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CASE REPORT

Primary Hypogamaglobulinemia
A Case to Look for Failure to Thrive in Children

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Primary Hypogammagobulinaemia is a rare condition but it is one of the commonest causes of immune deficiency. It is usually non-familial but one of its variety is X-linked. Prevalance of the syndrome is about 15 per million for the male and 4 per million for the female population. The association of hypogammaglobulinemia and thymoma constitute a distinct syndrome. Secondary hypogammaglobulinaemia commonly occurs in chronic lymphatic leukemia and myeloma..(1 & 2).

CASE REPORT

A four and a half year old child was brought to the clinic with a history of failure to thrive. Her mother gave history of child having recurrent chest and gastrointestinal infections. He had been given prolonged courses of antibiotics and antiamoebic drugs. On examination he was a thin and frail child, weight 28 lbs., height 3 ft. 6 inches. He had slight pallor. His abdomen was distended bowel sounds were active. Chest was clear on auscultation. He had small lymph nodes paplable in the groin, and the anterior cervical chains. Lab data revealed Hb 12.5 grams. W.B.C.-9800/mm^3, D.L.C. -polys 34, Lymphos 57, Mono 4 and Eosinophils 5%. Peripheal smear was reported as normal. Bone marrow aspiriation examination and lymph node biopsy were also normal. Stool examination on three consecutive days revealed vegetative forms of Giardia lambia. Chest X-ray did not suggest a thymoma.

Serum immunoglobulins levels were IgM 42.7 mg% (N.V. 70-210 mg%), 1gG 275 mg% (N.V.700-1700 mg%) and 1gA 38.5 mg% (N.V. 70-350 mg%).

A request for barium meal examination was turned down by the mother.

A diagnosis of primary hypogammaglobulinaemia alongwith giardial infection was made. He was treated with immunoglobulins replacement therapy and metronidazole. He made a good recovery. He is now receiving his replacement therapy regularly(2,3,4). In the last three months he had gained 12 lbs of weight and had a single episode of chest infection.

COMMENTS

Primary hypogammaglobulinaemia although a rare disorder, should be kept in mind in children who are suffering from recurrent chest and G.I. Tract infections.

Primary hypogammaglobulinaemia is associated with frequent Giardiasis, Cryptosporodial infection and Campylobactor enteritis. It may also be associated with malabsorption syndrome which may be secondary to giardial infection or to bacterial overgrowth. Bacterial overgrowth is related to the hypo or achlorhydria commonly associated with hypogammaglobulinaemia. Achlorhydria and hypogammaglobulinaemia are assciated with a higher risk of cancer of stomach, fifty folds increase as compared to general population. In pernicious anaemia there is achlorohydia and increased risk of cancer of stomach is five folds).

Nodular lymphoid hyperplasia occurs in different parts of G.I. tract in adult onset hypogammaglobulinaemia Dental caries is less common in hypogammaglobulinae mia (1).

This case of primary hypogammaglobulinaemia is being reported to alert the general physicians that it should always be kept in the back of our minds because it is manageable disorder.

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