Intestinal lymphangiectasia: presentation in pregnancy and association with herpes zoster and alopecia

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We report a woman with intestinal lymphangiectasia whose symptoms were wrongly attributed to pregnancy; the diagnosis was made in the postpartum period. She also developed alopecia and herpes zoster. [Indian J Gastroenterol 1997; 16: 153-154]

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Intestinal lymphangiectasia (IL) is an uncommon disease which usually manifests with diarrhea and hypoproteinemia due to protein-losing enteropathy. The diagnosis is based on characteristic small bowel mucosal histology and demonstration of enteric protein loss. The onset of symptoms is usually in the first two decades of life although diagnosis may be delayed for several years. However, appearance of symptoms for the first time during pregnancy is uncommon. We report a woman in whom the disease manifested for the first time in pregnancy and was associated with alopecia and herpes zoster.

A 35-year-old woman presented in the immediate postpartum period of her fourth pregnancy. She had developed abdominal distension (which she felt to be exceptionally excessive as compared to her previous pregnancies) and pedal edema. However, she was not investigated for these symptoms. She delivered a full-term, low-birth-weight infant elsewhere; the baby died a day after birth. Abdominal distension and pedal edema persisted in the postpartum period. There was no history of seizure, decreased urine output, hematuria, facial puffiness, cough, alteration in bowel habits, abdominal pain, gastrointestinal bleeding or joint pains. She had noticed excessive loss of scalp hair since two months.

Examination revealed cachexia (body weight 25.3 kg), pedal edema, glossitis, angular stomatosis, moderate ascites and alopecia almost amounting to alopecia totalis (eyebrows and eyelashes preserved). There was no peripheral sign of chronic liver disease.

Investigations: Hemoglobin 7.8 g/dL; total leucocyte count 10,300/μL (polymorphs 95%, lymphocytes 5%, monocytes 2%); platelets 320,000/μL; ESR 6 mm in first hour; blood urea 37 mg/dL; serum creatinine 1.1 mg/dL; urinary protein 28 mg/24 h. Serum bilirubin 0.8 mg/dL (direct 0.12); proteins 5.3 g/dL (albumin 2.1); aspartate and alanine aminotransferases 47 U/L and 72 U/L respectively; alkaline phosphatase 336 U/L, prothrombin time 17.3 s (control 12.7). X-ray chest showed left pleural effusion. Abdominal ultrasonography was unremarkable except for ascites. Ascitic fluid analysis revealed 5 cells/mm³, protein 0.4 g/dL, glucose 100 mg/dL, smear and culture negative for tubercle bacilli. Pleural fluid showed protein 0.6 g/dL, glucose 96 mg/dL, 2 cells/mm³; fecal fat (72 h collection) 5.8 g/24 h, urinary d-xyllose 0.3 g/5 g/24 h.

In view of the lymphocytopenia, low serum protein and albumin, absence of splenomegaly or collaterals on ultrasonography and lack of significant proteinuria, a diagnosis of protein-losing enteropathy was considered. An isotope study using 99m Tc-labelled human serum albumin revealed a progressive collection of radio-pharmaceutical in the small bowel from 20 minutes onwards, moving distally with time, confirming enteric protein loss. 99m Tc labelled sulfur colloid scan showed normal hepatic uptake. Upper gastrointestinal endoscopy revealed white punctate spots in the second part of the duodenum. Biopsy from these areas revealed mild broadening of villi with dilated lymphatic channels in the lamina propria (Fig). Barium meal follow-through showed thickening of folds in the small bowel. Contrast-enhanced CT scan revealed ascites and thickening of small bowel folds without lymph node enlargement; liver and spleen were normal and no collateral vessel was seen at the splenic hilum.

With a diagnosis of primary IL the patient was treated with fat-restricted, high-protein diet with medium-chain triglyceride supplementation and low-dose diuretics. Her ascites and pedal edema subsided in one week. She remained ascites-free on follow up.

While in hospital she developed herpes zoster in the right lower thoracic nerve distribution. These lesions healed with scarring and depigmentation.

IL frequently affects young adults: therefore, pregnancy is not unexpected in these patients. However, occurrence of the symptoms for the first time during pregnancy, as in our patient, is uncommon. In such a situation, the diagnosis may be a challenge as many of the manifestations, like abdominal distension and pedal edema, occur in pregnancy. Unusual abdominal distension may even be wrongly attributed to hydramnios. In our patient, investigations were undertaken as abdominal distension and pedal...
edema persisted in the postpartum period.

The second interesting feature was the development of herpes zoster. To our knowledge, this is the first report of primary IL complicated by herpes zoster. Rarely, chicken pox and viral warts have been reported in IL, and are probably related to lymphocyte depletion which leads to depressed cell-mediated immunity, resulting in anergic skin response.4-5

Marked alopecia almost amounting to alopecia totalis has also not been described previously in this disease.

References

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BOOK REVIEW


This book is written from the perspective of developing countries. It is divided into seven sections, viz. chemical nature, physiology and metabolism of nutrients; assessment of nutrition status and nutrition requirements; functional significance of nutrition; nutritional deficiency disorders; diet, nutrition and degenerative diseases; food toxicities; and, reaching nutrients to the community. As is apparent, the book is loaded with information, and is well laid out. Most of the authors are past or present faculty of the National Institute of Nutrition, Hyderabad, and so have research and teaching experience in the field. But as a dietician, I am tempted to offer my comments on areas which I feel could have been written differently (my way!).

There could, for example, have been a chapter on biochemical investigations and their correlation with clinical states. In the chapter on nutritional requirements, energy requirements for adolescents should have been given on the basis of height.

The deficiency states of various nutrients have been covered; however, their toxic effects do not find mention, except for vitamin A. Though it is believed that water-soluble vitamins do not cause toxicity easily, this is not really true; a mention of this would have been useful.

The chapter on nutrition and immune response is well covered. In pregnancy, the energy requirement should be given as per the weight of the mother, and not as fixed requirements.

In protein-energy malnutrition, alternatives for milk-based diets, especially for lactose intolerant patients, would have been a valuable addition to a well-written chapter. The chapter on investigations for anemia is well written, as is the one on iodine. Nutrition in relation to thyroid requirements is dealt with very well.

Though the chapters on nutrition and systemic diseases are well written, most of the references are old, and recent advances in these disorders, many of which are relevant to management, have not been included. Wilson's disease and alcoholic liver disease have not been dealt with at all.

This is not to take away from the virtues of this great effort. Overall, the book is well written; it would have been even more readable if the font size used by the printer was a little easier on the eyes. But I suppose that is the price for information, which this book has in good measure. It should be a good adjuvant to any library: students interested in dietetics can easily afford this treat.

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