

Pseudo-Bartter Syndrome as an Atypical Presentation of Intestinal Malrotation: A Case Report

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Abstract

Intestinal malrotation is a congenital intestinal rotation anomaly and can present with various symptoms. Electrolyte disorders are very common in childhood. Pseudo-Bartter syndrome (PBS) is one of the conditions that causes electrolyte disorders and can be seen due to intestinal malrotation in children.

A 3.5-month-old boy who was diagnosed as having malrotation is reported. The patient had PBS because of non-bilious vomiting.

We could find only two reports on PBS related to malrotation. It is emphasized that intestinal malrotation should be considered in patients presenting with gastrointestinal symptoms such as vomiting, abdominal pain, and also PBS.

Introduction

Electrolyte disorders are one of the most common problems encountered by pediatric physicians. Vomiting is one of the most common causes of electrolyte disorders during infancy. There are many reasons for vomiting in infants such as gastroesophageal reflux, food poisoning, post-tussive emesis, overfeeding, feed intolerance, otitis media, pneumonia, urinary tract infection, renal tubular acidosis, obstruction, intussusception, malrotation, milk allergy, hydrocephaly, raised intracranial pressure, and acute gastroenteritis [1]. Although most infants and children with vomiting have a non-serious etiology, it may sometimes be the initial symptom in life-threatening conditions [1]. Bartter syndrome (BS) is a clinical condition characterized by hypokalemic metabolic alkalosis, hyperreninemia and hyperaldosteronism, normal blood pressure, and hyperplasia of the juxtaglomerular apparatus, first described by Frederic Bartter in 1937 [2]. Pseudo-Bartter's syndrome (PBS) is a condition that may be caused by diuretic or laxative abuse, prolonged gastric drainage without adequate electrolyte support, diarrhea-vomiting, pyloric stenosis, and cystic fibrosis, but without primary renal tubule abnormalities [3]. Electrolyte imbalance, especially after severe vomiting, can be helpful for the diagnosis. Herein, we report an infant who presented with severe vomiting, dehydration, and hypokalemic hypochloremic metabolic alkalosis who was diagnosed as having PBS due to malrotation.

Case Presentation

A 3.5-month-old infant boy was admitted to a local hospital because of non-bilious vomiting, which he had had for 15 days. The vomiting frequency was 10 to 12 times per day. On admission, dehydration was marked, intravenous physiologic saline was initiated, and he was referred to our hospital for further evaluation. His past medical history was unremarkable, as was the family history.

At admission to our hospital, he had two generalized tonic clonic seizures. The patient was dehydrated, lethargic, and pallor. He had fever (36.5°C), his heart rate was 138/min, blood pressure was 118/78 mm Hg, and oxygen saturation was 94% in room air. The capillary refilling time was 3 seconds. His blood glucose was 88 mg/dL.

Hemoglobin 9.5 g/dL, white blood cells 23,660/ μ L, platelets 390,000/ μ L, sodium 133 mEq/L (normal: 135–145), potassium 2.5 mEq/L (normal: 3.6–5.8), chloride 89 mEq/L (normal: 98–118), calcium 5.8 mEq/L (normal: 8.8–10.88), blood urea nitrogen 44 mg/dL (normal: 0–10), blood glucose 97 mg/dL, C-reactive protein 3.4 mg/dL (normal: 0–4), and albumin, creatinine, aspartate aminotransferase, alanine aminotransferase, and gamma glutamyl transferase were normal. The blood pH was 7.58 (normal: 7.35–7.45), pCO₂ was 34.7 mm Hg (normal: 32–46), and HCO₃ was 33.3 mmol/L (normal: 18–23). Urine electrolyte values were in the normal range. The blood culture was sterile. The patient was diagnosed as having PBS because of metabolic alkalosis and electrolyte imbalance. Corrective fluid and electrolyte supplementation were administered. Abdominal ultrasonography (USG) was normal. Abdominal Doppler USG showed that the superior mesenteric artery was vertically placed to the inferior of the superior mesenteric vein and the patient was suspected of having intestinal malrotation. Barium contrast medium did not pass to the left side, and intestinal malrotation was diagnosed under esophagogastroduodenography (Fig. 1). The patient was referred to the pediatric surgery department. Surgery was performed for the intestinal malrotation. After a successful Ladd procedure, the symptoms completely resolved. He was discharged from hospital with no further vomiting episodes.

