Association between infantile nephrotic syndrome and Intussusception, case report

Naif F. Abdulmajeed [1], Ayesha A. Aldhamer [2], Fatimah A. Aldhamer [3]
Majed S. Alofi [1]

(1) Prince Sultan Medical City, King khalid University, Pediatric Nephrology Consultant, Saudi Arabia.
(2) Prince Sultan Military Medical city, King khalid University, General practitioner, Saudi Arabia.
(3) Prince Sultan Cardiac Center, General Practitioner, Saudi Arabia

Correspondence
Dr. Ayesha Adel Aldhamer
Prince sultan medical city, Al-Riyadh,
Saudi Arabia

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Abstract

Intussusception refers to the invagination (telescoping) of a portion of the intestine into itself. It is the most frequent abdominal emergency among children. Infantile nephrotic syndrome is a triad of proteinuria, hypoalbuminemia, and oedema that present between the age of 3 month to 12 month of age. Although both intussusception and infantile nephrotic syndrome are common in early childhood, there coexisting is unusual. We present here a rare case of 8-month-old, baby girl, known case of infantile nephrotic syndrome that was admitted due to distended abdomen, irritability and poor oral intake. Upon examination she was active, well hydrated, slightly tachypenic and distressed. Abdominal exam showed distended yet soft and fluctuant abdomen. Laboratory investigation upon admission showed leukocytosis, thrombocytosis, high serum creatinine and urea with low albumin. Abdominal ultrasound reveal ileocolic intussusception. Patient was treated with fluoroscopy and barium enema. Patient's original disease was treated conservatively and two doses of albumin infusion was given.

Keywords: intussusception, case report, infantile nephrotic syndrome

Introduction

Intussusception refers to the invagination (telescoping) of a portion of the intestine into itself. It is the most frequent abdominal emergency among children. Intussusception typically presents between 6 and 36 months of age. It usually presents with a classic triad of symptoms: paroxysmal abdominal pain, bloody stool, and vomiting. However, this classic triad appears only in 10–20% of intussusception cases. Many patients present with only irritability or other non-specific complaints.

Infantile nephrotic syndrome is a triad of proteinuria, hypoalbuminemia and oedema that presents between 3 months to 12 months of age. In contrast to idiopathic nephrotic syndrome presenting in children >12 months of age, the renal outcome of infantile nephrotic syndrome is generally poor with the majority developing chronic kidney disease stage 5, although the age at which this develops can be variable.

