

Rapunzel Syndrome An Uncommon Problem In Children

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Citation

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Abstract

A Trichobezoar represents a mass of accumulated hair within the gastrointestinal tract. Isolated gastric Trichobezoars, those with extension into the duodenum, and small intestinal Trichobezoars have all been described (1). We present two cases of Trichobezoars; 7 1/2 and 11 years old girls with palpable abdominal mass. The diagnosis was clinically suspected, confirmed by investigation and through gastrotomy retrieved. Both patients went uneventful recovery. These cases are presented because of its rarity, unique presentation, and simplicity of management.

INTRODUCTION

The Rapunzel syndrome, first described by Vaughn and coauthors in 1968, is a rare form of gastric trichobezoar with a tail-like extension into the duodenum or proximal small intestine. (2) Up to 2003 only 14 cases of this entity have been reported. (3)

CASES PRESENTATION

We report two cases; The 1st case a 7 1/2 year's old girl was admitted in oncology department as a case abdominal mass with loss of appetite for last week, marked wasting, BW 15 Kg. and some loss in scalp hair; Abdomen shows epigastric mass oblong in shape occupying upper abdomen, firm in consistency, smooth surface mobile up and down. The 2nd case is 11-year old girl, complaining of intermittent colicky abdominal pain since one year, increased in severity last one week; with no evidence of psychological problems related to family affairs and parental marital problems. The patient was generally stable; no alopecia there is hard non-tender epigastric intra abdominal mass. In both cases abdominal X-rays (fig 1) Shows the gastric shadow occupied by radiopaque like mass with absent gastric bubble. Barium meal (fig 2)

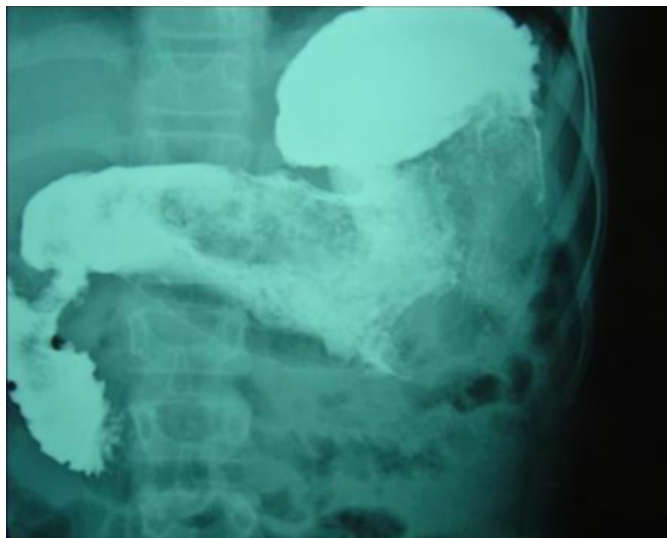
Figure 1

Figure 1: Standing AP radiograph showing Trichobezoars opacity occupying most of the gastric shadow.



Figure 2

Figure 2: shows two large filling defects in the stomach



And C.T scan (fig 3) Showed large intragastric mass which extend into duodenum. Gastroscopy was performed in case 2 (fig 4) revealed a large ball of hair occupying most of the stomach which could not be removed endoscopically

Figure 3

Figure 3: abdominal CT scan (with IV & oral contrast) shows large heterogeneous intragastric mass surrounded with contrast & extended to the duodenum

