Case Report

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Recurrent Intussusceptions in both Twins with Congenital Nephrotic Syndrome

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Congenital Nephrotic Syndrome (CNS) is an autosomal recessive (AR) disorder characterized by massive proteinuria, hypoalbuminemia, and severe edema, starting in the first 3 months of life. Intussusception on the other hand is a surgical condition requiring a leading point in most of the cases. The occurrence of intussusception in congenital nephrotic syndrome may be due to bowel edema as the leading point. In this report, we present monozygotic twins with congenital nephrotic syndrome, who experienced the first episode of intussusception at ten months of age and then had repeated episodes until they were 18 months old. In our patients, mesenteric lymphadenopathy was reported by a radiologist that could serve as a leading point. Upon laboratory examinations, we could not find any explanation for this report. Intussusception must be one of the differential diagnoses of abdominal pain in nephrotic syndrome patients since prompt diagnosis and appropriate treatment can prevent further complications.

Keywords: Congenital nephrosis; Intussusception; Child

Running Title: Recurrent Intussusceptions in both twins

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Introduction

Congenital Nephrotic Syndrome (CNS) is an AR disorder, defined as proteinuria manifesting in the first three months of life. Primary CNS is typically caused by mutations in genes encoding for the components of the glomerular filtration barrier. Because edema of the bowel and reactive mesenteric lymph nodes are common in these patients, they are susceptible to intussusceptions. Therefore, intussusception should be one of the differential diagnoses of abdominal pain other than bowel ischemia, thrombosis of the renal vessels, and subacute bacterial peritonitis in these patients [1,2].

Case Report

Our patients were monozygotic female twins who were ten months old when they had the first episode of intussusceptions. Prenatal history was unremarkable with a questionable large placenta. Alpha-fetoprotein (αFPr) was not assessed in the amniotic fluid. After birth, the pediatrician faced unexpectedly edematous twins. They were term with birth weights of 2700 and 2400 gr. Their parents were non-consanguineous. Urine analysis revealed 2+ proteinuria and random Pr/Cr for both twins was significantly higher than normal (Pr/Cr>20/1). During these 10 months of life, they were admitted for anasarca several times and received intravenous 20% albumin and furosemide. The lab data at three months of age was as follows: CBC: WBC=7900 /µl, RBC=4000000 /µl, Hb=13.7 gr/dl, MCV=95.5, platelets=372000 /µl, BUN=5, Cr=0.35, uric acid=4.1, Ca=7.4, P=5.5, Na=128, K=5, Total protein=2.7, Albumin=1.2, Globulin=1.5, A/G ratio=0.8, and αFPr=53789 IU/ml. On urinalysis, WBC=6- 8, RBC=4-5, Ph=7, SG=1015, Pr=+3, Blood=+1. Thyroid function test results were

T3=1.1 (0.8-2.8), T4=2.9 (5.3-16), and TSH=6.6 (0.6-8.1). On ultrasound evaluation at that time, the kidneys were normal in size with increased paranchymal echogenicity. The medications were as follows: Captopril 12.5 mg/day, Losartan = 6.25 mg/day, hydrochlorthiazide=6.25 mg/day, Multi-Vit=10 drop/day, Levothyroxin=2.5 µg/day. At about five months of age, they gradually developed hyperphosphatemia, hyperkalemia, hyperuricemia, hypertriglyceridemia, and hypercholesterolemia. The indices at that time were as follows: CBC: WBC=10800/µl, Hb=7gr/dl, MCV=72.8, platelets=267000/µl, BUN=15, Cr=0.5, uric acid=7.7, Ca=10.3, P=9.5, Na=140, K=6, Total protein=1.8. Albumin=1.3, TG=857, Cholesterol=226. Random urinalysis showed Pr=158 mg/dl and Cr=15 mg/dl. Thus, phosphate binders, kayoxalate and Gemfibrozil were added to their medications. Renal function gradually aggravated in both twins. At ten months, their height and weight was 70 cm and 9 kg, respectively. The lab data in first twin was: BUN=62, Cr=1.8(GFR=20), Na=142, K=6.8, Ca=8, P=8.5, uric acid=6.5, TG=1335, Cholestrol=276, Albumin=1.4, CBC: WBC=10000, Hb=9.5, Plt=416000, U/A: (WBC=8-10, RBC=5-6, PH=6.5, SG=1015, PR=+3, Blood=+1). In the second twin: BUN=40, Cr=1.2, Na=135, K=5.6, Ca=7.8, P=9, uric acid=80, TG=950, Cholesterol=250, Albumin=1.5, VBG (PH=7.37, PCo2=27, HCO3=15.6).

