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CASE REPORT

A RARE FETAL ANOMALY CAUSING OBSTRUCTED LABOUR

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ABSTRACT: Prune belly syndrome is a rare congenital disease. Its incidence being 1 in 30,000 to 1 in 50,000 births. Males are more affected than females. A third of cases are usually stillbirths. Herewith, we present to you a baby who was diagnosed to have prune belly syndrome and its obstructed labour course which we managed by fetal abdominal tapping.
KEYWORDS: Prune belly syndrome, hydronephrosis.

INTRODUCTION: Prune belly syndrome is a rare congenital disease characterized by abdominal wall muscle abnormalities, urinary tract abnormalities and cryptorchidism. It is also associated with cardiovascular, genital, respiratory and other musculoskeletal malformations. The prognosis is usually guarded. The syndrome is named for the mass of wrinkled skin that is usually present on the abdomen of affected baby.

CASE REPORT: Mrs. S.D, 25 years old lady with history of third degree consanguineous marriage, reported to the outpatient department of Obstetrics with history of amenorrhoea for five months. By dates she was 21.6 weeks of gestation. Her obstetric history being, G4 P1+2 with history of one caesarean section done for acute fetal distress, this baby had an unexplained neonatal death at one month of age, followed by 2 spontaneous abortions at 4 months of gestation each.

Ultrasound done revealed a single live fetus in the intrauterine cavity with BPD of 22.3 weeks and FL of 21 weeks. Placenta was posterior and fundal. Severe oligohydramnios was present with massive distension of fetal urinary bladder with bilateral massive hydronephrosis and hydroureter. Fetal ascites was also there. No significant other structural anomaly noted. Diagnosis of prune belly syndrome was made.

The patient was explained about the poor fetal prognosis. She then returned in early labour at 23 weeks, with leaking per vaginum and abdominal pain for four hours. On examination, height of the uterus was 28 weeks. Presentation was footling breech, and fetal heart sounds were present. Patient went to full dilatation, and after the delivery of the lower limbs, patient went into obstructed labour.
After aseptic precautions a spinal needle was introduced through the vagina into the tense fetal abdomen and 1.5 liters of fluid was drained from the fetal abdomen.

The fetus was then easily delivered by assisted breech delivery. The fetus was a male stillborn, with a birth weight of 2.45 kg. Grossly the fetal abdomen was over distended with genital showing a single phallus without scrotal development. Talipes equinovarus was also present. Post-mortem could not be done as patient was not wiling.

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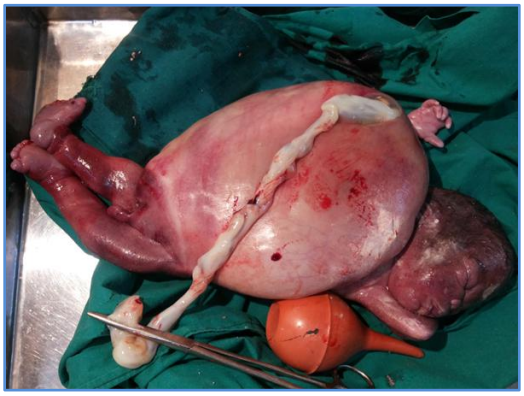

Gross appearance of the fetus with prune belly syndrome

DISCUSSION: Prune belly syndrome is a rare, genetic birth defect affecting about 1 in 30,000 to 1 in 50,000 births and 3.8 in 100,000 live births.[1] It is also known as Eagle Barret Syndrome or Obrinsky syndrome or mesenchymal dysplasia or triad syndrome.[2] About 95% of those affected are males.[3]
Prune belly syndrome is characterized by a triad of symptoms:

1. A partial or complete lack of abdominal muscles. There may be wrinkly folds of skin covering the abdomen like a prune.
2. Undescended testicles in males.
3. Urinary tract abnormalities such as large bilateral hydronephrosis, tortuous dilated ureters, distended urinary bladder and variable degree of renal dysplasia.[2] In addition to classical triad, associated musculoskeletal abnormality, cardiovascular abnormality and genital malformations are noted.[1,2]

ETIOLOGY: The exact cause of this congenital anomaly is not known. Diseases inherited in autosomal recessive pattern appear to occur at higher frequency in consanguineous marriages, additional evidence for this inheritance pattern is suggested[6]. But it has recently been suggested that it is a 2 step autosomal dominant mutation with sex-linked expressions, partially mimicking X-linkages[7]. Few sporadic cases are also reported.[6]

