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Prune belly syndrome in an adult Nigerian: case report

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Summary
Prune Belly Syndrome is a rare congenital anomaly characterized by deficient anterior abdominal wall musculature, bilateral cryptorchidism, bilateral megaureters and often unilateral or bilateral vesico-ureteric junction obstruction. The report of prune belly syndrome in the adult is scanty. We report a case of prune belly syndrome in a 24 year old Nigerian who presented with 3 year history of recurrent right loin pain. Examination showed wrinkled abdominal skin, bilateral undescended testes and an hypoplastic rectus abdominis, below the umbilicus. Further evaluation revealed enlarged bladder, bilateral megaureters and right intra-abdominal testis. A diagnosis of Prune Belly Syndrome was made. The challenges in the diagnosis and management of this rare condition are highlighted in this presentation.

Keywords: *Prune belly, cryptorchidism, megaureter, abdominal wall defect*

Résumé
Le syndrome de l'estomac ballonné est une anomalie congénitale rare caractérisée par une insuffisance musculaire de la paroi abdominale antérieur, le cryptorchidie bilatéral, les méga uretères bilatéraux et souvent, l'obstruction de la jonction vesico-uretère unilatérale ou bilatérale. Le rapport du syndrome de l'estomac ballonné chez les adultes est rare. Nous avons reporté un cas de syndrome de l'estomac ballonné chez un Nigérian de 24 ans qui présentait depuis 3 ans des douleurs récurrentes a droite. Les examinassions ont montre des plis sur la peau de son abdomen. Les évaluations supplémentaires ont révélées une vessie élargie, les méga-uretères bilatéraux et le testicule droit intra-abdominal. Le diagnostique du syndrome de l'estomac ballonne était fait, le défit dans le diagnostique et le gestion de cette condition rare était éclairé dans cette étude.

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Introduction

Prune Belly Syndrome was first described by Frolich in 1839 and has been known to Urologist since 1895. It is also referred to as Eagle Barette Syndrome [1], Obrinsky Syndrome [2] and Triad Syndrome [2]. The classical presentation consists of abdominal wall defects, urinary tract abnormalities and bilateral cryptorchdism. Other clinical features are pulmonary hypoplasia, cardiac and bony abnormalities. This condition is common amongst black Americans, twins, neonates and children with incidence of 1/35,000 to 1/50,000 births [3]. However it is seldom reported in Nigeria [4,5,6] and there are no figures on prevalence and incidence. We report the presentation in a young adult Nigerian.

Case report

A 24 year old male Nigerian, presented at the out patient clinic, with a 3 year history of recurrent colicky right loin pain. There were no constitutional symptoms. He had bilateral undescended testes for which left orchidopexy was done about 3 years prior to this presentation. There was no family history of similar complaint. He was a healthy looking man of normal built and average height. He had wrinkled abdominal skin, hypoplastic rectus abdominal musculature below the umbilicus, healed left groin scar with palpable testicule in the left inguinal canal, atrophic scrotum with normal phallus and normally situated external meatus (fig. 1). There was no abnormality in the other systems. A clinical diagnosis of Prune Belly Syndrome with right ureteric colic was made. Chest X Ray and electrocardiogram were normal. Abdominal ultrasound showed bilateral hydronephrosis and hydroureter, there was no stone. An intravenous urogram done showed prompt excretion on the right and the left kidneys with narrowing at (R) PUJ and grade 4 hydroureter on the left (fig. 2). Subsequent right retrograde ureterogram (fig. 4) showed grade 4 hydroureter and kinked pelvi-ureteric junction. He however defaulted from clinic followed up and only to re-represent 10 months later with the same symptoms. Micturating cystourethrogram (MCUG) showed dilated bladder and posterior urethra with no demonstrable vesico

ureteric reflux (fig. 5). Serum creatinine, urea and electrolytes were normal.

