Selective IgM deficiency presenting as chronic diarrhea: A case report

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Abstract

Primary immunodeficiency (PID) is a heterogeneous disorder resulting from reduced or absent function of one of the components of immune system like T-cells, B-cells, NK cells, neutrophils, its receptor, immunoglobulins, complements and genes responsible for rearrangement of T-cell receptor and immunoglobulin and its signaling molecule. Among the clinical manifestation sinusitis, pneumonia, bronchiectasis, allergic disease, autoimmune disease and diarrhea are common manifestation in Antibodies deficiency.

A 4-years-old boy presented with chronic diarrhea and failure to grow. On endoscopy, there was nodule in D2 region which, on histopathological examination showed features mimicking as celiac disease. His serum anti-TTG Ab was within normal limits and serum Ig M was deficient.
Introduction
Primary immunodeficiency (PID) is a heterogeneous disorder resulting from reduced or absent function of one of the components of immune system like T-cells, B-cells, NK cells, neutrophils, its receptor, immunoglobulins, complements and genes responsible for rearrangement of T-cell receptor and immunoglobulin and its signaling molecule \[^{[1]}\] \[^{[2]}\]. PID registry in Australia have shown that antibody deficiency (Ab D) is the commonest form of immunodeficiency which comprises 77% of the total PID. Among the antibody deficiency, commonest is Common Variable Immunodeficiency (CVID – 38.4%) followed by IgG subclass Deficiency (19.4%), IgA deficiency (7.8%), X linked agammaglobulinemia (6.6%), specific antibody deficiency (3.0%) and others \[^{[3]}\]. Among the clinical manifestation sinusitis, pneumonia, bronchiectasis, allergic disease, autoimmune disease and diarrhea are common manifestation in Ab D.

Selective IgM deficiency is uncommon. Here we are reporting a rare case of selective IgM D who presented with chronic diarrhea and failure to thrive.

Case Report
A 4 years old male child presented to clinician with history of recurrent diarrhea for last 2 years along with failure to thrive. He had mild pallor with cachectic look. He was very lean and thin with weight of 6.5 kg and had monkey shaped face. There was no positive family history of similar manifestation. His urine examination and complete blood count were within normal limit except with mild anemia (Hb-7.8 g/dl). Clinician suspected him to be a case of Celiac disease. Endoscopic examination revealed presence of nodules in D2 region. Histopathology revealed atrophy of villi, increase in intraepithelial lymphocytes and plasma cells & eosinophils in lamina propria, all of which favouring towards celiac disease (fig. 1).

![Fig 1: Photomicrograph showing subtotal villous atrophy, increase in lymphocytic infiltration (>30/100 epithelial cells) and increase in eosinophils (H & E Stain X 200).](http://www.pacificejournals.com/aabs)
rum Ig E was increased (233.8 mg/dl; normal range <60IU/mL). Serum IgM was repeated after one month which was found to be again low (1.4 mg/dl). Patient was kept on antibiotics but he did not turn up for follow up.

**Discussion**

Failure to thrive, recurrent infections and diarrhea are the common manifestation of antibody deficiency [3,4]. Prevalence of gastrointestinal disorder is present in 5 to 50% patients of PID[5,6]. Diarrhea is more common in CVID and IgA D[7]. Selective IgM D presenting as diarrhea and failure of growth is uncommon.

In past Delacencha et al described 2 patients with IgM D who presented with dermatitis, chronic diarrhea, recurrent respiratory infections, failure to thrive and elevated serum IgE. Their B lymphocytes with surface IgM were within normal limit and in vitro B lymphocytes, when co cultured with normal T cells, produced normal IgM. They proposed that the IgM D was due to the defect in T regulatory cells. Our case is similar to that, because lamina propria contained plenty of plasma cells also probably there is some defect in T helper cell level. Presence of normal IgG and IgA cannot be explained. It may be due to either some auto antibody to IgM causing its catabolism, or due to T-helper cell defect which has produced defective isotype switching.

**Conclusion**

Our present study concludes that isolated serum IgM deficiency may present with recurrent diarrhea and failure to growth.

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**Competing Interests**

None declared.

**References**