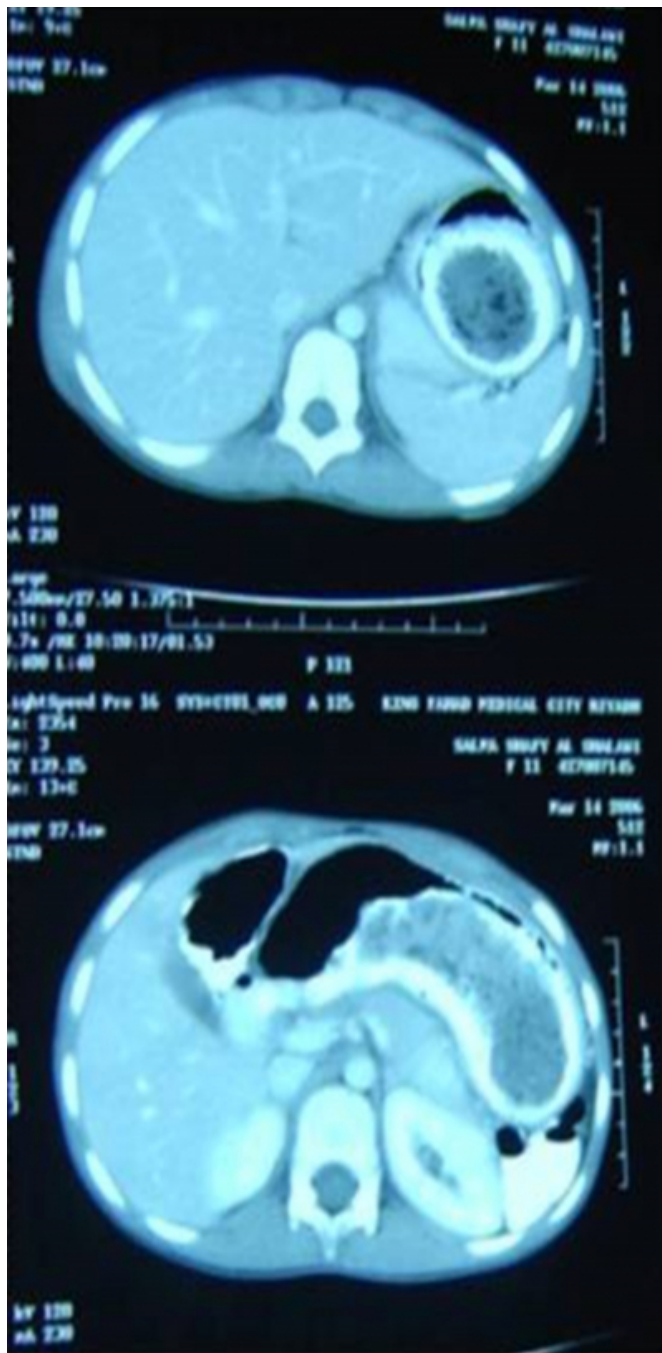
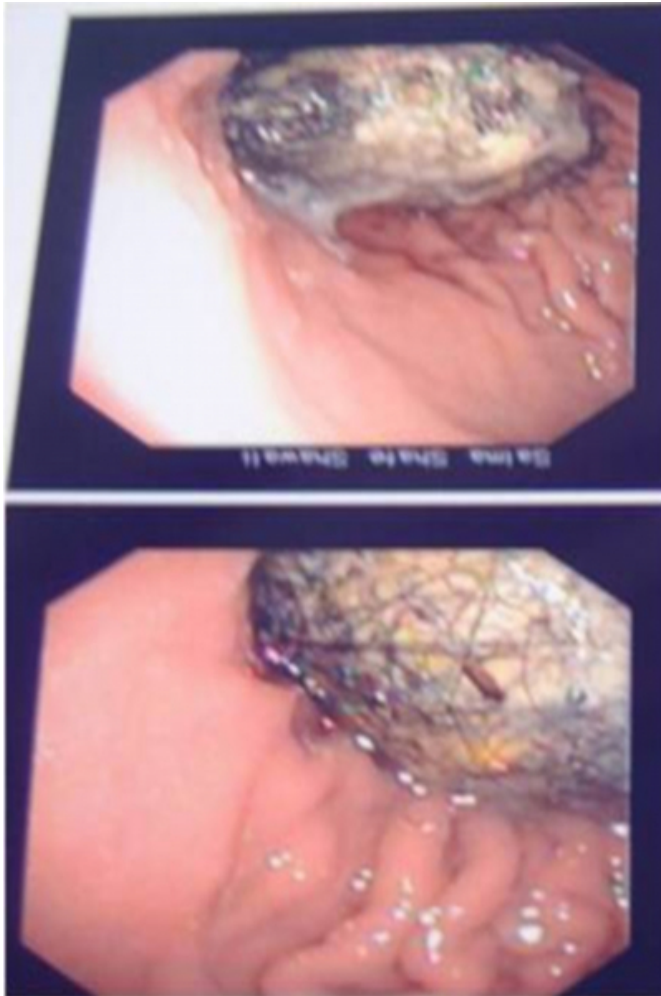


Figure 4

Figure 4: Showed a large, greenish black Trichobezoars occupying most of the stomach



In both cases the Trichobezoars was removed by gastrotomy (fig 5), which revealed; around 20x15cm J shaped, foul smelling, black loose Trichobezoars, weighting 1.540 grams in the 1st case and 2,152 grams in 2nd case; with extension into the duodenum and jejunum (Fig 6) .

Figure 5

Figure 5: At Laparotomy a large 20x15 cm j shaped Trichobezoars black in color was retrieved via a longitudinal anterior gastrotomy and you can see extension of the bezoars into the duodenum and jejunum.



Figure 6

Figure 6: shows Postoperative specimen of Trichobezoar



The patients made uneventful recovery, discharged home at 3rd postoperative day and referred to the pediatric behavioral clinic for psychological evaluation and support.

DISCUSSION

The Rapunzel syndrome (a trichobezoar with a long tail extending from the stomach to the small bowel) is an uncommon disease. (4) Which can cause small bowel obstruction, jaundice or even acute pancreatitis (5); our cases were lucky that presented before occurrence of this complication.

DeBakey and Ochsner reviewed 172 cases of trichobezoar of which 90% were females in the 10 to 19 years age group and they noted that a palpable abdominal mass was present in

(87.7%). (6) Like in Our two cases were females (7 ½ and 11 years old), presented mainly with abdominal mass.

Diagnosis may be aided by abdominal plain films, contrast upper gastrointestinal radiography, ultrasound, CT scan, or upper endoscopy . (5) As in our cases the diagnosis was Clinical suspected, confirmed radiological but in the 2nd case we used the endoscopy only as a trial for non invasive removal of trichobezoar.

Larger gastric trichobezoars must be removed either laparoscopically or by laparotomy (1). Because the treatment of choice for trichobezoar is Surgical (6) we decide to retrieve the huge trichobezoars through open gastrotomy.

Most importantly, although surgery addresses the immediate issue, psychiatric follow-up is essential to prevent recurrences (1); we referred our cases to the pediatric behavioral clinic for psychological evaluation and support.

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