Case Report

Prune belly syndrome: A rare case report of ultrasound-guided prenatal diagnosis

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Abstract

Background: Prune belly syndrome is described as a rare congenital anomaly with an uncertain aetiology. It is characterised by abnormalities like deficiency in abdominal muscles, urinary tract abnormalities and also there is bilateral cryptorchidism in males. This case report describes the early diagnosis of Prune belly syndrome by ultrasonography.
Conclusion: This case report highlights the importance of ultrasonography in identifying and documenting congenital abnormalities in early antenatal period.
Keywords: Bilateral cryptorchidism, Urinary tract abnormalities, Congenital anomalies

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INTRODUCTION

Prune belly syndrome was first described in the year 1839 by Frolich[1]. It is synonymous with Eagle-Barrett syndrome[2] and also known as Obrinsky syndrome[3]. It is a rare congenital disorder which affects about 1 in 30,000 births[4] and 96% among the affected are male subjects. The characteristics include bilateral cryptorchidism, deficient development of the abdominal muscles which causes prune like wrinkling of the abdominal skin and also urinary tract abnormalities like bilateral and gross hydronephrosis, megacystitis and megaureter[5]. The exact cause is not known but some studies have revealed that there is a chance of genetic inheritance and maybe an association with genetic abnormalities like trisomy 18 and trisomy 21[5,6]. The prognosis of Prune belly syndrome is reported to be poor with early infant deaths and stillbirths being very common[7]. This case report describes the early diagnosis of Prune belly syndrome by ultrasonography.

CASE PRESENTATION

A 22 years old married female, gravida 1, para 0 with history of 4 months of amenorrhea came for regular antenatal visit. She had no significant past medical history or family history. There was no history of consanguineous marriage. On examination, general condition was fair, per abdomen examination revealed uterus size corresponding to 16 weeks with relaxed abdomen and external ballotment present. Haemoglobin level was 7.9g/dl and other routine blood and urine reports were within physiological limits. Ultrasonography scan revealed single live intrauterine pregnancy with a changing lie, adequate amniotic fluid, fetal heart activity was present, placental site was anterior with maturity Grade 0. Fetal growth parameter of Bi parietal diameter (BPD) was 3.6cms suggesting 17wks 01day gestational age. Congenital anomalies noted were deficient anterior abdominal wall, large bladder extrophy, mild hydronephrosis, defect in lower spine and fused lower limb. Repeat ultrasonography scan was done and it showed similar findings. Neonatologist opinion was taken regarding the prognosis of baby; he mentioned that as it has a poor outcome, hence termination of pregnancy can be suggested. Also, as this condition does not have a predictable risk of recurrence; hence future pregnancy is reported to be usually safe. Following counselling, informed and written consent was taken from the couple for termination of pregnancy. Patient received one unit of blood. Induction with misoprostone 100ug was done. On 24th December 2014 she aborted a male baby of 125g

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along with placenta of 115g per vaginally. Patient refused for further examination, namely karyotyping and autopsy. Macroscopic examination of the baby showed absent anterior abdominal wall, large bladder extrophy, fused lower limb and undescended testis which further confirmed the diagnosis of Prune belly syndrome.

DISCUSSION

The present case showed the classical presentation of Prune belly syndrome. However, it has been reported to be associated with a variety of defects including cardiovascular, pulmonary, musculoskeletal and genital malformations[8]. When the urinary tract abnormality is associated with a severe obstructive uropathy, Prune belly syndrome can lead to oligohydramnios and also pulmonary hypoplasia[9]. Prune belly syndrome's pathogenesis is not clearly known. There is a mesodermal defect theory which suggests that there exists a defect in mesoderm of urinary tract and anterior abdominal wall. Aberrant development of derivatives of the first lumbar myotome between 6 and 10 weeks of gestation leads to a patchy muscular deficiency or hypoplasia of the abdominal wall and also causes urinary tract abnormalities[10]. There is an alternate theory about the urethral obstruction malformation complex which proposes that there occurs pressure atrophy of muscles of abdominal wall when there is urethral obstruction leading to massive distension of bladder as well as ureters. Descent of the testes may also be affected by bladder distension and lead to bilateral cryptorchidism[11]. There is comparatively a very high incidence of Prune belly syndrome in males which can be explained by the more complex morphogenesis of male urethra, possibly leading to obstructive anomalies at several levels[9]. In the present case, ultrasound sonography helped in diagnosis and after counselling and consent of parents as well as neonatologist opinion, termination of pregnancy was done. Ultrasound, plain X-ray, and intravenous pyelogram have been reported to be very useful investigations to diagnose the condition. Although many ethical questions are raised when innovative fetal therapy is discussed, the insults that result from urinary tract obstruction often lead to stillbirth or neonatal death. Many infants are either stillborn or die within the first few weeks of life from severe lung or kidney problems, or a combination of congenital anomalies[9].

CONCLUSION

This case report highlights the importance of ultrasonography in identifying and documenting congenital abnormalities in early antenatal period. Women undergoing serial antenatal ultrasound examinations must be carefully counselled regarding the purpose of the scan. The increasing use of obstetric ultrasound will inevitably result in a rise in prenatal detection of congenital abnormalities. This need is to be met with adequate training, referral services and better knowledge of women's attitude and beliefs on birth defects and ultrasonography.

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