Primary Intestinal Lymphangiectasia with Predominant Ileal involvement Diagnosed by Ileocolonoscopy

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ABSTRACT

Intestinal lymphangiectasia is a rare disease of intestinal lymphatics presenting with hypoproteinemia, bilateral lower limb edema, ascites, and protein losing enteropathy. It is most commonly diagnosed on the endoscopic appearances and corresponding histology of the duodenum. We report a 8-year-child with chronic diarrhea, anasarca and hypoalbuminemia in whom duodenal endoscopic findings and histology were normal and the diagnosis of intestinal lymphangiectasia was made by endoscopic ilial examination and biopsy. (J Dig Endosc 2010;1(3):155-7)

Key words: Intestinal lymphangiectasia – Protein losing enteropathy – Hyproteinemia – Ileoscopy - Duodenal biopsy

Introduction

Intestinal lymphangiectasia is a rare disease characterized by diffuse dilatation and proliferation of enteric lymphatic channels which was originally described in 1961 by Waldmann et al in a series of 18 patients as “idiopathic hypoproteinemia”. Dilated intestinal lymphatics causes leakage of lymph into the small intestinal lumen and results in protein-losing enteropathy leading to hypoproteinemia, lymphopenia and hypogammaglobulinemia. The hypoproteinemia leads to bilateral leg edema, ascites, and pleural effusion. It is either primary (idiopathic) or secondary. Primary intestinal lymphangiectasia (PIL) as described by Waldmann et al predominantly occurs in children and adolescents due to congenital deformity of the lymphatics of the small intestines, whereas secondary intestinal lymphangiectasia is commonly seen in adults as a result of raised lymphatic pressure as seen in lymphoma, malignancy, inflammatory bowel disease constrictive pericarditis and cardiac surgery. PIL is most commonly diagnosed by endoscopic appearances of lymphangiectasia with characteristic histology of the duodenal biopsy; however in some instances due to patchy nature of the disease duodenal mucosal findings as well as corresponding biopsy may be normal and other modalities like enteroscopy may be required to diagnose this entity.

We report a case of chronic diarrhea with anasarca in a 8-year-old child in whom the duodenal biopsy was normal and diagnosis of PIL was made by endoscopic ilial examination and biopsy.

Case report

A 8-year-old boy presented with history of pain abdomen on and off for six months which was colicky in nature without history of vomiting or features suggestive of subacute intestinal obstruction. There was progressively increasing anasarca for 4 months. There was no history of decrease in urine output, diarrhea, rash, blood in stool, worm infestation, cough, or orthopnea. CBC revealed a hemoglobin of 12.4 g/dl with normal other indices. Serum protein was 4.2 gm/dl, and serum albumin was 2.2 gm/dl with normal
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transaminases. Serum urea and creatinine were normal. The results of virologic markers such as HBsAg, Anti-HCV and HIV were negative. Anti-tissue transglutaminase antibody (IgA Ttg) assay was normal. Urine analysis was normal. Abdominal ultrasonography showed gross ascites with mild thickening of small bowel loops, Upper GI endoscopy was normal and biopsies from the duodenum did not show any abnormality.

In view of continuing symptoms and presence of low total protein and albumin, normal hemoglobin, normal upper GI endoscopy and duodenal biopsy, a possibility of protein loosing enteropathy was entertained. Barium meal follow through was suggestive of malabsorption with thickening of intestinal folds but no features of strictures. Due to non-availability of enteroscope, a colonoscopy with ileoscopy was performed. The whole colon was normal but the whole terminal ileum showed multiple whitish creamy irregular patches of variable sizes scattered in whole of the terminal ileum (Figure 1). Biopsy report from the ilial lesions showed the dilated lymphatics in the lamina propria with dilated and enlarged villi (Figure 2), the histologic features typical of the intestinal lymphangiectasia.

Discussion

PIL is a rare disease which primarily affects children usually before 3 years of age but some cases may be present in

Table 1: Published data from India on the Intestinal lymphangiectasia

<table>
<thead>
<tr>
<th>Authors</th>
<th>No of cases</th>
<th>Age at Diagnosis</th>
<th>Presenting</th>
<th>Associations</th>
<th>Mode of diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Suresh et al, 2009</td>
<td>Four</td>
<td>36 days –9 years</td>
<td>Diarrhea and anasarca</td>
<td>Hennekam syndrome</td>
<td>Duodenal biopsy</td>
</tr>
<tr>
<td>Katoch et al, 2008</td>
<td>One</td>
<td>Six mo</td>
<td>Intussusception in small bowel</td>
<td>-</td>
<td>Post-operative biopsy</td>
</tr>
<tr>
<td>Makharia et al, 2007</td>
<td>Two</td>
<td>-</td>
<td>Diarrhea</td>
<td>Autoimmune poly glandular syndrome (APS) type 1</td>
<td>Duodenal biopsy</td>
</tr>
<tr>
<td>Sethuraman et al, 2006</td>
<td>One</td>
<td>-</td>
<td>-</td>
<td>Familial pachydermo- periostosis</td>
<td>Duodenal biopsy</td>
</tr>
<tr>
<td>Riyaz et al, 2004</td>
<td>One</td>
<td>8 year</td>
<td>Diarrhea</td>
<td>Hemihypertrophy and Incontinentia pigmenti acromians</td>
<td>Duodenal biopsy</td>
</tr>
<tr>
<td>Ranjan et al, 2004</td>
<td>One</td>
<td>-</td>
<td>Diarrhea</td>
<td>-</td>
<td>Duodenal biopsy</td>
</tr>
<tr>
<td>Sharma et al, 2000</td>
<td>One</td>
<td>-</td>
<td>Diarrhea</td>
<td>-</td>
<td>Jejunal biopsy by push enteroscopy</td>
</tr>
<tr>
<td>Puri et al, 1992</td>
<td>One</td>
<td>40 year*</td>
<td>Diarrhea</td>
<td>-</td>
<td></td>
</tr>
</tbody>
</table>

Secondary intestinal lymphangiectasia
young adults or even later in adults. It aetiology remains unknown.

Intestinal lymphangiectasia is diagnosed based on typical endoscopic findings of “snow flake appearance” with the corresponding characteristic histology of intestinal biopsy of dilated lymphatics. Upper GI endoscopy may be negative when intestinal lesions are segmental or localized. In such cases enteroscopy is a useful tool to detect the presence of intestinal lymphangiectasia and to specify its localization. Similarly in our case the upper GI endoscopy and duodenal biopsy were normal. In our country where the facilities for pediatric enteroscopy and capsule endoscopy are not readily available, the endoscopic ilioscopy can be useful in picking up the lesions of intestinal lymphangiectasia as in our case. These different diagnostic tools have been used to acquire the tissue for the diagnosis of IL but in none of the published data from India the iloscopy and ileul biopsy has been used for its diagnosis. Table summarizes the Indian experience on the disease.

In conclusion patients with features of protein-losing enteropathy and suspicion of intestinal lymphangiectasia, a normal upper GI examination of the duodenum and duodenal biopsy can not rule out intestinal lymphangiectasia but it may be diagnosed on iloscopy because the gut involvement may be more distal.

References


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