Case Reports

Intestinal Lymphangiectasia

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In the pediatric age group hypoproteinemia and its consequences are commonly attributed to protein energy malnutrition, hepatic disease and nephrotic syndrome. We report a child where loss of proteins was occurring from a most unusual site, the gut, inspite of diarrhea not being the major presenting symptom.

Case Report

A 4 year old girl presented with generalized swelling of the body and diarrhea, with previous 2 similar episodes 6 months and 18 months back. Past records revealed hypoproteinemia with normal hepatic and renal functions with a partial and transitory response to diuretics and high protein diet. Examination revealed a stunted child (height 25-50th percentile) with anasarca (mild pitting edema of the feet and marked ascites). Investigations revealed an absolute lymphocyte count of 2460/cu mm, total serum proteins 2.8 g/dl, albumin 1.5 g/ dl, globulin 1.3 g/dl, and normal hepatic and renal functions. The ascitic fluid was a transudate and not chylous. Narrowing down the Site of protein loss as the gut, protein losing enteropathy was considered, the most likely entity being intestinal lymphangiectasia. Esophago-gastroduodenoscopy revealed nodular duodenum with loss of normal folds. Duodenal biopsy showed bloated nonatrophic villous tips with dilated lymphatic channels in the superficial duodenal mucosa, strongly suggestive of intestinal lymphangiectasia *(Fig. 1).* Serum ceruloplasmin and IgG were low with normal IgA and IgM. Right lower limb lymphangiogram showed obstruction of lymphatics at upper lumber level with retrograde opacification of intestinal lymphatics in the right iliac fossa *(Fig. 2).* Lower lumbar lymph nodes were small and upper lumbar lymph nodes were absent. Ultrasonography and CT scan abdomen showed uniformly thickened jejunal folds with spiculation and distal dilution of barium without dilatation of loops.

On arriving at the diagnosis of intestinal lymphangiectasia the child was taken off diuretics and put on a milk free high protein, low fat, medium chain triglyceride diet in the form of coconut oil. In three weeks, there was marked decrease in ascites and increase in serum albumin from 1.5 g/dl to 2.5 g/dl. On the same dietary regime, her first follow up after 3 months showed a symptom free child and a further increase in serum albumin to 3.3 g/dl. Subsequently as the child insisted for milk, the mother introduced cow's milk in the child's diet. Within 10 days, the child rapidly became symptomatic with ascites. The serum albumin had dropped to 1.6 g/dl [total proteins 3 g/dl].

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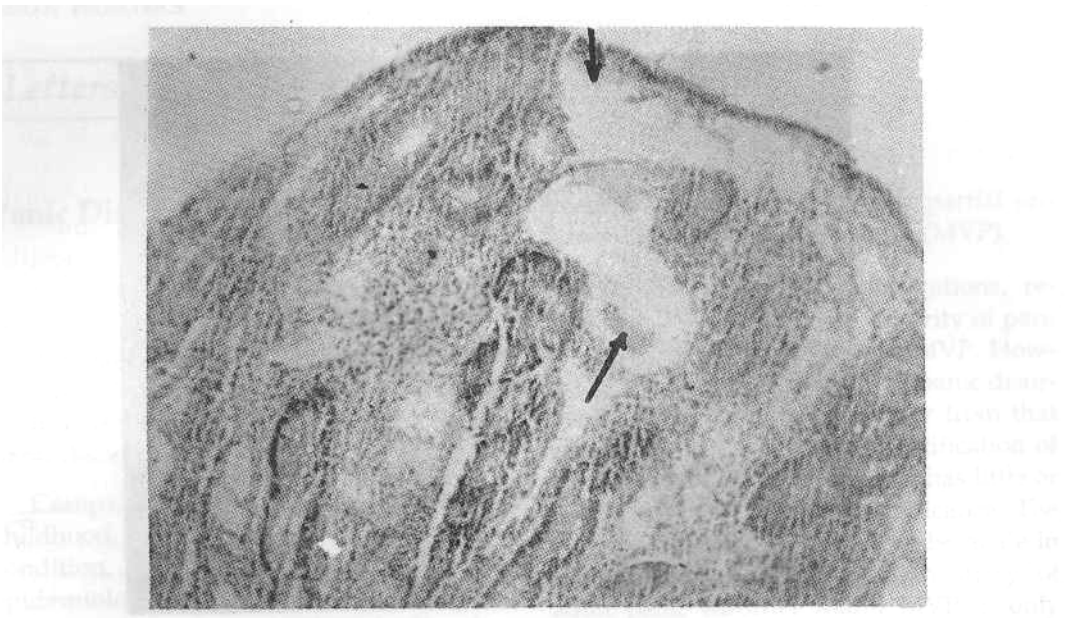

Fig. 1. Bloated non atrophic villous tips with dilated lymphatic channels in the superficial duodenal mucosa.

