Bartter Syndrome Masquerading as Acute Kidney Injury in a Neonate

Infants and children with Bartter syndrome present with polyuria and polydipsia, whereas older children present with constipation, salt craving and muscle cramps. The symptomatology is mainly due to renal concentrating defect [1]. This disorder is characterized by hypokalemia, hypochloremia, hypercalcuria, salt wasting with metabolic alkalosis. A 10-day-old male child born out of third degree consanguineous marriage presented with severe respiratory distress. The antenatal history was uneventful. The neonate continued to have polyuria inspite of measures to decrease urine output. The infant developed metabolic alkalosis despite acute kidney injury and polyuria. The blood pressures were in normal range. The urine examination showed red blood cells, granular casts and proteinuria. The biochemical features reflect defect in sodium, chloride and potassium transporter on ascending limb of loop of Henle [3]. Serum calcium, vitamin D and parathyroid hormone levels were within normal range. Ultrasonography of kidney and bladder showed calcifications in apex of medullary pyramids suggesting bilateral medullary nephrocalcinosis. We diagnosed our case as type 2 Bartter syndrome.

The classical Bartter syndrome (type 3) is perinatal in onset and presents with polyhydramnios, neonatal salt wasting and recurrent episodes of dehydration. Antenatal Bartter syndrome (type 1, 2 and 4) typically manifests in infancy with severe phenotype compared to the classical syndrome [2]. The biochemical features reflect defect in sodium, chloride and potassium transporter on ascending limb of loop of Henle [3].

Various genes are associated with Bartter syndrome [4]; MAGED2 mutation described recently is associated with transient Bartter syndrome which starts antenatally with severe phenotype and usually resolves by six weeks of age. Our case presented at around six weeks with acute kidney injury without hypomagnesemia [5]. The diagnosis of Bartter syndrome in neonate or infant is suggested by severe hypokalemia, hypochloremia and metabolic alkalosis. Hypercalcuria is typical and nephrocalcinosis is seen resulting from hyper-

REFERENCES
1. Fuloria M, Aschner JL. Persistent pulmonary hypertension of the newborn. Semin Fetal Neonatal Med. 2017;22:220-6.
2. Masarwa R, Paret G, Perlman A, Reif S, Raccab BH, Matok I. Role of vasopressin and terlipressin in refractory shock compared to conventional therapy in the neonatal and pediatric population: A systematic review, meta-analysis, and trial sequential analysis. Crit Care. 2017;21:1.
3. Rodriguez-Nuñez A, López-Herce J, Gil-Antón J, Hernández A, Rey C, RETSPED Working Group of the Spanish Society of Pediatric Intensive Care. Rescue treatment with terlipressin in children with refractory septic shock: A clinical study. Crit Care. 2006;10:R20.
4. Radicioni M, Troiani S, Camerini PG. Effects of terlipressin on pulmonary artery pressure in a septic cooled infant: an echocardiographic assessment. J Perinatol. 2012;32:89-5.
5. Evora PR, Pearson PJ, Schaff HV. Arginine vasopressin induces endothelium-dependent vasodilatation of the pulmonary artery. V1-receptor-mediated production of nitric oxide. Chest. 1993;103:1241-5.
6. Stathopoulos L, Nicaise C, Michel F, Thomachot L, Merrot T, Lagier P, et al. Terlipressin as rescue therapy for refractory pulmonary hypertension in a neonate with a congenital diaphragmatic hernia. J Pediatr Surg. 2011;46:e19-21.

Indian Pediatrics

3rd Edition

Volume 57—September 15, 2020
calciuria in type 1 and 2. Hypomagnesemia is seen in minority. Urinary levels of chloride are also very much elevated which helps in differentiating this picture from chronic vomiting and cystic fibrosis. The tubular defect in Bartter or Gitelman syndrome cannot be corrected [6], but with careful fluid and electrolyte management, long term prognosis is good. We treated the child with proper fluid and electrolyte correction following which hyperkalemia improved. The potassium levels normalised after a period of eight days without any therapy for potassium corrections except for restriction. Urinary electrolytes continued to remain elevated. The child was discharged in a stable condition after establishing oral feeds.

The child followed-up with us two weeks after the discharge which was uneventful. Our case focuses lights on the rare presentation of Bartter syndrome with acute kidney injury probably due to nephrocalcinosis which might have started in utero.

