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

Orkun Aydin1 · Burak Ardicli2 · Selman Kesici3

Accepted: 10 December 2021 / Published online: 27 December 2021 © The Author(s), under exclusive licence to Springer Nature Switzerland AG 2021

Abstract
Intestinal malrotation is a congenital intestinal rotation anomaly and can present with various symptoms. Pseudo-Bartter syndrome (PBS), which mimics the manifestations of Bartter’s syndrome, can be caused by a severe chloride deficiency and can be seen due to cystic fibrosis, hypertrophic pyloric stenosis, diuretic abuse, and so on. We presented a 3.5-month-old boy who had convulsion after multiple vomiting, followed up in the intensive care unit, and was diagnosed with malrotation. The patient complained of vomiting 10 times and diarrhea 3–4 times a day in the last 15 days. He went to another urban hospital after having a tonic–clonic seizure. Hypochloromic hypokalemic metabolic alkalosis was detected and he was admitted to the intensive care unit of our hospital. He was examined for differential diagnosis. The intestinal malrotation was diagnosed with esophagogastroduodenography. After surgery, he made a good recovery and was discharged symptom free. Cystic fibrosis, Barter syndrome, and hypertrophic pyloric stenosis are known diagnoses in patients presenting with PBS. Only two patients diagnosed with malrotation have been reported in the literature. We wanted to present this case so that the diagnosis of malrotation should be considered in patients presenting with PBS.

Keywords Electrolyte disorders · Intestinal malrotation · Pseudo-Bartter syndrome

Introduction
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 1962 [1]. After that, several articles have reported mimicking manifestations of Bartter syndrome due to diuretic or laxative abuse, prolonged gastric drainage without adequate electrolyte support, diarrhea-vomiting, pyloric stenosis, and cystic fibrosis, but without primary renal tubule abnormalities [2–4]. Then, this hypokalemic hypochloremic metabolic alkalosis picture turned into a laboratory picture called pseudo-Bartter syndrome.

Herein, we report an infant who presented with severe vomiting, diarrhea, and convulsion. Hypokalemic hypochloremic metabolic alkalosis was detected in the patient. With these laboratory findings, the patient was diagnosed with PBS and was examined for differential diagnosis. He was diagnosed with malrotation after the tests were performed. We wanted to present this rare case of the pseudo-Bartter syndrome, diagnosed with malrotation, who presented with seizures.

Case Presentation
A 3.5-month-old infant boy was presented to a local hospital for having had a seizure. The patient has complaints of vomiting and diarrhea 10 times a day for the last 15 days. Physical examination revealed dehydration and lethargy. Biochemical investigations documented electrolyte abnormalities (Na: 126 mEq/L, K: 2.4 mEq/L, creatinine: 1.7 mg/dL). Intravenous physiologic saline was performed due to hyponatremia to correct dehydration and electrolyte...
abnormalities, and he was referred to our hospital for further evaluation.

At admission to our hospital, he had two other generalized tonic–clonic seizures. The patient was dehydrated, lethargic, and pallor. Body temperature was 36.5 °C, 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 s. His blood glucose was 88 mg/dL. His past medical history was unremarkable, as was the family history. The patient did not have a history of the previous admission to the hospital with vomiting and diarrhea. He defecated on the first day after he was born. He had no complaints until 15 days ago. His weight was 5.5 kg (10 to 25th percentile for age).

Laboratory studies was performed and hemoglobin 9.5 g/dL, white blood cells 23,660/μL, platelets 390,000/μL, 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 sodium < 20 mEq/L, potassium 23.1 mEq/L, chlorine 22 mEq/L, and calcium < 2.0 mEq/L. All urine electrolyte values were normal. The blood culture was sterile. A sweat test was wanted to perform, but it was not successful. The patient was diagnosed as having PBS because of metabolic alkalosis and electrolyte imbalance. A plain abdominal radiograph taken after the seizure was under control. Abnormal distribution of intestinal gas was seen (Fig. 1). Abdominal ultrasonography which was performed both because of abnormal distribution on the graph and for differential diagnosis of PBS showed that the superior mesenteric artery was vertically placed to the inferior of the superior mesenteric vein. As a consequence, the patient was suspected of having intestinal malrotation and an esophagogastroduodenography was examined. Barium contrast enema did not pass to the left side (Fig. 2).

The patient was referred to the pediatric surgery department. Surgery was performed for intestinal malrotation. After a successful Ladd’s procedure, the symptoms completely resolved. He was discharged from the hospital with no further vomiting episodes.

**Discussion and Conclusions**

Clinical conditions that cause the biochemical findings of BS without pathology of kidneys are defined as PBS [5]. Cases reported in the literature with PBS were mostly diagnosed with cystic fibrosis, hypertrophic pyloric stenosis, excessive diuretic use, and a small number of Hirschsprung disease [6]. As in our case, PBS due to malrotation is less documented. In our research, we could only find two clinical reports of PBS due to malrotation [7, 8]. Basically, the mechanism can be considered as the development of metabolic alkalosis instead of metabolic acidosis due to vomiting from the proximal of the ligament of Treitz. However, malrotation is a diagnosis that can be missed in infancy and, like cystic fibrosis and hypertrophic pyloric stenosis, should be considered in patients with PBS. Koshida et al. presented a 6-year-old patient who was admitted to the hospital because of vomiting for the third time [9]. The patient who was diagnosed with PBS and later diagnosed with malrotation is the first case in this regard. Subsequently, Gonzalez-Rivero et al. reported a patient with PBS due to malrotation [10]. As far as we know, our patient is the first patient diagnosed with PBS during infancy and diagnosed with malrotation.

