Jodhpur disease revisited: a rare cause of severe protein energy malnutrition

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Abstract
A 3.5-year-old grossly cachectic female child presenting with recurrent vomiting, fever, abdominal distention, abdominal pain and severe weight loss was evaluated for the cause of severe protein energy malnutrition. Investigation revealed a massively dilated stomach with delayed gastric emptying and normal pylorus. On exploratory laparotomy, diagnosis of primary acquired gastric outlet obstruction (Jodhpur disease) was confirmed and she underwent pyloroplasty with uneventful post-operative period. To conclude, this entity should always be included in the differential diagnosis of gastric outlet obstruction with severe malnutrition especially in older children.

Keywords
Jodhpur disease, protein energy malnutrition, gastric outlet obstruction

Introduction
Protein energy malnutrition is rampant worldwide especially in developing countries. Most cases are due to poor nutrition and infections. Rarely, it may be due to structural gastrointestinal abnormalities. Jodhpur disease is one such rare structural abnormality of unknown etiology causing severe malnutrition. We present a case of this potentially curable entity, highlighting the importance of proper work up of all cases of malnutrition.

Case report
A 3.5-year-old unimmunized developmentally normal female child presented with vomiting for 6 months, intermittent fever for 5 months, abdominal distention for 4 months and painful abdomen for 1 month. Vomiting was non bilious, non projectile, 4-5 episodes/day, occurring 1-2 h after meal and contained food particles. Fever was on and off, low-grade, not associated with chills and rigor. Pain was localized to upper abdomen, non colicky, aggravated by food intake, relieved by vomiting or medications. The child had significant weight loss (>5 kg) during the past 4 months and had received one unit packed red cell transfusion at some other center. There was no history of loose stools at any point of time, constipation, jaundice, bleeding from any site or contact with a tuberculosis patient. On examination, she was malnourished with her weight only 6.5 kg and height 84 cm, both below the 3rd percentile of WHO standards. She was pale and dehydrated, with sparse and lusterless hair. Abdomen was soft and distended, with tympanic note on percussion, normal bowel sounds and without organomegaly. The rest of the systemic examination did not show any abnormality.

On investigation, her hemogram and chest X-ray were normal, but serum electrolytes were abnormal (hyponatremia with hypokalemia). HIV serology, PPD test and gastric aspirates (n=3) for tuberculosis were negative. Stool examination was negative for ova, cysts or opportunistic pathogens. X-ray abdomen showed no air-fluid level but only gaseous distention of stomach and intestine. Ultrasound of abdomen showed distended stomach with normal pyloric canal. Barium meal showed grossly distended stomach with slow passage of barium from the antero-pyloric region distally (Fig. 1). Suspecting gastric outlet obstruction, exploratory laparotomy was done that revealed hugely distended stomach with normal smooth muscle thickness but no intra-luminal valve, ring or diaphragm or extra-luminal compression was seen (Fig. 2). Heineke-Mikulicz pyloroplasty was done. Pyloric biopsy showed sub-mucosal edema and mild congestion and no inflammatory cells were seen. The diagnosis of primary acquired gastric outlet obstruction during infancy and childhood (Jodhpur disease) with severe malnutrition was made. The child recovered well postoperatively, became asymptomatic gradually and gained 2.5 kg in one month post surgery.
after the place of its first description [1]. Recently, it has been renamed “primary acquired GOO during infancy and childhood”. Until now, 22 cases have been reported in the world literature, among which 14 are from India (13 from Jodhpur itself) [2-8]. It is a rare entity with an incidence of 1 in 100,000 live births, with predilection for male sex [2]. Mean age of presentation is 2.9 years (range 1 month–6 years) [2].

The exact etiology is still unknown. Various hypotheses have been put forward. One states that lack of nitric oxide synthase in neurons leads to failure to relax pyloric smooth muscle causing neuromuscular incoordination and later GOO, while another claims that the cause is an abnormality of the interstitial cells of Cajal that act as electrical pacemakers of GI tract smooth muscles [9].

Usual presentations include intractable and recurrent non-bilious projectile vomiting, weight loss, pain abdomen, abdominal distention and growth retardation. Diagnosis is aided by ultrasonography that shows normal pyloric canal with no pyloric muscle hypertrophy, while upper GI contrast study shows large stomach with increased gastric emptying time (Fig. 1). Gastroscopy reveals no intra-luminal pathology with normal gastric mucosal rugosities in a large-size stomach with narrow pyloric canal. CT scan is usually of no further help as it shows only typical picture of GOO with narrow pyloric canal. Histopathologically there is normal cellular pattern of all coats without inflammatory, fibro-proliferative or neoplastic cells with a normal number of ganglion cells [2]. Management is essentially surgical with “Heineke-Mikulicz pyloroplasty” as the procedure of choice with excellent outcome [2]. Pneumatic dilatation by balloon inflation via upper endoscopy has also been tried successfully [10].

In conclusion, clinical presentation mimicking idiopathic hypertrophic pyloric stenosis in older infants and children should be strongly considered for Jodhpur disease.

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