In conclusion, HCV co-infection is common among HIV-infected persons in northern India. Further studies are needed to assess the impact of HCV co-infection on clinical outcome and response to treatment in these patients.

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Desquamative enterocolitis: an intestinal variant of Carmi syndrome presenting as protein-losing enteropathy

Carmi syndrome is a rare autosomal recessive disorder comprising congenital pyloric atresia (PA) and epidermolysis bullosa (EB). This association (EB–PA) was described by Swinborne and Kolher in 1968, whereas the genetic analysis was contributed by Carmi and hence the name.[1] The basic feature is a defect affecting the integrity of the basement membrane and hemidesmosomes and the control over the normal process of fibrosis which occurs during wound healing. It is associated with a high mortality and the majority of patients succumb to sepsis, dyselectrolytemia and hypoproteinemia. We report a 2-year-old boy with desquamative enterocolitis, an intestinal variant of this EB–PA syndrome presenting as protein-losing enteropathy (PLE).

A 2-year-old boy presented with diarrhea, generalized anasarca and poor appetite of 3 weeks' duration following the intake of uncooked meat. He was the second child of consanguineous parents and his mother had polyhydramnios during the antenatal period. He was operated for pyloric atresia on post-natal day 2. Small bullous lesions, which appeared in crops and healed with topical medications, were noticed on his forehead, chest and legs since the newborn period. He was an apathetic, undernourished child with anasarca, weight and height were less than third percentile. The liver was palpable 3 cm below the costal margin. Few healed hypopigmented macular lesions were seen over the face, trunk and extremities. During hospitalization, he passed strips of tissue, the longest was 2.5×1.0×0.5 cm, tubular, with a smooth, reddish external surface, resembling intestinal casts (Figure). He also developed fresh bullous lesions on his chest and ears.

Investigations: Total protein was 3.3 g/dL with albumin level of 2.2 g/dL. Immunoglobulin profile was normal and HIV status was negative. Barium studies showed malabsorptive pattern. Endoscopy showed that the prepyloric area appeared friable with a retained suture. The duodenal mucosa showed mild scalloping. Histology of duodenal biopsy showed villous fragments with no increase in intraepithelial lymphocytes or villous atrophy. Colonoscopy was normal and biopsy was reported as subacute colitis. Histology of the skin lesions confirmed epidermolysis bullosa, and that of the intestinal casts showed focal ghost outlines of the intestinal wall with epithelial cells in clusters.

The patient was managed with intravenous fluids, albumin, antibiotics, parenteral nutrition and oral phenytoin for skin lesions. Four months later he succumbed to sepsis. A diagnosis of Carmi syndrome was suggested

Figure: Showing intestinal cast

with "desquamative enterocolitis" presenting as PLE.

Epidermolysis bullosa and congenital pyloric atresia are both rare autosomal recessive conditions occurring in 1 in 100,000 and 1 in 300,000 live-births, respectively.[2] Congenital PA can occur in isolation, with EB or with intestinal atresia. Similarly, EB can be simplex, dystrophic or junctional (JEB). All three types of EB have been reported with PA, the most common being JEB.[3] The occurrence of EB–PA or Carmi syndrome in children is rare. Two cases of desquamative enterocolitis, an intestinal variant of this syndrome where the child passes visible strips of intestinal tissue per rectum have been reported previously.[4] Pyloric atresia in EB as proposed by Chang[5] is due to the separation of the epithelium from its basement membrane in the developing pylorus leading to sloughing, scarring and fibrosis with narrowing of the pyloric canal.

There is no definite treatment for EB though phenytoin and steroids have been tried. Children with EB–PA syndrome and presenting with passage of intestinal tissue have been reported and designated as an overlap syndrome. The episodic nature of the relapses may be secondary to an autoimmune response and deposition of IgG and C1q in the basement membrane. In this child the event was probably triggered following a gut infection; however immunofluroscence of colonic mucosa was not done. Since this disease can affect any epithelium, children may present with features of obstructive uropathy and should be screened for renal disease even if asymptomatic.

The basic inherited defect is that the integrity of the basement membrane and hemidesmosomes, and control over the normal process of fibrosis which occurs during wound healing are disturbed. PA therefore occurs as an intrauterine complication of EB.[2] EB–PA is caused by the ITGA-6 or ITGB-4 genes encoding for the integrin alpha-6 or beta-4 subunits, respectively. Mutation genes encoding the epithelial integrin a6b4 (ITGA-6 and ITGB-4) have been identified in patients with Carmi syndrome.[6] Prenatal diagnosis is possible by fetal DNA analysis and fetal skin biopsy using EM and indirect immunofluorescence. Ultrasound features of gastric dilatation due to PA, and the snow flake sign of amniotic fluid in the second trimester in EB, if present, are characteristic. This combination of EB–PA can therefore present with failure to thrive, PLE, respiratory compromise, obstructive uropathy, electrolyte imbalance and septicemia. In this child it was associated with an intestinal component, namely desquamative enterocolitis presenting as hypoproteinemia, dehydration, dyselectrolytemia and sepsis.

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