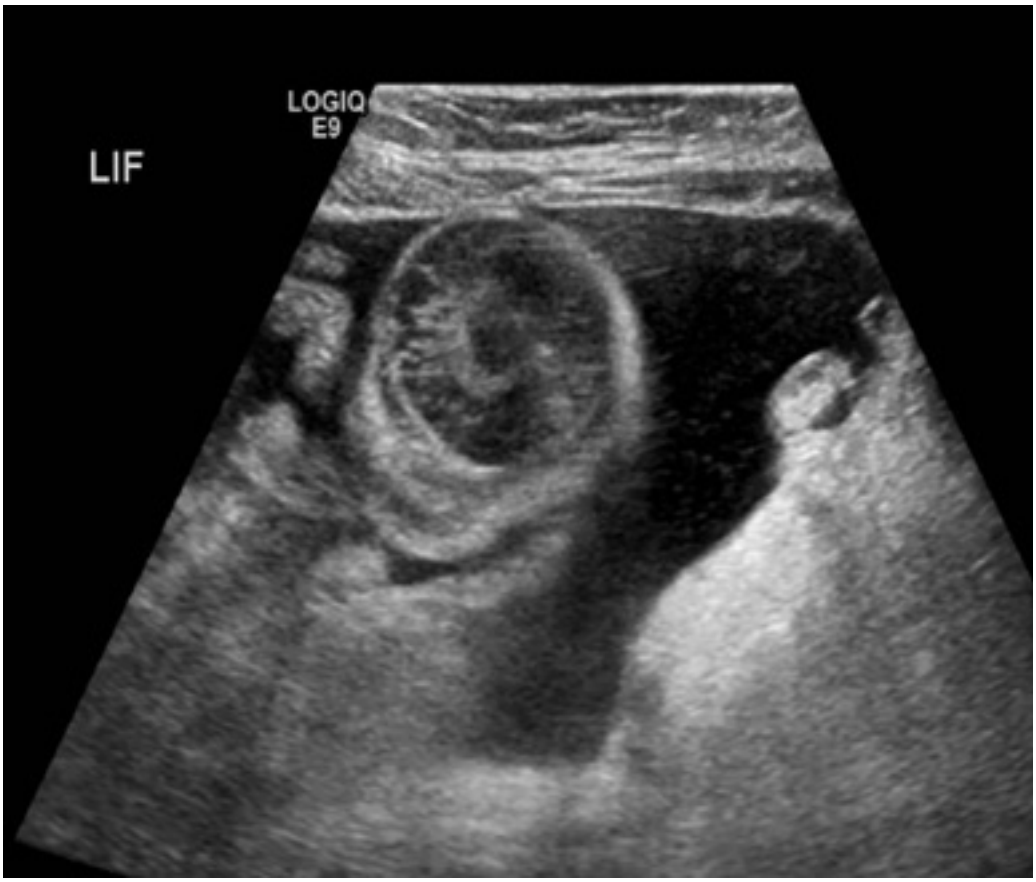
Case Report

An 8-month-old baby girl, full term, was delivered via C-Section after uneventful pregnancy. No neonatal intensive care unit (NICU) admission, No family history of any renal, autoimmune, or genetic disease. She was diagnosed as Nephrotic syndrome on October 2022, and started on Prednisolone. Percutaneous renal biopsy was performed on November 2022 and demonstrated features consistent with diffuse mesangial sclerosis. The diagnosis of infantile nephrotic syndrome was established, and prednisolone was tapered.

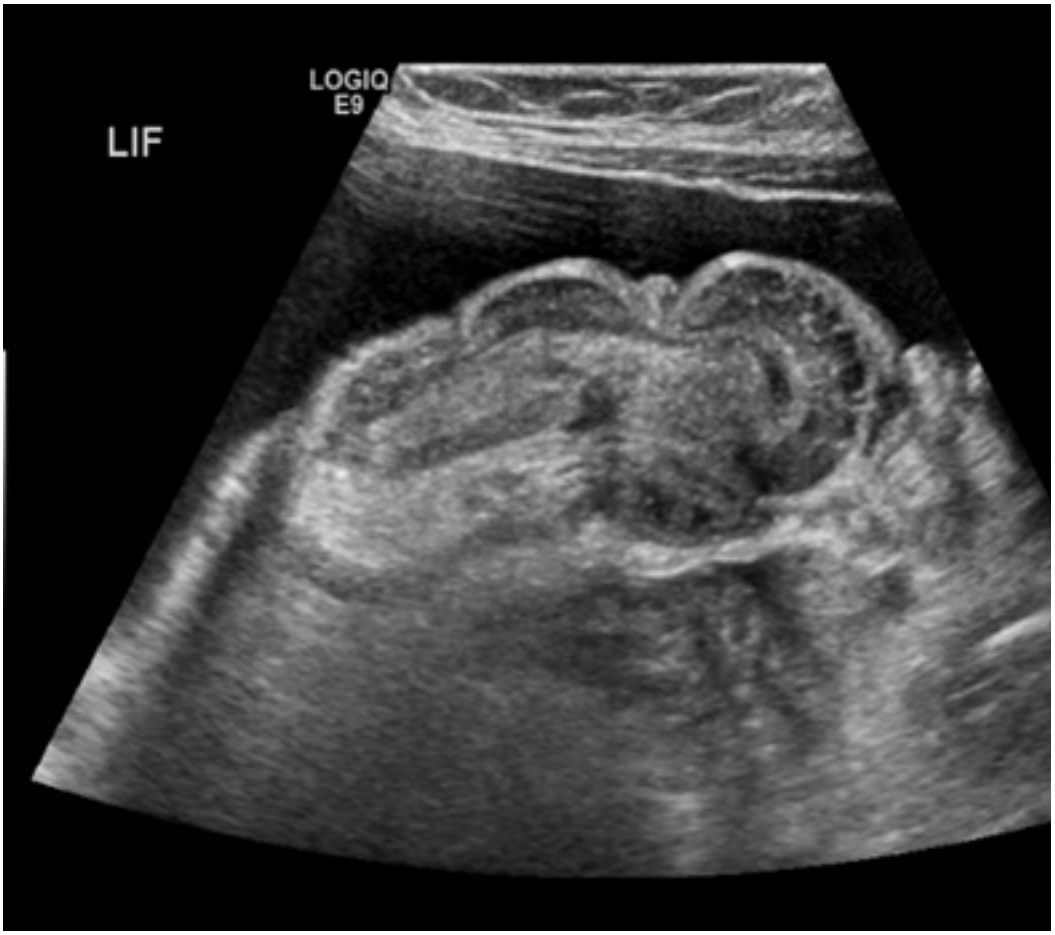
In December 2022, patient was admitted due to abdominal distension (abdominal girth = 63.8 cm), lower limb oedema, cushioned face with excessive crying, irritability and poor oral feeding. Upon examination, she looked well, active, slightly tachypnic, in distress with normal oxygen saturation. She was well-hydrates, afibril , not pale, with normal capillary refill and warm extremities. Her length was 68cm and her weight was 11.5kg, Her Blood pressure was on the 90th blood pressure percentile for her age and length. she had moon face (cushingoid face) with no other dysmorphic feature. She also had mild periorbital oedema, bilateral lower limb pitting oedema reaching her knee and no labial or sacral involvement. abdominal examination revealed distended, round, symmetrical contoured abdomen with no scarring on inspection. On palpation, abdomen was soft and fluctuant with no guarding. Chest exams revealed clear chest with normal breath sound. Cardiac exam was normal.

