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Case report

Secondary Pseudohypoaldosteronism: Salt Wasting in Infant with Posterior Urethral Valves

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HIGHLIGHTS

- A UTI can cause transient secondary type 1 PHA.
- PHA type 1 should be considered in the existence of hyponatremia, hyperkalemia, and metabolic acidosis in newborns with UTI and UTM.
- It can be concluded that the same work is feasible for the following patients to investigate them and be vigilant before operating on them.

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ABSTRACT

Introduction

One of the uncommon diseases and serious one in an infant is hyponatremia with hyperkalemia. Secondary Pseudo hypoaldosteronism (PHA) is congenital adrenal hyperplasia that has been described in babies with urinary tract infection (UTI) and urinary tract malformation (UTM).

Case presentation

Here, we report a CRE guideline-based case of a 39 days infant with failure to thrive and lastly identified with transient PHA because of UTI with posterior urethral valves. Congenital adrenal hyperplasia was excluded in front of cortisol; 17 OH progesterone and adrenocorticotropic hormone (ACTH) were within normal ranges. The infant was put under double antibiotic and sodium replacement therapy and cation exchange resin at the rate of one gram per kilogram, and after stabilizing his clinical condition and sterilizing the urine, the infant underwent endoscopic valve resection.

Conclusions

Transient secondary type 1 PHA caused by a UTI should be considered in the existence of hyponatremia, hyperkalemia, and metabolic acidosis in newborns with UTI and UTM.

Keywords: Secondary PAH; Infant; Urinary Tract Infection; Posterior Urethral Valves

Introduction

TPHA type 1 is an uncommon disorder that can happen in one of 80,000 newborns (1). The renal PHA type I (an autosomal dominant disease) is limited to the kidney with fewer severe disorders and no systemic association. Patients with sodium wasting in the renal, lungs, colorectal, sweat, and salivary glands can have a multisystem disease. Consequently, the reduced sodium-dependent liquid concentration in the infant's lungs can develop pulmonary symptoms like congestion, wheezing, and recurrent pulmonary infections. Renal PHA type I is generally weaker and is likely to diminish as patients get older, while systemic PHA type I stays and needs permanent therapy (1, 3). PHA has a variance diagnosis of Congenital Adrenal Hyperplasia (CAH) in newborns with hyperkalemia, hyponatremia, and metabolic acidosis. PHA also is a famous disorder for pediatricians, mainly pediatric nephrologists and endocrinologists (4, 5). In 1983, Rodriguez-Soriano et al., (6) presented the first report of transient PHA with obstructive uropathy, and Watanabe additionally reviewed studies on infants aged ≤ 7 months who suffered from both UTM and UTI, the same

Table 1. Evolution of blood and urine parameters

	First day	Second day	Third day	Fourth day	Fifth day	Sixth day	Seventh day
Urine Volume	400	800	600	500	500	400	400
Serum Sodium	135	128	129	130	130	132	135
Serum Potassium	6.5	6	7	5.4	5.2	5	5

as our case (7).

Secondary PHA may happen mostly in newborns connected to UTI and UTM, such as hydronephrosis, ureteropelvic junction obstruction, vesicoureteral reflux, and posterior urethral valve (8). In this case report, we described an Infant boy aged 39 days with physical abnormalities of the urinary tract diagnosis with transient type 1 PHA caused by a UTI.

Case presentation

Infant boy aged 39 days, bilateral hydronephrosis was detected in the antenatal ultrasonography (USG), delivered over the cesarean section in the 37th week of pregnancy (3700g weight). Our case reports are based on CARE guidelines, and the patients' parent agreed to report his case after signing the written informed consent. He had an excellent adaptation to extrauterine life, under mixed breastfeeding from birth, polled since the third day of life because he had difficulty urinating and interrupted urination. On his clinical examination, the infant was febrile at 39ºC with suitable weight growth:4.5 kg. His Height is 55cm, and he has a cranial perimeter of 37 cm. We noticed a cloudy appearance of urine.

