PRUNE BELLY SYNDROME
A RARE CASE REPORT

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ABSTRACT

A case of a rare congenital anomaly of Prune Belly Syndrome (PBS) predominant in males is presented. Incidence is 1 in 40,000 lives births.[1]

The complete syndrome is characterized by ; dilatation of the upper urinary tract, lateral deviation of the dilated ureters large bladder with urachal anomalies, vesicoureteral reflux, dilatation of the prostatic utricle, undescended testis and patchy agenesis of the anterior abdominal wall.[2]All the above features may not necessarily be present in one single case.

The major anomaly is due to failure of proper development of both the urinary tract and anterior abdominal wall. Respiratory, cardiac, skeletal and anorectal malformations may be present as well.

CASE REPORT

A 2 month old baby was referred to University of Nigeria Teaching Hospital, Enugu from a private hospital. It is alarming that despite the gross anomalies staring at this patient's parents, their main aim for seeking medical attention was diarrhoea and fever. The referring doctor observed the following; anterior abdominal wall was deficient, undescended testes, manual expression of urine, and asymmetry of the thoracic cage. Laboratory investigations and results were as follows; Sodium =124meq/l, Potassium=2-8meq/l, Urea=53meq/l, Haemoglobin=10.9g/dl.The patient had excretory urography(IVU) and micturating cystourethrography.The IVU showed distension of the abdomen with fullness at the flanks. Contrast films showed delayed excretion of contrast by the left kidney. The 6hour and 8hour films showed gross hydronephrosis of the left kidney. Function was not visualized on the right kidney.Micturating cystourethrogram showed irregular outline of the bladder, fourth degree reflux, and megaureters with marked tortuosity and lateral displacement of the ureters.Treatment included antibiotics, continuos bladder irrigation and antibiotics.Unfortunately, this patient was so poorly that his parents did not foresee any hope for survival. They insisted on taking him home.

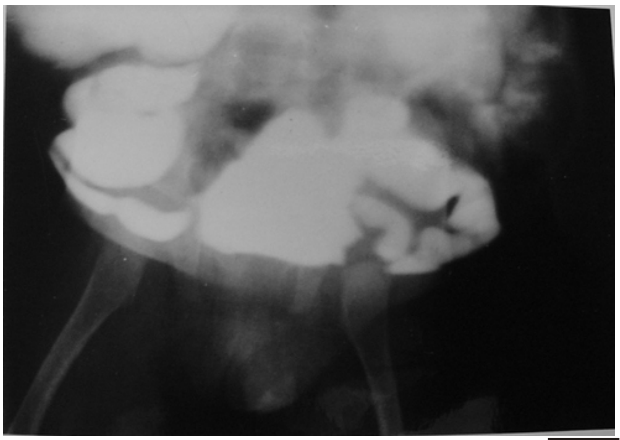

Fig 1

Cystogram showing triangular shaped bladder with irregular outline and wide bladder neck. Reflux filling of laterally displaced dilated, tortous and Redundant ureters is shown. Bilateral hydronephrosis is also Present.

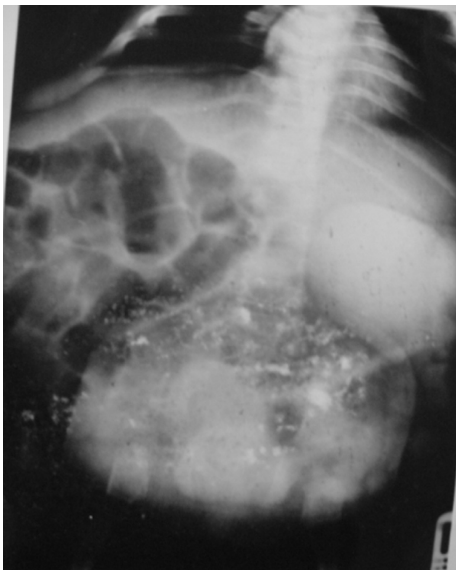

Fig. 2
Intravenous Urography (IVU)Full length IVU showing dilated bowel loops on the right which might have obscured the right hydronephrotic kidney and hydroureter. Bladder is moderately opacified.