Laboratory tests showed leukocytosis (WBC = 25.07 10^9/L) normal range (6.0-17.5), thrombocytosis (PLT =737 10^9/L) normal range (150-450), high serum creatinine (CREA = 87 mcmol/L) normal range (14 -34), high urea (18.3 mmol/L normal range 2.8 -8.1) and low albumin (26 g/L normal range 38-54). Urine analysis showed (UA WBC= 25.40mcl, normal value is equal or below 5.01) and (UA Cast =2.88mcl, normal value is equal or below 2.0) urine culture was positive to two type of bacteria, Escherichia coli and Enterobacter asburiae. Patient was diagnosed with urinary tract infection (UTI) and full course of intravenous (IV) antibiotics was given according to her culture and sensitivity. During her admission, patient defecation decreased in frequency and became harder in consistency. A few days later, patient passed currant jelly stool. Abdominal ultrasound (US) was requested and showed a mass of intussuscepted bowel loops seen in all 4 quadrants of the abdomen demonstrating a target sign. The intussuscepted bowel loops were thickened. Color Doppler of interstitial septal bowel loops showed preserved color vascularity. There were a large amount of ascites. No identifiable debris or septation of the ascites was visualized. These findings are suggestive of ileocolic intussusception (likely in the long segment).

Pediatric surgery team was consulted, and water-soluble contrast enema examination was performed via 16 Fr Foleys catheter utilizing 100 cc of Telebrix contrast. Scout image of the abdomen and pelvis showed paucity of bowel loops which were displaced to the left side. During the filling stage, contrast passed through the rectum and intussusception was identified in the proximal sigmoid colon. Afterwards, contrast passed slowly, and pushed the intussusceptum through the colon. The cecum was located medial to the midline, no focal stricture, fistula or contrast extravasation were noticed. The contrast was refluxed into the terminal ileum. The procedure went smoothly, and reduction of ileocolic long-segment intussusception was achieved by fluoroscopy with barium enema. The patient recovered well and was further managed for nephrotic syndrome with albumin infusion and conservative management.



Abdominal ultrasonography showing target sign of the intussusception in transverse plane associated with adjacent free fluid.



Telescoping of bowel loops with alternating hypoechoic and hyperechoic layers resulted in pseudo-kidney sign on the longitudinal plane.

Discussion

The top differential diagnosis for abdominal pain in nephrotic syndrome patient is usually UTI (25- 66.7%)1 or spontaneous bacterial peritonitis (10.8%)2. Although only few cases of intussusception in association with nephrotic syndrome were reported worldwide, the association between these two diseases is not absurd. Hence nephrotic syndrome causes hypoproteinemia which sequentially will result in ascites. This may cause functional bowel obstruction, disturb normal bowel peristaltic and then intussusception. Furthermore, nephrotic syndrome affects potassium secretion which causes hyperkalemia and other electrolytes disturbances which could also result in disturbed bowel movement. Finally, nephrotic syndrome patients with ascites have a high risk of peritonitis and that means multiple infections then fibrosis, which can work as an anatomical leading point, then intussusception.

Ultrasound abdomen is highly sensitive and specific in the diagnosis of intussusception. It is also safe for the pediatric age group. It has a crucial role in ruling out any anatomical source of abdominal pain, bowel disease, or urinary tract abnormalities which could be associated with UTI, and it is also helpful in confirming the presence of ascites fluid.

Conclusion

We presented a rare case of Intussusception in an infant with nephrotic syndrome. High index of suspicion with early consideration of abdominal US are crucial in diagnosing this rare presentation.

Consent for publication

Informed consent was obtained from the patient parents to publish this case report in a medical journal.

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