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Intestinal Lymphangiectasia

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Intestinal lymphangiectasia is a rare congenital disorder associated with protein losing enteropathy. It presents more often

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in the first two years of life with diarrhea and failure to thrive, and later with generalized edema due to hypoproteinemia(I). We report here a boy with intestinal lymphangiectasia.

Case Report

A 2 year and 11 month old boy presented in December 1994 with a history of recurrent episodes of generalized swelling of the body and loose stools for two years. He had been treated with plasma transfusion and antituberculous treatment in the past. He was the only child to his non-consanguineous parents. At birth, he was noticed to have facial asymmetry. He was breastfed for one year and at admission was on diet consisting of cow's milk, rice, dhal, boiled

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eggs, fish, bananas and biscuits. His performance in nursery school was good.

On examination, his weight was 14 kg (50th centile), height 93 cm (25th centile) and head circumference 51 cm (75th centile). He was noted to have hemihypertrophy of the right side of face with prominent metopic suture (*Fig. 1*), bilateral pitting pedal and periorbital edema, gross ascites and a firm hepatomegaly of 3 cm. The rest of the physical examination was normal.

His hemoglobin was 11.9 g/dl, total white blood cell count was 4,900/cu mm, with a differential count of bandforms 2%, neutrophils 74% and lymphocytes 24%. Absolute lymphocyte count was 1176/cu mm (norm: 2000). Platelet count was

4,96,000/cu mm. Liver function tests showed a serum total bilirubin 0.4 mg/dl (norm: 0.2-1), direct 0.2 mg/dl (norm: 0.2-0.5), total protein 2.7 g/dl (norm: 6.5-8.5), albumin 1.4 g/dl (norm: 3.7-4.9), AST 356 U/L (norm: 40), ALT 327 U/L (norm: 40) and alkaline phosphatase 163 U/L (norm: 145-200). Serum Hepatitis B surface antigen was negative. Prothrombin time, serum electrolytes, calcium, cholesterol and creatinine were normal. Serum copper was 80 Hg/dl (norm: 70-170), and ceruloplasmin 63 U/L (norm: 60-140), alpha-1 antitrypsin 133 mg/dl (norm: < 150). Serum amylase was 153 Somogyi units (norm: 60-200) and 24 hour urinary amylase was 1540 Somogyi units (norm: 1500-7500). Stool occult blood was present in traces. Seventy two hour stool fat excretion was 6.3 g (norm: 6-7 g/



Fig. 1. Hemihypertrophy of the right side of face.

day). D-xylose absorption in blood after 1 hour was 2.91 mmol/L (norm: > 1.33 mmol/L). Sweat sodium and chloride were 35 mmol/L and 29 mmol/L, respectively (norm: sodium -58 + 16 and chloride-45 + 16). A repeat liver function test done two weeks later showed AST and ALT to be 21 and 12 U/L, respectively.

Mantoux test was negative. Chest radiograph showed mild haziness in the right paracardiac region, and abdominal radiograph was normal. Skull radiograph showed an incomplete posterior arch of atlas. Ultrasound examination of the abdomen showed bulky tail of pancreas (a normal variant) and ascites. Liver biopsy showed mild non specific portal inflammation. Barium meal examination showed mild gastro-esophageal reflux and nodular filling defects in the duodenum. Endoscopic appearance of the upper gastrointestinal tract was grossly normal. Duodenal mucosal biopsy showed dilated lacteal with distortion of the villi (Fig. 2), characteristic of intestinal lymphangiectasia(2,3).

Therapy was started with a high protein, low fat diet and unrestricted amounts of medium chain triglycerides as coconut oil was used. Oral calcium lactate 300 mg, Vitamin A 6000 U and Vitamin D 1,000 U once daily were added.



Fig. 2. Duodenal mucosal biopsy illustrating distortion of the villi and dilation of the lacteals (arrow).

He came back for review 1 year and 5 months later. He had continued to have recurrent diarrhea although the frequency was less. He had developed 2 episodes of sudden loss of consciousness associated with tonic posturing in November 1995 and June 1996. His weight had increased marginally from 14 kg to 14.6 kg. He had mild pitting pedal edema. There was no ascites. Liver was palpable 2 cm below the costal margin. His serum protein had increased to 3.5 g/dl and albumin to 2.3 g/dl; AST, ALT and alkaline phosphatase were normal. Serum calcium and phosphorus were normal. Serum immunoglobulins done then showed low IgG and IgA levels: IgG - 374 mg/dl (norm : 800-1700); IgA - 25 mg/dl (norm : 140-420); and IgM - 59 mg/dl (norm: 50-190).