On the second day of his hospitalization, the infant presents signs of dehydration with the excavation of the eyeballs; the losses are then estimated at 7%, diuresis of 800 cc/24h soit 08 cc/kg/h. The blood pressure was 80/50 mmHg; the heart rate was 130/min. The examination of the genitourinary organs did not note the existence of malformations

Laboratory examinations showed normal blood counts (CBCs), and a C-reactive protein level (CRP) of 96 mg /l. A urinalysis and microscopic urine examination found a urinary tract infection with a klebsiella cloacae. Urea was 0.20g/l; Creatinine was 6.5mg/l, potassium 6,5meq / l, and sodium 135meq / l; blood gas analysis was normal. below is the table of the evolution of the infant's blood and urinary parameters (Table 1). Congenital adrenal hyperplasia was excluded in front of cortisol, 17 OH progesterone and ACTH were within normal ranges, and secondary PHA1 due to obstructive uropathy was diagnosed with renin level at 50 mg/ml (Nl: 20), and the aldosterone level was at 2942 pml/l (Nl: 01 Mois – 02 Ans: 20-1100 pm/ml). The infant was put under double antibiotic and sodium replacement therapy and cation exchange resin at the rate of one gram per kilogram. After stabilizing his clinical condition and sterilizing the urine, the infant underwent endoscopic valve resection. At the last visit, after one month of endoscopic resection, the patient did not receive any additional therapy for one month, and the level of potassium was average.

Discussion

Several studies have previously described the Renal tubular resistance to aldosterone in patients with several UTM, which, in some cases, the electrolyte imbalance was problematic and even life-threatening (5, 6, 9, 10). The S-PHA can happen more frequently in boys, but no apparent reason has been offered in the texts yet. More incidence of UTI in boys is suspected of playing a role, together with the higher frequency of obstructive uropathies (11, 14). Its mechanism is unclear, but it is hypothesized that it might be the consequence of parenchymal scarring secondary to obstruction and tubular aldosterone resistance secondary to endotoxin damage of the aldosterone cytokinesis from cytokines such as TGF-β (7). Then, it is thought that tubular immaturity can cause the pathogenesis of the disease for the reason that this would involve elevated aldosterone levels to keep electrolyte balance (15). It is believed that UTI and urinary tract obstruction can trigger the increased aldosterone resistance in renal tubules directly (unusual cellular response) and indirectly (hormone levels change) (16).

PHA is a renal tubular resistance to aldosterone activity with a heterogeneous group of disorders of electrolytes. The impact of urinary tract obstruction on the growing kidney can be defined based on the time of onset, location, and degree of obstruction. Ureteral obstruction in the first trimester of pregnancy can cause dysplasia of the renal parenchyma and decrease the nephrons. In newborns, incomplete ureteral obstruction initiates renal vasoconstriction, glomerular hypoperfusion, impaired ipsilateral renal growth, and interstitial fibrosis. The healing of renal function following relief of urinary tract obstruction is connected to the patient's age (17). The seriousness of medical and laboratory indications at the diagnosis is contrarywise connected to the patient's age, varying from a newborn life-threatening salt-wasting syndrome to an asymptomatic adult increase of plasma aldosterone concentrations. Some additional indications suggested the revealed obstructive uropathy as the original cause of Pseudohypoaldosteronism. Two-sided obstructive uropathy is the leading cause of PHA because

of the aldosterone insensitivity to the tubules. Acute pyelonephritis in companies with urinary tract anomalies enhances the risk of PHA, even though both can produce aldosterone unresponsiveness independently, like our case report (18). It results in Pseudohypoaldosteronism, mainly affecting nonspecific symptoms like reduced feeding, inadequate weight gain, nausea, failure to thrive, and dehydration.

Some cases have indicated uncontrollable seizures with sequelae, electrolyte abnormalities, and cardiac arrest (19). The symptoms can solve with intravenous fluids (IV), electrolyte correction, and antibiotic therapy. Therefore, early identification is necessary to avoid severe complications and sequelae. Some symptoms get milder as the patient aged, and salt replacement usually is not necessary for patients aged ≥ 2 years. In a comprehensive study of sixty patients less than seven months of age, secondary PHA-1 was reported. The treatment of PHA consists of 0.9% sodium chloride infusion, normalization of potassium levels, antibiotic therapy, and surgical intervention when indicated (relief of urinary obstruction) (19).

Conclusions

Transient secondary type 1 PHA caused by a UTI should be considered in the existence of hyponatremia, hyperkalemia, and metabolic acidosis in newborns with UTI and UTM. The primary treatment strategy on the first to adjust electrolyte defects and, after that, deal with the underlying infection and malformation.

Authors' contributions

All authors contributed equally.

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Conflict of interest

The author declares that there is no conflict of interest.

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Ethical statement

Our case reports are based on CARE guidelines, and the patient's parent agreed to report his case after signing the written informed consent.

Data availability

Data will be provided on request.

