3 and total parenteral nutrition-1). Lymphedema (3/5 vs. 1/23), pleural effusion (4/5 vs. 1/23) and need of additional therapy (4/5 vs. 2/23) was significantly more common in patients with chylous ascites (n=5) than those without chylous ascites (n=23). 24 cases were in follow-up for 39 (6-120) months and showed improvement, however 8 required readmission (symptom recurrence-6 [25%], complication- 2 [8.3%, Budd Chiari Syndrome-1 and abdominal B cell lymphoma-1]).

Conclusion: Presence of chylous ascites suggests severe disease in children with PIL. Majority of PIL children respond to dietary therapy; only 20% need additional therapy. Symptom relapse and complications occur in 1/3rd of cases and need long-term follow-up.

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