Electroencephalogram showed primary generalized epileptiform activity. He was started on sodium valproate. Diet restrictions, calcium and vitamin supplements were continued.

Discussion

Intestinal lymphangiectasia was originally described in 1961 by Waldmann *et al.*(4). Very few cases have been reported from India so far(5,6). It is a disorder characterized by diffuse or localized ectasia of the enteric lymphatics often in association with lymphatic abnormalities elsewhere in the body. The pathogenesis of abnormal lymphatic structure is uncertain. Ectatic lymphatics may be located in the mucosa, submucosa or subserosa, leading to loss of protein and lymphocytes into the gut or the peritoneal cavity. The mechanism of this lymphatic loss is believed to be from rupture of lymphatics across the mucosa with subsequent leakage of lymph into the bowel lumen(7).

Children with this disorder present throughout childhood but most often in the

first 2 years of life(1). It has been noted to occur in families. It may occur as an isolated abnormality or as part of a syndrome such as Noonan's syndrome or Klippel-Trenaunay-Weber syndrome. The former is a sporadic disorder with the salient features of short stature, short neck with webbing, cardiac anomalies, pectus carinatum and a characteristic facies; the latter is a sporadic condition featuring cutaneous nevus in an asymmetrical distribution, varicosities and asymmetrical hypertrophy of all or part of a limb(7).

Edema may be asymmetric and non pitting due to lymphatic abnormality of the affected extremity. The whole lymphatic system below the diaphragm may be grossly abnormal and outflow towards the thoracic duct may be obstructed(8).

In addition to the loss of albumin, patients with lymphangiectasia also frequently have reduced levels of immunoglobulins and a low circulating lymphocyte count, particularly T-lymphocytes due to lymphatic loss(9). Steatorrhea may be demonstrable on a fecal fat estimation. Barium studies may demonstrate thickening of the jejunal folds, fluid hypersecretion and nodular or punctate lucencies in the mucosa of the small bowel(10). Endoscopic abnormalities such as scattered white plaques or the presence of chyle-like substances covering the mucosa have been observed, but may not always be present.

Our patient was symptomatic from late infancy. He had generalized edema and marked hypoalbuminemia. No other cause for the hypoproteinemia could be demonstrated apart from intestinal lymphangiectasia. The diagnosis of intestinal lymphangiectasia in him is suggested by the clinical findings, supported by the presence of hypoproteinemia, lymphocytopenia, hypogammaglobulinemia, filling defects

in barium meal, and confirmed by dilated lacteals on small intestinal biopsy.

The characteristic abnormalities of the above mentioned syndromes were absent in our patient. Further, there was no history of similar illness in the family. He had a firm hepatomegaly with elevated liver enzymes which resolved spontaneously in a few weeks. Liver biopsy showed mild nonspecific portal inflammation. Therefore a concurrent resolving hepatitis was considered the most likely cause for the hepatomegaly.

The mainstay of treatment for intestinal lymphangiectasia is the use of a low fat, high protein diet rich in medium chain triglycerides. Medium chain triglycerides are not reesterified within the intestinal cell and thus bypass the enteric lymphatics and directly enter the portal system. It is believed that the reduction in long chain fats reduces lymphatic flow and pressure within the lymphatic system and decreases the amount of lymph leakage(7). Severe steatorrhea may result in hypocalcemic tetany. Therefore, some children require additional supplementation with calcium salts, in addition to water soluble forms of fat soluble vitamins(11).

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Arthritis in Hypogammaglobulinemia

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Majority of children with hypogammaglobulinemia present with recurrent and

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severe lower respiratory tract infections and the diagnosis in such cases is usually clinically obvious. In some children, however chest infection may not be very prominent at presentation and instead joint involvement may be the major clinical finding. Such patients have been misdiagnosed as having juvenile rheumatoid arthritis(l). We report two such cases whom we treated recently. This type of presentation of hypogammaglobulinaemia has not been previously reported in the Indian literature.

Case Reports

Case 1: A five-year-old boy presented with history of swelling in various joints for last 9 months for which he had been receiving treatment elsewhere To begin with he had swelling of left shoulder joint which was painful and was associated with fever. Arthrocentesis was done as aspirate showed a purulent exudate. He underwent arthrotomy and received antimicrobials for