INTESTINAL LYMPHANGIECTASIA A CASE REPORT

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Abstract

A 28 year old woman with protein losing enteropathy due to intestinal lymphangiectasia is presented. The patient had diarrhoea, oedema restricted to the lower extremities, hypoproteinemia and an abnormal radiological appearance of the small intestines.
Jejunal biopsy and the histopatho-logy report of the resected jejunum showed dilated lymphatics in the submucosa.
Post-operatively patient showed a marked improvement in her general condition with an improvement in appetite, disappearance of oedema and an increase in her serum protein levels. Introduction of a medium chain triglyceride diet had a beneficial effect with a significant reduction in the frequency of bowel movements.
Hypoproteinemia of obscure origin may be due to loss of plasma proteins into the gastrointestinal tract and the presence of a lymphatic disorder of the small bowel as a cause of the syndrome in such cases should be entertained. (JPMA 30:231, 1980).

Introduction

An abnormal gastrointestinal protein loss has been seen in association with a wide variety of gastrointestinal disorders including giant gastric rugae, gastric carcinoma, sprue, Whipple's disease, regional enteritis and ulcerative colitis. Loss of proteins into the gastrointestinal tract has been shown to be a major factor in the so called idiopathic hypoproteinemia, a syndrome of hypoproteinemia with oedema but without proteinuria and with normal hepatic function. In most of these patients, the excessive protein loss into the bowel has been shown to be secondary to a disorder of the lympahtics of the small intestine. This condition has been termed as Intestinal lymphangiectasia (Waldmann et al., 1961), and is characterized by marked dilatation of lymphatics of the mucosa and mesentery of the small intestine, manifested by an extreme loss of proteins into the gastrointestinal tract, resulting in hypoalbuminaemia, hypogammaglobulinemia, oedema and effusions (Pomerantz and Waldmann, 1963).
The present report describes a case of intestinal lymphangiectasia, together with a comparative study of the clinical features with other cases of intestinal lymphangiectasia reported in the literature.

Case Report

S.Z. a 28 year old married female gave a history of anorexia and postprandial abdominal distention together with passage of 15 to 20 bulky stools per day, since childhood.
In August 1965, she was admitted to a hospital with an attack of severe epigastric pain, and was treated symptomatically for about a month during which she also received psychiatric treatment. She was also noted to have oedema of her legs, which responded to treatment.
Four years later, she again developed severe oedema of her lower extremities during her lirst pregnancy, which persisted after the delivery. She also had bouts of diarrhoea accompanied with abdominal distention. Oedema was treated with diuretics, and as a result she developed cramps and body aches.
Five years later in May 1974, she was seen at Ganga Ram Hospital Lahore, where she was noted to

have a waddling gait, swollen and infected gums, abdominal distention with ascites and an umbilical hernia, hepatosplenomegally and oedema of her legs together with bone and calf muscle tenderness. She weighed 95 lbs. Investigations revealed a haemoglobin of 10 gm% with moderate hypochromia and total proteins amounted to 3.7 gm'%, albumin being 1.3 gm% and globulin 2.4 gm% (Table I). Liver biospy revealed no significant changes except for a slight cellular infiltration in the portal areas. Patient was seen at the P.M.R.C. Research Centre, Jinnah Postgraduate Medical Centre, Karachi in December 1974. She was noted to have a sallow complexion and dry skin, with thin upper limbs as compared to her oedematous lower limbs. She had divarication of her recti and an umbilical hernia, but no ascites. No visceromegaly was detected and her systemic examination was normal. She then weighed 86 lbs and had a blood pressure of 90/60 mm of Hg. Investigations revealed a haemoglobin of 13.4 mg% with a PCV of 39%. Total and differential leucocyte and platelet counts were normal. Bone-marrow was hyperplastic with a mild normoblastic hyperplasia. Mega karyocytes were normal and only traces of marrow iron were detected. Total bilirubin was 0.5 mg%, alkaline phosphatase 2.6 BLU, SGOT 20 mu/ml and SGPT 6.5 mu/ml. Total proteins were 4.2 gm/% with albumin being 2.2 gm/%only (Table I). Serum calcium was 3.0 meq/litre(4.4-5.3) and cholesterol 105 mg%. Glucose tolerance test showed a flat curve and D-xylose excretion was 1.0 gm/5 hours. Faecal fat content was 3.86 gm/24 hrs. Barium meal examination revealed coarse oedemaiious jejunal mucosa with some dilated loops. X-rays of chest and long-bones were normal. Capsule biopsy of the jejunum showed dilated lymphatics, as shown in the accompanying figure.