Intestinal malrotation is a disorder resulting from the lack of normal intestinal physiologic rotation during organogenesis in embryonic life [11]. Most malrotation cases are present within the first month of life. The incidence of malrotation is approximately at 1 in 500 live births [12]. More than 40% of cases are diagnosed within the first week of life and 75–85% within the first year of life [13]. 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 [14].
Laboratory changes that are mainly emphasized in pseudo-Bartter syndrome are hypokalemia, hypochloremia, and metabolic alkalosis. The pathogenesis of this state might be explained as follows: originally, the chloride and water loss occurred as a result of severe massive vomiting, and then, hypochloremia and hypovolemia were conducted to stimulation of the juxtaglomerular apparatus and activation of the renin–angiotensin–aldosterone system. Aldosterone would then develop reabsorption of sodium and excretion of potassium into the urine in the distal renal tubules. Serum potassium reduction boosts the synthesis of renal prostaglandin E2 [15], which then stimulates renin secretion, exacerbating the hyperaldosteronism [16]. In addition to hypokalemia and hypochloremia, hypocalcemia can also be seen in PBS and may cause seizures, as in our case. The reason for this can be explained as follows: because of intracellular hypokalemia, hydrogen and sodium ions move from the extracellular to the intracellular space. This replacement of intracellular protons (actually movement of hydrogen ions) induces a decrease in serum calcium ion concentration.

Although vomiting is common in infancy, the diagnosis of malrotation causing PBS in a patient presenting with severe vomiting and diarrhea is a rare condition. This patient had a seizure due to hypocalcemia. Then, when the whole picture was evaluated, a diagnosis of PBS was made. While investigating the etiology of PBS, he was diagnosed with malrotation. So 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 syndrome; USG: Ultrasonography

**Author Contribution**  
O.A. drafted the initial manuscript. O.A. retrieved the pertinent literature. O.A. and S.K. contributed to patient management. S.K. critically reviewed the manuscript. All authors have read and approved the final submitted manuscript.

**Data Availability**  
Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

**Declarations**

**Ethical Approval and Consent to participate**  
No ethical committee approval is required for this case report.

**Consent for Publication**  
Written informed consent was obtained from the patient’s legal guardian(s) for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.
Competing Interests  The authors declare no competing interests.

References

1. Bartter FC, Pronove P, Gill Jr JR, MacCardle RC. Hyperplasia of the juxtaglomerular complex with hyperaldosteronism and hypokalemic alkalosis: a new syndrome. Am J Med. 1962;33(6):811–28.
2. Saneian H, Bahraminia E. Congenital chloride diarrhea misdiagnosed as pseudo-Bartter syndrome. J Res Med Sci. 2013;18(9):822–4.
3. Faraji-Goodarzi M. Pseudo-Bartter syndrome in children with cystic fibrosis. Clin Case Rep. 2019;7(6):1123–6.
4. Yu H-R, Huang S-C, Hsieh C-S. Infantile hypertrophic pyloric stenosis presenting as pseudo-Bartter’s syndrome and seizures: report of one case. Zhonghua Minguo Xiao Er Ke Yi Xue Hui Za. 1998;39(3):195–7.
5. Vanhaesebrouck S, Allegaert K, Vanhole C, Devlieger H, Gewillig M, Proesmans W. Pseudo-Bartter syndrome in a neonate on prostaglandin infusion. Eur J Pediatr. 2003;162(9):569–71.
6. Vanhaesebrouck S, Van Laere D, Fryns JP, Theyskens C. Pseudo-Bartter syndrome due to Hirschsprung disease in a neonate with an extra ring chromosome 8. Am J Med Genet A. 2007;143(20):2469–72.
7. Koshida R, Sakazume S, Maruyama H, Okuda N, Ohama K, Asano S. A case of pseudo-Bartter’s syndrome due to intestinal malrotation. Acta Paediatr Jpn. 1994;36(1):107–11.
8. Gonzalez-Rivero MA, Bonet Alcaina M, Vall Combelles O, Cesena Santiago Y, Martinez-Roig A, Garcia AO. Pseudo-Bartter syndrome as a complication of an undiagnosed intestinal malrotation. An Esp Pediatr. 1998;49(5):523–4.
9. Koshida R, Sakazume S, Maruyama H, Okuda N, Ohama K, Asano S. A case of pseudo-Bartter’s syndrome due to intestinal malrotation. Pediatr Int. 1994;36(1):107–11.
10. González-Rivero M, Alcaina MB, Combelles OV, Santiago YC, Martínez-Roig A, Algar OG. Pseudo-Bartter syndrome as a complication of an undiagnosed intestinal malrotation. An Esp Pediatr. 1998;49(5):523–4.
11. Aslanabadi S, Ghalehgolab-Behbahan A, Jamshidi M, Veisi P, Zarrintan S. Intestinal malrotations: a review and report of thirty cases. Folia Morphol. 2007;66(4):277–82.
12. Applegate KE, Anderson JM, Klatte EC. Intestinal malrotation in children: a problem-solving approach to the upper gastrointestinal series. Radiographics : a review publication of the Radiological Society of North America, Inc. 2006;26(5):1485–500.
13. 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.
14. Millar AJ, Rode H, Cywes S. Malrotation and volvulus in infancy and childhood. Semin Pediatr Surg. 2003;12(4):229–36.
15. Dunn MJ, Hood VL. Prostaglandins and the kidney. Am J Physiol-Renal Physiol. 1977;233(3):F169–84.
16. Hornyh A, de Barochez YH, Bariety J, Branca G, Vigeral P, Girard J, et al. Bartter’s syndrome with normal chloride reabsorption during indomethacin treatment. Nephron. 1987;46(2):137–43.

Publisher’s Note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.