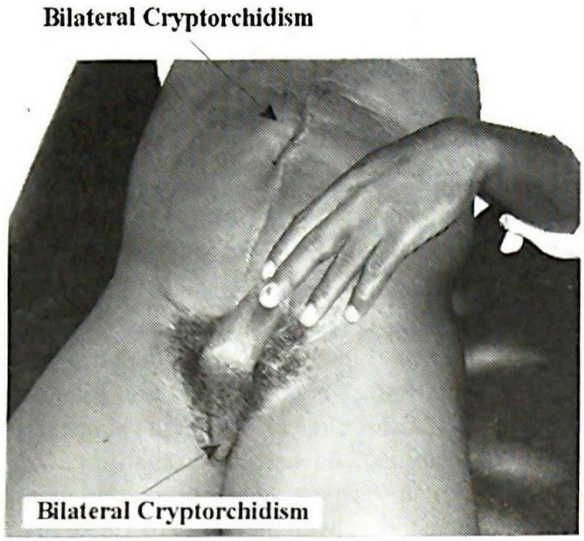

Fig. 1: Clinical photograph showing wrinkled abdominal skin and bilateral cryptorchidism. The rectus abdominis is hypolastic.

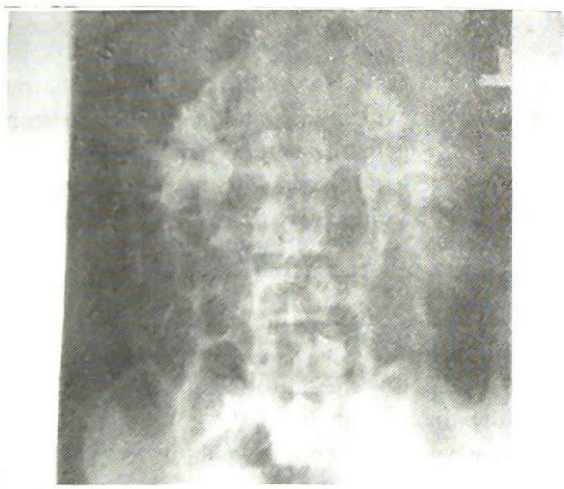

Fig. 2: Intravenous urography showing bilateral hydronephrosis and left megaureter

Pre-operative diagnosis of torsion/detorsion of the (R) undescended testis was made, with recurrent appendicitis as a differential diagnosis. At exploratory laparatomy (fig. 5) we found a floating caecum and non-inflamed appendix in the right sub hepatic region , normal looking right testis on psoas muscle, dilatation of both ureters with normal peristalsis, grossly dilated thick walled urinary bladder up to the umbilicus with fibrotic urachus, easily catherised ureteric orifices with 6fr whistle tipped

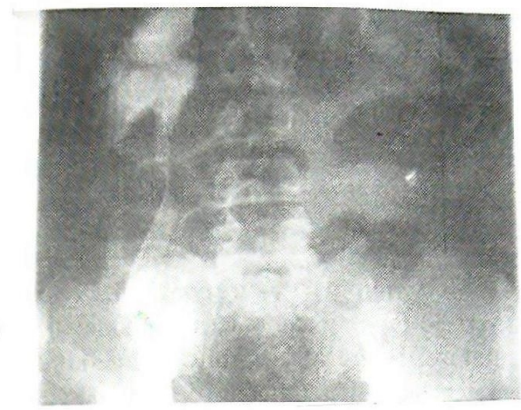

Fig. 3: Right retrograde ureterogram showing R megaureter

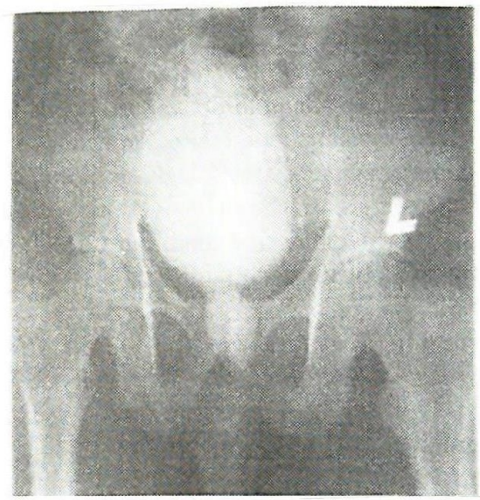

Fig. 4: Micturating cystourethrogram showing grossly dilated bladder and posterior urethra.