Discussion

Intestinal lymphangiectasia (IL) is characterized by congenital hypoplastic visceral lymphatic channels leading to obstruction of lymphatic flow and dilatation of intestinal lymphatics. Rupture of the dilated lymphatic venules with discharge of lymph into the bowel lumen results in hypoproteinemia, steatorrhea and lymphopenia(l). Dietary long chain triglycerides stimulate lymphatic flow thus worsening the condition.

IL classically has protein losing enteropathy, hypoalbuninemia and it's clinical manifestations. Symptoms may be severe and chronic or mild and transitory and may vary widely in its manifestations and severity(2). Diarrhea is a common symptom(2) though steatorrhea may be mild(l) and altered bowel rhythm may be conspicuous by its absence(3). Vomiting, growth retardation, peripheral lymphedema and hypocalcemic seizures are variable features(2).

Hypoproteinemia is a constant finding throughout the course of the disease. Chylous effusions is related to the severity of the disease and may first appear years after the protein loss begins(4).

In addition to proteins, immunoglobulins and ceruloplasmin are also lost leading to their low levels as also seen in our patient. Upper GIT series shows thickened bowel folds, spiculation, dilution of barium (increased secretion), without dilation. Lymphangiogram shows hypoplastic and blocked lymphatics as seen in our case. Biopsy is diagnostic and helps to rule out other conditions such as intestinal tuberculosis, Whipples disease, Crohns disease, eosinophilic gastroenteritis and celiac disease. Other methods of diagnosing IL include fecal alpha-1 antitrypsin estimation and radioactive albumin loss in the gut, both of which were not available in our city. The diagnosis of IL in our patient was confirmed by lymphagiogram and biopsy.

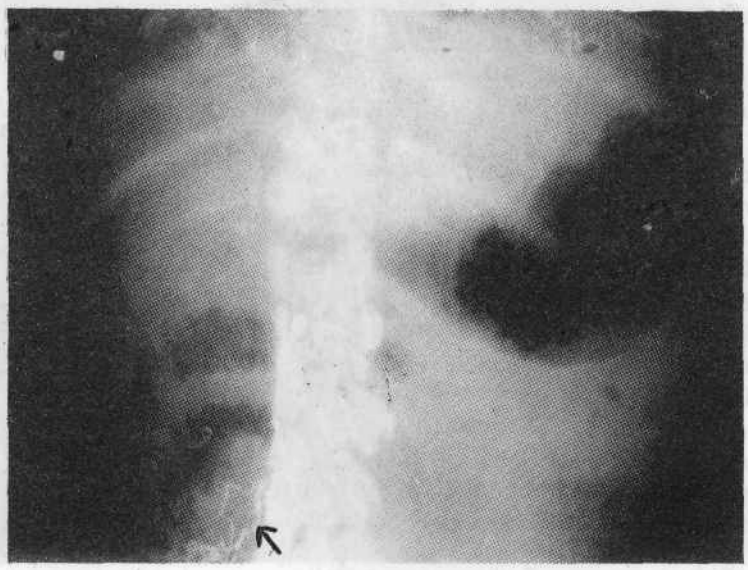

Fig. 2. Lymphangiogram showing obstruction of intestinal lymphatics at upper lumbar level with retrograde opacification of intestinal lymphatics in the right itiac bossa. Lower lumbar lymphnodes are small and upper lumbar lymphnodes are absent.

Treatment of IL with a high protein, fat free diet with added MCT, while supposedly having no effect on the underlying pathology, is usually effective in preventing or alleviating the diarrhea and hypoproteinemia. Absence of fat in the diet prevents engorgement of the intestinal lymphatics with chyle (fat is the stimulus for increased lymphatic secretion) thus preventing their rupture with its concomitant T cell and protein loss. MCT is mostly absorbed directly into the portal system, thereby providing nutrient fat but avoiding lacteal engorgement(5). Introduction of even small amounts of fat rich food or milk can precipitate relapse as seen in our patient.

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