Discussion

Intestinal malrotation is a disorder resulting from the lack of intestinal physiologic rotation during organogenesis in embryonic life [4]. Most malrotation cases present within the first month of life. More than 40% of cases are diagnosed within the first week of life and 75–85% within the first year of life [5]. Although the symptoms in newborn infants are those of intestinal obstruction, such as biliary vomiting, almost 50% of cases initially present with non-bilious vomiting [6].

Clinical conditions that cause the biochemical findings of BS without pathology of kidneys are defined as PBS [3]. PBS can be caused by a severe chloride deficiency secondary to vomiting [7]. The renin-angiotensin-aldosterone system is activated due hypovolemia that occurs after severe vomiting, then the reabsorption of sodium and excretion of potassium is increased [7]. Electrolyte abnormality and dehydration due to PBS may be a critical condition such as convulsions that was seen in this report. As in our case, PBS due to malrotation is less documented [8]. In our PubMed search, we could only find two clinical reports of PBS due to malrotation [8,9].

Conclusion

Pediatricians should consider the diagnosis of malrotation in the presence of PBS and look carefully for clinical signs of the disease.

Abbreviations

BS: Bartter syndrome

PBS: pseudo-Bartter's syndrome

USG: ultrasonography

Declarations

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Ethics approval and consent to participate: No ethical committee approval is required for this case report.

Consent for publication: Consent was obtained from parents.

Availability of data and material (data transparency): Data sharing is not applicable to this article because no datasets were generated or analyzed during the current study.

Code availability: Not available.

Authors' contributions: OA drafted the initial manuscript. OA, BA, and SK contributed to the patient management. SK critically reviewed the manuscript. All authors have read and approved the final submitted manuscript.

References

1. Singhi SC, Shah R, Bansal A, et al. Management of a child with vomiting. *Indian J Pediatr.* 2013;80(4):318-325.
2. Bartter FC, Pronove P, Gill JR, et al. Hyperplasia of the juxtaglomerular complex with hyperaldosteronism and hypokalemic alkalosis. A new syndrome. *Am J Med.* 1962;(33):811-828.
3. Saneian H, Bahraminia E. Congenital chloride diarrhea misdiagnosed as pseudo-Bartter syndrome. *J Res Med Sci.* 2013;18(9):822-824.
4. Aslanabadi S, Ghaleh Golab-Behbahan A, et al. Intestinal malrotations: a review and report of thirty cases. *Folia Morphol.* 2007;66(4):277-282.
5. Ayane GN, Kadimo K. Diagnosis and surgical management of congenital intestinal malrotation presenting with midgut volvulus in an adult: high index of suspicion (case report). *Pan Afr Med J.* 2018; (29):154.
6. Millar AJ, Rode H, Cywes S. Malrotation and volvulus in infancy and childhood. *Semin Pediatr Surg.* 2003;12(4):229-236.

7. Kintu B, Brightwell A. Episodic seasonal Pseudo-Bartter syndrome in cystic fibrosis. *Paediatr Respir Rev.* 2014;15 Suppl 1:19-21.
8. Koshida R, Sakazume S, Maruyama H, et al. A case of pseudo-Bartter's syndrome due to intestinal malrotation. *Acta Paediatr Jpn.* 1994;36(1):107-111.
9. Gonzalez-Rivero MA, Bonet Alcaina M, Vall Combelles O, et al. [Pseudo-Bartter syndrome as a complication of an undiagnosed intestinal malrotation]. *An Esp Pediatr.* 1998;49(5):523-524.

Figures



Figure 1

Esophagogastroduodenography demonstrating the intestinal malrotation

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