At this time, the first twin was admitted for abdominal pain and vomiting; the other twin manifested the same symptoms three days later. On ultrasound evaluation, there was a mass in the RUQ and epigastric and LUQ regions had the appearance of the pseudokidney (Fig.1). This mass contained several reactive lymph nodes. Intussusception was reduced with barium enema in both twins (Fig.2). Twenty days later, they were admitted with previous symptoms and currant jelly stool. Intussusception was detected on sonography and treated with closed reduction. The first twin had four episodes until 14 months of age when she was admitted with confusion and respiratory distress. Lab data showed she had severe metabolic acidosis and anemia: CBC: WBC=6500, Hb=5.5, Plt=360000, BUN=4, Cr=2.7, Na=134, K=6, Ca=6, P=9.5, uric acid= 14, VBG (PH=7.20, PCo2=12, HCO3=5.5). CXR revealed pulmonary edema. She was admitted in PICU and received antibiotics, packed cell, bicarbonate, and phosphate binders; peritoneal dialysis began at this time. The second twin was admitted three months later with the fifth episode of intussusception. This time, she was treated with open reduction. Because of uremia and metabolic acidosis, the Tenckhoff catheter was inserted simultaneously and peritoneal dialysis began with low volume. Her lab data was as follows:

CBC: WBC=7700, Hb=6.1, Plt=230000, BUN=65, Cr=3.8, Na=133, K=6.1, Ca=6.6, P=9. VBG (PH=7.23, PCo2=11, HCO3=4.5).

We would like to report another case of nephrotic syndrome and intussusceptions. She was an 8-year-old girl who was diagnosed as steroid resistance nephrotic syndrome since she was 5 years old. She presented with edema, proteinuria, and hypoalbuminemia. She had acute renal failure at that time, therefore she was dialyzed for three months. Percutaneous renal biopsy was performed and demonstrated mesengial proliferation with a focus of crescent formation. She received prednisolone 2 mg/kg and mycophenolate 250 mg/8h orally. She was at remission for ten months. Her drug history was atenolol 25 mg/12h, losartan 25 mg/daily, prednisolone 37.5 mg/EOD, penicillin-V 250 mg/daily, and captopril 6.25 mg/12h. She admitted with fever and edema at 8 years of age. Physical examination revealed high blood pressure, lower limb edema, and ascites. Three days later, she developed acute colicky abdominal pain and rectorrhagia. On physical examination, her abdomen was generally tender. Peritoneal fluid was tapped and analyzed (WBC=9800; 90% poly and 10% lymph, RBC=10000, Glucose=133 mg/dl, protein=480 mg/dl). Other lab data were as follows: CBC: WBC=12000, Hb=7.4, Plt=604000, BS=84, BUN=33, Cr=0.9, Na=139, K=3.9, Ca=8.7, P=3.8, Total Pr=5.4, Albumin=2.8. U/A results were SG=1015, PH=5, Pr=+3, Blood=+1, WBC=8-10, RBC=15-20. Complements were normal and autoimmune serology was negative. The patient was treated with intravenous meropenem, vancomycin, and metronidazol; 48 hours later, tap of the peritoneal fluid was repeated (WBC=480; 80% poly and 20% lymph, RBC=16000). Despite treatment of peritonitis, abdominal pain progressed. The abdominal x-ray was unremarkable. Abdominal ultrasound showed an iliocolic intussusception. Segmental resection of the ileum and cecum was performed. Pathology reported hemorrhagic necrosis in the small and large intestinal wall due to intussusception.

Discussion

Gastrointestinal disturbances are encountered frequently in the course of nephrotic syndrome, but intussusception is a rare association. It may be the result of uncoordinated motility and bowel wall edema. Intussusception should be included in the differential diagnoses of relapsing nephritic syndrome presenting with acute abdominal pain. Abdominal ultrasound is helpful for confirming this condition [3,4].

Conclusions

We concluded that in nephrotic syndrome, we should consider intussusception in the differential diagnosis of abdominal pain, since early diagnosis may improve prognosis.
References:

Conflict of Interest

None declared

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None declared

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