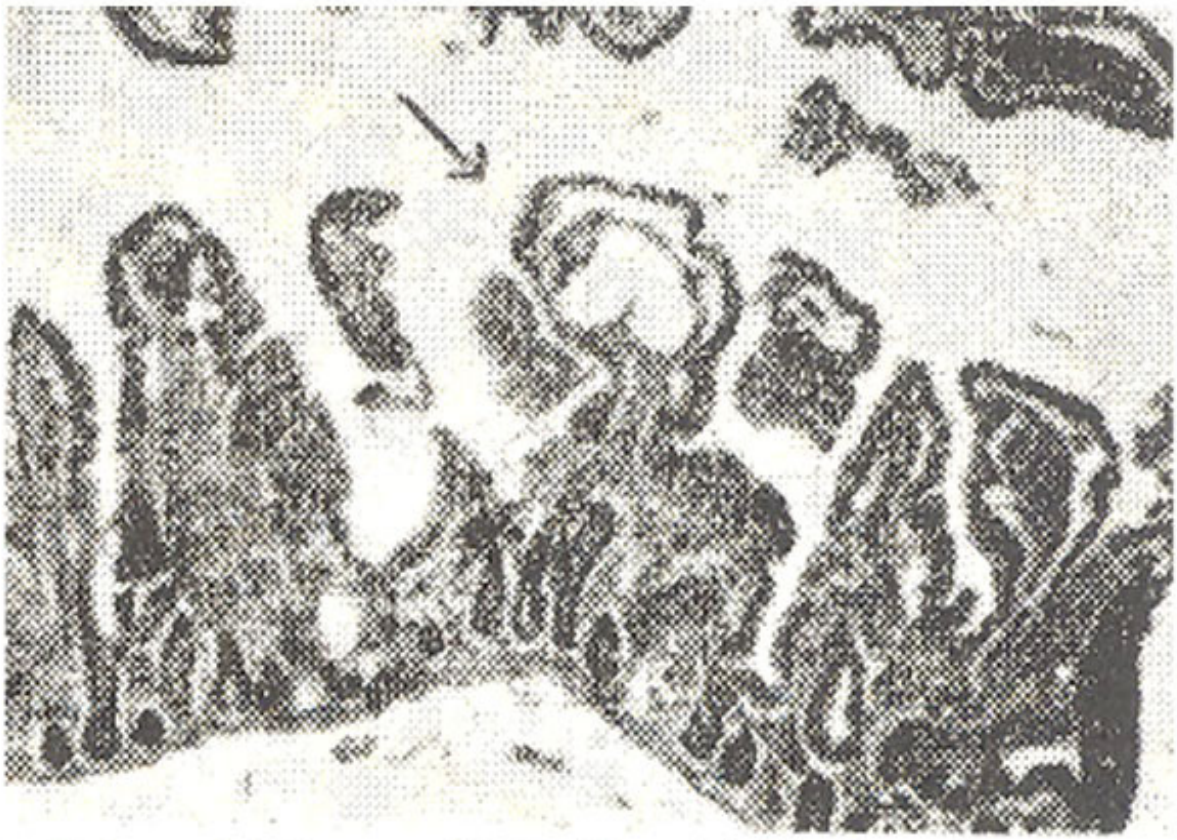

Fig. Jejunal biopsy (H & E x 100) showing dilated lymphatics in a villous.

She was given several infusions of saltfree albumin and her subsequent serum protein levels in August and September 1975 are shown in Table I.

