Gastrointestinal lymphoid pseudotumoral hyperplasia: report of four pediatric cases

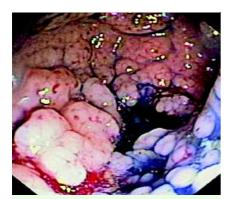


Fig. 1 Endoscopy aspect of lymphoid nodular hyperplasia in the left colon after chromoendoscopy using indigo carmine dye.

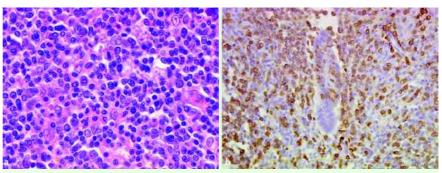


Fig. 3 a Microscopic features: small lymphoid cells, mainly centrocytic-like, rarely with plasma cell differentiation (hematoxylin–eosin–saffron stained section, magnification × 400). **b** Immunostaining with anti-CD20: cells expressed CD20. Focal invasion of the crypt epithelium reminiscent of lymphoepithelial lesions (magnification × 100).



Fig. 2 Video capsule appearance of the jejunal lesions.

We report on four pediatric patients (Table 1, Fig. 1–3) with lymphoid pseudotumoral hyperplasia (LPH), two with an atypical localization (diffuse disease). The lesion diameters varied from 7 to 30 mm.

In our patients, the treatment was: conservative (patient 1), surgery (patient 2), corticosteroids (patient 3), and a milk-free diet (patient 4). Complete remission of lesions and/or clinical improvement were seen in all cases. In the three clinically managed cases, there were no changes during the follow-up (6 years for patient 2, 4 years for patient 3, and 6 months for patient 4).

Lymphoid nodular hyperplasia (LNH) is a common, nonpathologic finding in children [1]. The ileum and colon are commonly affected [1,2]. An atypical form is

the pseudotumoral presentation, defined as a tumoral presentation of LNH associated with inflammatory mesenteric lymph nodes [1,2]. The imperative differential diagnosis is low-grade lymphoma. In such cases, a diagnosis of malignancy is based on morphologic and immunophenotypic (histologic) features with molecular evidence of clonality [3].

LPH and LNH are similar, differing only in their endoscopic appearance. LNH appears as micronodules 1-2 mm in size, of a whitish-rose color, surrounded by normal mucosa [1]. In LPH, the micronodular endoscopic appearance is absent, giving a misleading picture of tumor [2] - as was observed in our cases. It is very important to take a large number of biopsies (sometimes macrobiopsies) to establish the diagnosis [2,4]. The physiopathology is not completely understood, but probably reflects an unspecific exacerbated lymphoid response to various stimuli (e.g., infections, allergies, immunodeficiency, autoimmunity) [1,2], which were identified in three-quarters of our patients.

There is no consensus about treatment, which is reserved for patients with severe symptoms. Treatments include surgery and hydrostatic barium enema (in the case of intestinal obstruction or intussusception), or the use of corticosteroids (with previous histologic confirmation of LPH) [2,5]. Due to its rarity, the natural history, and the increased risk of malignant transformation, many aspects such as clinical and endoscopic features and treatment follow-up remain uncertain.

Endoscopy_UCTN_Code_CCL_1AD_2AC

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 Table 1
 Clinical, histologic, and endoscopic features of the patients.

| Age (y) | Symptoms | Topo- graphic data | Diagnostic method | Lesion diameters | Immunohisto- chemistry | | Etiology | Treat- ment | Response | Follow-up (years) |
|------------|--------------------------------|---|---|---------------------|---------------------------|----------|-----------------------------|----------------------|---|----------------------|
| | | | | | Bcl-2 | CD20+ | | | | |
| 4 | diarrhea, hemato- chezia | esophagus, stomach, duode- num, ileum, colon | gastros- copy, colonos- copy | 20 – 30 mm | negative | negative | allergy? | - | sponta- neous CI | 6 |
| 7 | intussus- ception | ileum, colon | surgery | 15 – 20 mm | negative | positive | HHV6 virus | surgery | good, no recurrence | 2 |
| 5 | abdominal pain | jejunum, ileum, colon | gastros- copy, colo- noscopy, video capsule | 7 – 15 mm | negative | positive | immuno- suppres- sion | cortico- steroids | good, LPH improve- ment, no recurrence | 4 |
| 12 | rectal bleeding | colon | gastros- copy, colo- noscopy | 15 mm | negative | positive | milk allergy | diet | Cl | 0.6 |

y = years, mm = millimetres, CI = clinical improvement.

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DOI 10.1055/s-2008-1077668 Endoscopy 2008; 40: E267 − E268 © Georg Thieme Verlag KG Stuttgart · New York · ISSN 0013-726X

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