Abbreviations

ACTH	Adrenocorticotropic hormone
CAH	Congenital adrenal hyperplasia
CRP	C-reactive protein
PHA	Secondary Pseudo hypoaldosteronism
USG	Ultrasonography
UTI	Urinary tract infection
UTM	Urinary tract malformation

References

1. Chang SS, Grunder S, Hanukoglu A, Rösler A, Mathew P, Hanukoglu I, et al. Mutations in subunits of the epithelial sodium channel cause salt wasting with hyperkalaemic acidosis, pseudohypoaldosteronism type 1. Nature genetics. 1996;12(3):248-53.
2. Hanukoglu I, Hanukoglu A. Epithelial sodium channel (ENaC) family: Phylogeny, structure–function, tissue distribution, and associated inherited diseases. Gene. 2016;579(2):95-132.
3. Fakhr Yasseri A, Mohammadi A, Aghamir SMK. Management of a lost stone during laparoscopic ureterolithotomy: video article. Translational Research In Urology. 2021;3(1):38-9.
4. Krishnappa V, Ross JH, Kenagy DN, Raina R. Secondary or transient pseudohypoaldosteronism associated with urinary tract anomaly and urinary infection: A case report. Urology Case Reports. 2016;8:61-2.
5. Melzi ML, Guez S, Sersale G, Terzi F, Secco E, Marra G, et al. Acute pyelonephritis as a cause of hyponatremia/hyperkalemia in young infants with urinary tract malformations. The Pediatric infectious disease journal. 1995;14(1):56-9.
6. Rodríguez-Soriano J, Vallo A, Oliveros R, Castillo G. Transient pseudohypoaldosteronism secondary to obstructive uropathy in infancy. The Journal of pediatrics. 1983;103(3):375-80.
7. Watanabe T. Reversible secondary pseudohypoaldosteronism. Pediatric Nephrology. 2003;18(5):486-.
8. Proctor G, Linas S. Type 2 pseudohypoaldosteronism: new insights into renal potassium, sodium, and chloride handling. American journal of kidney diseases. 2006;48(4):674-93.
9. Asano T, Abe M, Asai M, Imai T, Kamisago M, Kuwabara K, et al. Urinary tract malformation and infection with hyperkalemia and decreased fractional excretion of potassium in an infant. Journal of Nippon Medical School. 2006;73(5):289-91.
10. Tütüncüler F, Günöz H, Bas F, Bundak R, Saka N, Neyzi O. Transient pseudohypoaldosteronism in an infant with urinary tract anomaly. Pediatrics international. 2004;46(5):618-20.
11. Schoen EJ, Bhatia S, Ray GT, Clapp W, To TT. Transient pseudohypoaldosteronism with hyponatremia-hyperkalemia in infant urinary tract infection. The Journal of urology. 2002;167(2):680-2.
12. Bogdanović R, Stajić N, Putnik J, Paripović A. Transient type 1 pseudo-hypoaldosteronism: report on an eight-patient series and literature review. Pediatric nephrology. 2009;24(11):2167-75.
13. Aghamir SMK, Dadkhah Tehrani F, Khatami F, Zia H. Second-look percutaneous nephrolithotomy: access to the tract with direct vision and fluoroscopic guidance. Translational Research in Urology. 2020;2(4):118-22.
14. Aghamir SMK, Salavati A, Hamidi M, FallahNejad A. Primary Report of Totally Tubeless Percutaneous Nephrolithotomy Despite Pelvi-calyceal Perforations. Urology journal. 2017;14(4):4020-3.
15. Delforge X, Kongolo G, Cauliez A, Braun K, Haraux E, Buisson P. Transient pseudohypoaldosteronism: a potentially severe condition affecting infants with urinary tract malformation. Journal of Pediatric Urology. 2019;15(3):265. e1-. e7.
16. Giapros VI, Tsatsoulis AA, Drougia EA, Kollios KD, Siomou EC, Andronikou SK. Rare causes of acute hyperkalemia in the 1st week of life. Pediatric Nephrology. 2004;19(9):1046-9.
17. Chevalier RL, editor Effects of ureteral obstruction on renal growth. Seminars in Nephrology; 1995.
18. Belot A, Ranchin B, Fichtner C, Pujo L, Rossier BC, Liutkus A, et al. Pseudohypoaldosteronisms, report on a 10-patient series. Nephrology Dialysis Transplantation. 2008;23(5):1636-41.
19. Perez-Brayfield MR, Gatti J, Smith E, Kirsch AJ. Pseudohypoaldosteronism associated with ureterocele and upper pole moiety obstruction. Urology. 2001;57(6):1178.

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