Table I: Investigations

Date	Wt. lbs	Hb gm%	Total proteins gm%	Albumin gm%	Globulin gm%	Serum cholesterol mg%	Serum Calcium meq/lit
16 May'74	95	10.0	3.7	1.3	2.4	—	4.0
7 December'74	86	13.4	4.2	2.2	2.0	105	3.0
21 August'75	—	—	3.9	1.7	2.2	—	—
18 September'75	—	—	4.0	1.7	2.3	—	—
30 October'75	SURGERY						
15 November'75	88	11.9	4.4	2.9	1.5	155	4.2
30 December'75	90	—	5.4	3.4	2.0	184	5.1
24 March'76	—	12.0	6.1	3.55	2.65	—	—
17 November'79	100	12.3	6.4	3.6	2.8	130	—

On 30th October 1975, most of the affected jejunum was resected, leaving 2 feet of proximal jejunum. Histopathology report on the specimen also showed lymphangiectasia. Post-operatively, patient persisted to have frequent bowel movements ranging from 4 to 8 times per day, but by the end of December 1975 her total proteins rose to 5.4 gm% with albumin/globulin being 3.4/2.0. Serum cholesterol was 184 mg% and serum calcium 5.1 meg/litre. Only slight oedema over the shins was detected and she weighed 90 lbs. Serum proteins kept on improving gradually (Table 1) and on follow-up in March 1977, her appetite showed marked improvement and in August 1977 she weighed 99 lbs and showed no signs of oedema. She was subsequently advised to have medium chain triglycerides in her diet, and on follow-up examination in November 1979 she was having normal bowel movements, weighed 100 lbs and had normal protein levels (Table I).

Discussion

Loss of proteins into the gastrointestinal tract is commonly seen in a wide variety of gastro intestinal disorders. One of the causes of severe intestinal protein loss is intestinal lymphangiectasia. In this disorder there is marked dilatation of the lymphatic channels in the mucosa and serosa of the samll intestine. The lymphatic channels and mesentric lymph nodes frequently show fat filled macrophages (Pomerantz and Waldmann, 1963). There is no evidence of villous atrophy or infiltration of the intestinal wall with PAS positive material as seen in Whipple's disease (Holt, 1964).
The patient described had been keeping poor health with anorexia, and had been suffering from abdominal distention and diarrhoea since childhood. Sixteen patients suffering from intestinal lymphangiectasia, seen at the National Institutes of Health had a mean age of 10 years at the onset of symptoms (Waldmann et al., 1961). The majority of them also presented with mild diarrhoea or steatorrhoea at some time during the course of the disease (Table II).

Table II: Comparative Findings—Review of Literature

Authors	No. of Patients	Diarrhoea	OEDEMA		Effusions	Systemic lymphatic abnormalities	Hypoproteinaemia	Positive Radiology	Positive Histopathology
			Limited	Generalised					
Waldmann et al. (1961)	12	11		12	4		12	9	12
Pomerantz and Waldmann (1963)	4	1	2	2	1	4	4	4	4
Stoelinga et al. (1963)	1	1	1			Probably present	1	1	
Holt (1964)	1	1	—	1	—	1	1	1	1
Present Report	1	1	1	—			1	1	1

The hypoproteincmia seen in this syndrome, and as exhibited by our patient is due to the gastrointestinal loss of proteins. Albumin synthesis is not impaired but is normal or increased (Waldmann, 1961). Various theories regarding the mechanism for the increased loss of protein into the bowel in intestinal lymphangiectasia have been suggested. Dooren et al (1964) have suggested that protein may be taken up by foamy macrophages in the intestinal, wall, by demonstrating areas rich in these macrophages in one patient with exudative enteropathy. Stoelinga et al (1963) believe that rupture of the dilated intestinal lymphatics occurs, leading to leakage of lymph into the gut with considerable loss of protein and fat into the intestinal lumen. Sim-monds (1954) found that an increase in lymph flow resulted in the thoracic duct of rats on administering fats. In addition, protein concentration in the lymph was significantly higher following administration of fats. Lipids with long chain fatty acids were transported exclusively by the thoracic duct (Bloom et al., 1951), whereas fatty acids with a chain length of less than C12 pass into the portal vein (Borgstorm, 1955) and are not found in the thoracic duct (Fernandez, 1953). Holt (1964) therefore concluded that the absorption of lipids containing long chain fatty acids that are normally transported by the thoracic duct, enhances protein losses in intestinal lymphangiectasia, and that by feeding such a patient on medium chain triglycerides only, loss of albumin from the serum decreased markedly. In addition, the steatorrhoea and creatorrhoca present while consuming a normal diet was eliminated on taking a medium chain triglyceride formula. A similar improvement in protein loss together with reduction in the frequency of bowel movements, was also seen in our patient when she was subsequently placed on a medium chain triglycerides.