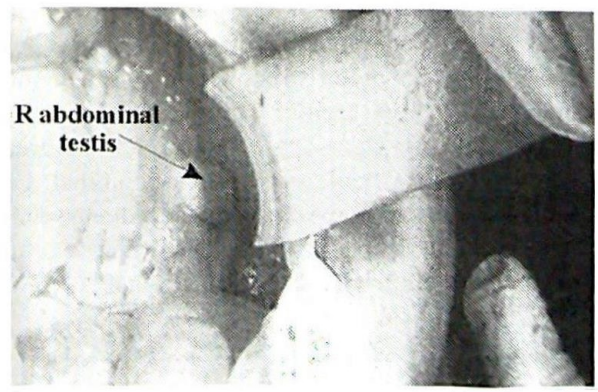

Fig. 5: Intraoperative retroperitoneal right testis

ureteric catheter. Other intra abdominal organs were normal. The right testes was mobilized and fixed with 4/0 vicryl suture to the anterior abdominal wall at the right iliac fossa. Post-operative recovery was uneventful and was discharged home one week. He has remained stable one year after discharged.

Discussion

Prune Belly Syndrome is a rare congenital abnormality with classical triads of abdominal wall defects, urinary tract abnormalities and bilateral cryptorchidism, all amenable to surgical corrections if detected early [1,8,9,11,13]. It is a clinical syndrome commonly diagnosed in childhood in the western world where imaging facilities are readily available and in environment where it is prevalent [1,3]. It is rare in the adult [7,8,9]. However this patient presented late as an adult. The wrinkled skin (worse in infancy and childhood) and the bilateral cryptorchidism(fig I) at birth are good pointers to diagnosis where there is a high index of suspicion. This is not surprising considering the cases reported in Nigeria [4,5,6]. Obstetric and Pediatric specialist care are not readily available to majority of the population in the under-developed world where patients are delivered outside the hospitals. The possibility of genetic basis was observed in the cases reported in Nigeria [5].

The exact etiology is unknown but studies have suggested the possibility of megacystitis [10] intermediate and lateral mesenchymal arrest during embrogenesis [11]. We postulate that there is an arrest of downward descent of both testis with over whelming ascent of the ureters and the kidneys in our case. The possibility of testis descent inhibitory factor (IDF) was also considered though could not be investigated.

This case presented late with urinary complication of the disease that we suspected to be urolithiasis or vesico-ureteric reflux (VUR). This is different from what obtains in the developed and developing world where congenital anomalies are diagnosed at birth during routine physical examination and or imaging techniques of Ultrasound, Computerized Tomographic Scan or Magnetic Resonance Imaging, where these are available. Any part of the urinary tract may be involved in this anomaly. In addition , secondary changes which may follow infection and or obstruction may complicate the picture so that it may be difficult to differentiate from consequent pathology [12,13,14]. The kidneys may be normal or hypoplastic and may be complicated by hydronephrosis [14]. This patient has bilateral hydronephrosis in normally located kidneys on ultrasound and the IVU(fig. 2). In extreme cases, there is a multicystic kidney with atretic upper ureter [14]. The hydronephrosis may result from pelvi ureteric junctional obstruction or coincide with gross ureteric dilation [14]. Due to the none visualization of the R ureter and the R hydronephrosis(fig. 2), we suspected PUJ obstruction on the (R) side and that necesitated the right retrograde ureterogram (fig III) , which showed dilated (R) ureter.

In this case, the preoperative IVU, retrograde R ureterogram and MCUG was not suggestive of pain from ureteral obstruction or vesicoureteric reflux but possibly from the retroperitoneal testis or recurrent appendicitis.Also to be considered was pain from the megaureter. The intraoperative finding of a normal appendix made one to consider torsion/detorsion of the right undescended testis. This patient had incidental appendectomy, fixation of the caecum and transabdominal orchideopexy. Abdominoplasty was not considered because the defect was mild, in other series this was performed by the plastic surgeons using the Randolph, Ehrlich or Monfort techniques [13]. The dilated posterior urethral was asymptomatic.

He poses a diagnostic challenge because of the age and the recurrent loin pain. In the typical African setting where most abdominal pain are not fully investigated and may resolve before a diagnosis is made, prune belly syndrome should be considered in the different diagnosis.

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