DISCUSSION

In 1839,Frohlich described a disease entity in a child which included congenital absence of abdominal muscles and urinary tract abnormalities. In 1950, Eagle and Barret[3] earned credit for recognizing this constellation of findings as a syndrome.
This syndrome (PBS) occurs as a result of failure of the lateral mesoderm to migrate or differentiate into the musculature of the abdominal wall and urinary tract[4]. The migration in normal circumstances occurs about the tenth week of intrauterine life. However, cases reported so far have shown normal chromosomal features. PBS therefore does not seem to have any genetic basis. The abdominal wall is wrinkled and this is responsible for the prune like appearance of the abdomen. The Abdomen diameter shows as a large ballooned out distended abdomen whose diameter is several times larger than the thoracic diameter. The abdominal wall is thin and membranous. The above features were observed in this patient. The urinary tract is predominantly affected and as such excretory urography is a very useful investigation. Dilatation of the renal pelvis, clubbing of the renal calyces and diminished number of calyces is characteristic of prune belly syndrome. Ultrasound will reveal diffuse parenchymal cysts and hypodysplasia Sometimes the kidneys may be small and globular with four to five stubby calyces and a small poorly developed pelvis tucked away at one end. At other times the kidney is grossly hydronephrotic with distended calyces as well as a thin shell of cortex peripherally as in the present case.
Micturating cystourethrogram will reveal enlarged bladder with a vertical orientation and thick walls as seen hereunder. A persistent urachus may be present and is represented by a bulging conical dome of the bladder.Vesicoureteral reflux and large residual volume are common features as seen in the case discussed herein with fourth degree reflux.
.Though the posterior urethra may be elongated or dilated, the presence of a valve is unusual in prune belly syndrome. A persistent prostatic utricle may be present in the posterior urethra at the verumontanum.
Histology shows sparse and patchy muscles in the urinary bladder. The muscles are replaced by mature fibrous tissue and collagen in varying amounts. In electron microscopy, myofilaments appear thin and scanty. The inferior aspect of the recti and oblique muscles are also replaced by thin fibrous tissue. Electron microscopy has revealed that there is arrest of normal muscle development.
Ultrasonography may reveal obstructive cystic renal dysplastic features in some cases.
Occasionally, prenatal ultrasound may reveal fetal ascites or perinephric urinoma. This is as a result of decompensation of the urinary tract. Oligohydramnios is a common feature and when seen, portends poor prognosis. As a result of olygohydramnios a myriad of congenital anomalies may arise as a result of pressure exerted by the uterine wall on the fetus. These anomalies

include the following; flattened cranium, facies and nose; low set ears and chin; limb anomalies like clubfoot, contractures and hip dislocation. Loss of normal stenting effect of the amniotic fluid results in pulmonary hypoplasia.The patient in question had asymmetry of the thoracic cage.

Renal scintigraphy using 99mTc-DMSA could be used to monitor function of the kidneys.[5]

A case reviewed by Leersum et al showed abnormalities of the respiratory system. This patient had hyper lucent right lung field resulting in displacement of the heart and mediastinum to the left as a result of the volume increase of the right lung field. Contrast enhanced computerized tomography at the level of the lower lobe showed multiple, medium sized lucent cystic lesions in the right lower lobe. A shift of the mediastinum to the left and compression of the normal left lung were observed. After surgery, histology revealed congenital cystic adenomatoid malformation type 2 coexisting with PBS.

PBS should be differentiated from lesions like Posterior Urethral valve or other causes of ureteral dilatation in infancy and childhood.[6,7]

Treatment is either surgical or 'hands-off' attitude. Surgical treatment may be supravesical diversion with subsequent reconstruction of the ureters, orchidopexy or primary orchidectomy.Some workers advocate surgical reconstruction of the anterior abdominal wall to improve respiratory function and also for cosmetic reasons This baby may have benefited from this surgical repair but his parents imposed the 'hands-off' attitude on him.

Prognosis is not very encouraging in PBS and other congenital renal anomalies. Death occurs commonly from urinary sepsis, renal failure or both. Fertility is not usually easily established in cases that have undescended testis in addition.

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