Barium meal examination of our patient showed coarse oedematous jejunal mucosa with some dilated loops. Of the 12 patients studied by Waldmann et al (1951), seven had a similar radiological picture, while 2 patients showed marked puddling and segmentation of the barium in the small intestine and 3 had negative roentgenograms (Table II).

It has been suggested that intestinal lymphangiectasia may be a manifestation of a more generalised lymphatic disorder (Holt, 1964). Lour of the patients reported by Waldmann et al (1961) had chylous effusions and three had asymmetrical oedema. Pomerantz and Waldmann (1963) studied 4 patients of intestinal lymphangiectasia and all of them had lymphangiographic evidence of systemic lymphatic abnormalities. Asymmetry in the oedema of a petient with idiopathic hypoproteineamia has also been reported by Stoelinga et al (1963). No such abnormality or any other evidence of systemic involvement was seen in our patient. Although ascites was noted previously" on physical examination, in May 1974 (Table II).

The pathogenesis of the lymphatic abnormalities seen in patients suffering from intestinal lymphangiectasia is not clear, though a congenital malformation seems to be the most likely explanation especially in those patients with onset at birth. In other patients one may attribute an acquired defect, such as blockage of the main lymphatic pathways, as a possible cause (Waldmann et al., 1961).

No definite treatment is available and none of the patients treated at the National Institutes of Health could be shown to respond satisfactorily to gluten restriction, steroids or resec- tion of the most severely a fleeted segment of the small intestine (Waldmann et al., 1961). Our patient was treated by resecting all but proximal 2 feet of the jejunum, and though postoperatively she continued to have frequent bowel movements, yet her serum protein levels showed a marked improvement as did her general condition, with disappearance of oedema and a markedly improved appetite. A subsequent introduction of medium chain triglyceride diet formula also benefitted the patient and the frequency of her bowel movements showed a significant reduction.

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References

1. Bloom, B.. Chaikoff, I.L., and Reinhardt, W.O. (1951) Intestinal lymph as pathway for transport of absorbed fatty acids of different chain lengths. Am. J. Physiol., 166:451.
2. Borgstrom, B. (1955) Transport of 14 C-decanoic acid in portal and inferior vena cava blood during absorption in the rat. Acta Physiol. Scand., 34:71.
3. Dooren, L.J., Van Gelderen, J.J., and Vink, C.L.J. (1961) Exudative enteropathy. Acta Paediat., 50:249.
4. Fernandez, J. Fat absorption studied in a child with chylothorax. Thesis. Leiden, 1953.
5. Holt, P.R. (1964) Dietary treatment of protein loss in intestinal lymphangiectasia. The effect of eliminating dietary long chain triglycerides on albumin metabolism in this condition. Pediatrics, 34:629.
6. Pomerantz, M. and Waldmann, T.A. (1963) Systemic lymphatic abnormalities associated with gastrointestinal protein loss secondary to intestinal lymphangiectasia. Gas troenterology, 45:703.
7. Simmonds, W.J. (1954) The effect of fluid, electrolyte and food intake and thoracic duct lymph flow in unanaesthetized rats. Austral. J. Exp. Biol. Med. Sci., 32:285.
8. Stoelinga, G.B.A., Van Munster, P.J.J. and Sloof, J.P. (1963) Chylous effusions into the intestine in a patient with protein loosing gastrocnteropathy. Pediatrics, 31:1011.
9. Waldmann, T.A., Steinfield, J.L., Dutcher, T.F., Davidson, J.D. and Gordon, R.S. Jr. (1961) Role of gastrointestinal system in "idiopathic hypoproteinemia". Gastroenterology, 41:197.