Pseudo prune belly syndrome

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Case report

A 2.5 month old male infant was admitted with a history of fever for 5 days. He had bilateral cryptorchidism with palpable right kidney. The abdominal wall was normal (Figure 1). There was no other systemic abnormality. Antenatal scan was not performed.

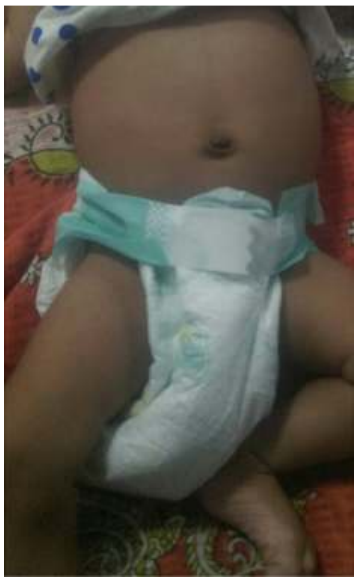

Figure 1: ***Normal abdominal wall***

The haemoglobin level was 10g/dl and the total leucocyte count 31,400/cu mm (neutrophils 58%, lymphocytes 38%). C-reactive protein was 25.2 mg/L, blood urea 50mg/dl, the serum creatinine 0.52mg/dl with normal serum electrolytes and normal liver function. Urine had plenty of pus cells per high power field and urine culture revealed growth of E.coli which was sensitive to carbapenems, amikacin, nitrofurantoin and cotrimoxazole. Magnetic resonance imaging (MRI) of kidney-ureter-bladder showed enlarged right kidney with hydronephroureterosis with grade IV vesico-ureteric reflux (VUR) and multi cystic dysplastic kidney with almost absent renal parenchyma on left side. The right ureter was dilated and tortuous and the left ureter was not visualised (Figure 1).

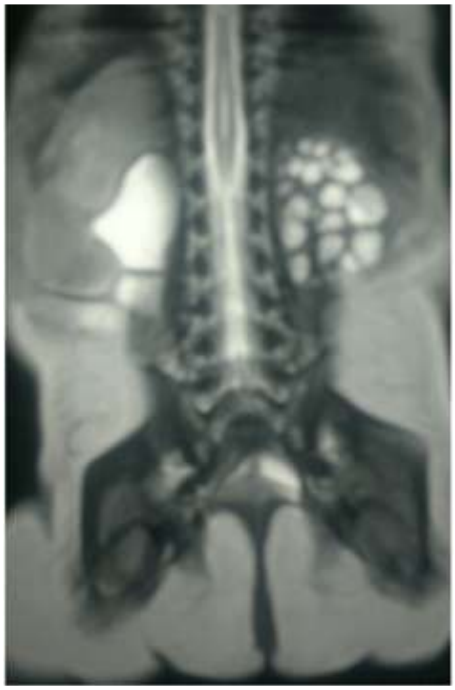

Figure 2: ***MRI abdomen showing enlarged right kidney with hydronephroureterosis***

Renal scintigraphy showed a functionality of 63.84% in the right kidney and 16.76% in the left kidney. Ultrasonography located the right testis within the inguinal canal, but the left was not visible. The presence of renal abnormalities leading to urinary tract infection (UTI) with cryptorchidism in an infant with normal abdominal wall confirmed the diagnosis of Pseudo Prune Belly syndrome (PPBS). He was treated with Meropenem and is presently in follow up with monitoring of blood pressure and renal function. Orchidopexy has been planned.

Discussion

Prune Belly Syndrome (PBS) is characterised by complete or partial abdominal wall deficiency, bilateral cryptorchidism and urinary tract

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abnormality. The incidence varies between one in 35,000 to one in 50,000 with greater predominance in males[1,2]. Mutation is usually sporadic although familial forms are seen[3]. It possibly occurs due to an aberration in mesenchymal development that combines to produce abdominal, urologic and testicular abnormalities. The paraxial mesoderm may be less affected causing sparing of the abdominal wall[4]. PPBS refers to those where one of the triad are absent. Our index case had normal abdominal wall. Urinary tract abnormalities were detected following imaging, similar to the study by Lebowitz and Griscom[5]. Diagnosis is often delayed because of normal appearance of abdominal wall in PPBS[5]. Prognostication is determined by urinary tract abnormalities and renal function.

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