RAVI TEJA JALADI,* ARNAB BISWAS AND SONALI MITRA
Department of Pediatrics,
Nil Ratan Sircar Medical College and Hospital,
West Bengal, Kolkata, India.
*jaladiraviteja@gmail.com

REFERENCES
1. Sinha A, Bagga A. Tubular disorders. In: Srivastava RN, Bagg A, editors. Pediatric Nephrology, 6th edition. Jaypee publishers; 2016. p. 312-16.
2. Dixon BP, Bartter syndrome, inherited tubular transport abnormalities. In: Kliegman, St Geme, Blum, Shah, Tasker, Wilson, editors. Nelsons Textbook of Paediatrics, 21st edition. Elsevier; 2019; p. 2767-9.
3. Kurtz I. Molecular pathogenesis of Bartter’s and Gitelman’s syndromes. Kidney Int. 1998;54:1396-410.
4. Simon DB, Karet FE, Rodriguez-Soriano J, Hamdan JH, DiPietro A, Trachtman H, et al. Genetic heterogeneity of Bartter’s syndrome revealed by mutations in the K+ channel, ROMK: Classic diseases revisited. Nat Genet. 1996;14:152-6.
5. Laghmani K, Beck BB, Yang SS, Seaayfan E, Wenzel A, Reusch B, et al. Polyhydramnios, transient antenatal Bartter’s syndrome, and MAGED2 mutations: N Engl J Med. 2016;374:1853-63.
6. Fujiita T, Ando K, Sato Y, Yamashita K, Nomura M, Fukui T. Independent roles of prostaglandins and the renin-angiotensin system in abnormal vascular reactivity in Bartter’s syndrome. Am J Med. 1982;73:71-6.

Fetal Ovarian Cyst Managed Laparoscopically in the Neonatal Period

Most antenatally diagnosed fetal cystic lesions are of renal or ovarian origin, and timely postnatal diagnosis facilitates early and appropriate management. We report early diagnosis of a fetal abdominal cyst with successful laparoscopic management.

A 1900 gram female baby was born vaginally at 38 weeks to a 24-year-old second gravid mother who had conceived spontaneously. Antenatal period was uneventful. Sonography at 30 weeks of gestation revealed a large well defined intra-abdominal fetal cystic lesion extending from pelvis to sub-hepatic region measuring, 4.8 cm × 4.2 cm × 4.8 cm with evident septations, with maximum wall thickness of 3.5mm. No subsequent antenatal scans were available. Baby did not need any resuscitation after birth but was detected with a palpable lower abdominal lump that was cystic in consistency. Rest of the examination including vitals was normal. A postnatal abdominal sonography showed a large cystic mass located in the right flank extending from sub-hepatic region to the pelvis measuring approximately 6.1 cm × 4 cm × 4.6 cm in size with internal solid areas (possibly fibrinous products) with no obvious vascularity or fluid debris level. Right ovary was not visualized, right kidney was seen distinctly separate from the cyst, and uterus, left ovary and left kidney were normal. Plain X-ray abdomen revealed displacement of bowel loops to left side. These findings were consistent with the diagnosis of ovarian cyst with internal hemorrhage (complicated). A thyroid scan performed later was normal. Laparoscopic excision of cyst with preservation of rest of the ovary was performed using three ports and a maximum of 10mm pneumo-peritoneum on day 8 of life. The cyst was seen to originate from right ovary, had a short pedicle and had undergone torsion on its own axis. Dark brown color fluid was aspirated from cyst, which was excised with Harmonic as energy source. Histopathological examination of excised cyst revealed complicated ovarian cyst with necrosed wall. Left ovary was normal. Intraoperative and postoperative course was uncomplicated. Breastfeeding was started on first postoperative day. The baby is currently on follow-up, is feeding and growing normally.

Fetal cystic masses in females are mostly benign and ovarian in origin. In a case series of 41 fetal abdominal cysts, 21 were ovarian cysts whereas 11, 6 and 3 cases were found to be bile duct cyst, intestinal duplication and mesenteric cysts respectively [1]. An antenatally detected isolated, non-lethal lesion should be monitored with repeated ultrasound examination, as the evolution of such a lesion in utero is extremely variable [2]. Serial antenatal ultrasounds help to determine the location and nature of the cyst and plan management. Accurate delineation of the mass may require fetal MRI.

Ovarian cysts are the commonest ovarian tumors in newborn period. Simple ovarian cysts are characteristically