An alternate theory states that, the infra-vesical obstruction leads to massive distension of urethra, distension of bladder and hydronephrosis. Bladder distension leads to atrophy of abdominal wall muscles due to venous infarction and prevents descent of testes into scrotum[8].Another theory of mesodermal arrest states that the defect exists in the mesoderm of anterior abdominal wall and urinary tract during 6th to 10th weeks of pregnancy.[9]

CLINCAL FEATURES: Abdominal wall skin wrinkling is secondary to abdominal muscle hypoplasia[2]. The bladder in patients with the prune belly syndrome is typically enlarged and thickened but trabeculations are usually absent. This is in sharp contrast to the bladder of a patient with posterior urethral valves, which is usually markedly trabeculated[10]. There is variable degree of renal dysplasia with hydronephrosis and hydroureter.[11]

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When there is severe obstructive uropathy due to urinary malformations it is associated with pulmonary hypoplasia and oligohydramnios[4], as seen in our case. About one third of infants are stillborns or die within few months of delivery due to pulmonary complications and renal complications. As many as 30% develop end stage renal disease and require renal transplant[11]. Other complications include urinary tract infections due to the inability to properly expel urine which can lead to septicemia. Later in life, a common symptom is post-ejaculatory discomfort.[12]

Bilateral cryptorchidism is considered a hallmark of this syndrome, with the majority of these testes being intra-abdominal above the level of the iliac vessels. Histologically these testes also demonstrate a variety of abnormalities. Children with prune belly syndrome must be closely monitored for the development of testicular tumors.[13]

Cardiac abnormalities have been reported to occur in 10% of prune belly patients. Associated cardiac findings include ventricular and atrial septal defects, patent ductus arteriosus and tetralogy of Fallot. Gastrointestinal anomalies appear to occur in up to 30% of affected prune belly children. Abnormalities such as malrotation, atresia, stenosis and volvulus appear to result secondary to persistence of the embryonic mesentery. Orthopedic abnormalities occur in 45-63% of prune belly patients. The most frequent anomalies are hip dislocation and talipes equinovarus.[5]

DIAGNOSIS: Prune belly syndrome can be diagnosed antenatally by ultrasound. An abnormally large abdominal cavity as the abdomen swells with the pressure of accumulated urine. Sonographic findings include oligohydroamnios or anhydramnios, hydroureter, hydronephrosis, a distended urinary bladder and a thin, attenuated abdominal wall.[14]

In newborn, the prune like abdomen usually leads to the diagnosis. Kidney function tests, chest X ray, renal ultrasound, voiding cystourethrogram and intravenous pyelography are diagnostic tests[14]. During a cystogram, the bladder is typically smooth walled due to absence of trabeculae and may demonstrate an urachal remnant or diverticulum.[10]

TREATMENT: In the patients, who survive, surgery is often required but it will not return the organs to a normal size. Reconstruction surgeries include abdominoplasty, urinary tract reconstruction and orchidopexy[15]. Bladder reductions have shown that the bladder will again stretch to its previous size due to lack of muscle. The type of treatment, like that of most disorders, depends on the severity of the symptoms. Cutaneous vesicostomy allows the bladder to drain through a small hole in the abdomen, thus helping to prevent urinary tract infections.

The diverticulum or patent urachus, if present may be excised at the time of vesicostomy. Supravesical diversions such as ureterostomy, pyelostomy and nephrostomy are indicated only if cutaneous vesicostomy is not effective in decompressing urinary system. Even with treatment, many patients experience renal failure and may require dialysis or renal transplant.[15] In case of mild disease self-catheterization, often several times per day, can be an effective approach in preventing infections.[16] Antibiotics are to be given to treat infections.

Diagnosis of prune belly syndrome necessitates thorough orthopedic and cardiological evaluation and treatment because of the high prevalence of associated musculoskeletal and cardiac abnormalities.[2,15] Long-term follow up is required which includes regular monitoring with renal function tests, ultrasonography, sexual functioning and urodynamic studies.[5,15]

PREVENTION: The routine use of screening for fetal anomalies has resulted in more affected pregnancies being terminated.[14] If an antenatal diagnosis of urinary obstruction is made, it may be possible to perform vesicoamniotic shunting and intrauterine surgery to prevent the development of prune belly syndrome. The results seem promising.[15,16]

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