Complete penoscrotal transposition

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Introduction
Penoscrotal transposition is an extremely rare congenital anomaly of the external genitalia, characterized by malposition of the penis in relation to the scrotum. In complete transposition, the scrotum covers the penis which emerges from the perineum. In incomplete transposition, which is more common, the penis lies in the middle of the scrotum. Both forms are most often associated with a wide variety of other anomalies [1]. We describe a case of a newborn with CPST, but normal upper urinary tract and other associated malformations.

Case report
The infant weighing 2.4 kg, was born at term, by vaginal route. He was asphyxiated at birth. The mother of the baby, 22 years of age, non-consanguineously married, had a previous normal childbirth and had no significant antenatal problem. No teratogen exposure was there. The antenatal ultrasound had missed the anomaly and the child was noted to have complete inversion of external genitalia at birth. Scrotum was well formed, blackish in colour with rugosities present all over. Both the testes were palpable in the scrotal sac. The penis was placed just anterior to the anal opening which itself was displaced anteriorly. (Figure A & B). Rectal atresia was there. There was no hypospadias or chordee. A diagnosis of complete form of penoscrotal transposition with ectopic anus was made. The child was evaluated for other associated anomalies. He had depressed nasal bridge with low set ears. Cranium and spine were normal except the presence of a pilonidal sinus overlying coccyx. Left foot was of rocker bottom type (Fig C). Echocardiography revealed a ventricular septal defect (VSD). Other investigations like...
Figure A, B and C: Complete penoscrotal transposition (A,B); deformed facial feature along with rocker bottom type left foot anomaly (C).

cranial and abdominal USG were normal. Unfortunately, the baby died 5 hours after birth due to effects of perinatal asphyxia, in spite of standard treatment provided.

Discussion

CPST is a very uncommon heterogeneous condition in which the scrotum is positioned superior and anterior to the penis. Less than 20 cases of CPST with normal scrotum have been reported in the literature [2]. CPST is frequently associated with major and often life-threatening malformations. Parida et al [3] had noted major renal anomalies in the form of agenesis, horseshoe kidney, ectopic and dysplastic kidney, obstructive uropathy and hydronephrosis. Other systemic abnormalities are mental retardation, anorectal malformations, central nervous system, skeletal and cardiological defects. The embryological explanation of this condition remains elusive. It is postulated that abnormal positioning of the genital tubercle in relation to the scrotal swellings during fourth to fifth week of gestation could affect the migration of the scrotal swellings [4]. Another theory suggests that the phallic tubercle is intrinsically abnormal and affects the corpora’s development, which explains the abnormal penis and frequent occurrences of the other genital abnormalities [1]. Kain et al. [5] proposed that abnormality in the gubernaculum could also result in defective migration of the labio-scrotal folds. It should be differentiated from the conditions like pseudohermaphroditism, penoscrotal hypospadias, micropenis, intrauterine penile amputation, and specially penile agenesis with a midline skin tag anterior to anus.

In presence of normal penis this anomaly does not cause any sexual dysfunction. However, its surgical repair is done for psychological reasons. It is performed usually between 12–18 months [6]. The basic principle is displacement of scrotal skin posteriorly and the penis anteriorly. McLvoy and Harris first performed surgery to move the penis into a more cranial position through a subcutaneous tunnel beneath the prepenile scrotum [7]. Forshall and Rickham used a different technique where cranially located scrotal flaps were elevated, rotated medially and caudally and sutured beneath the penis [8]. This technique was also used by Glenn and Anderson. To repair CPST we here use a circumferential incision at the base of the penis. This incision then extended vertically at 12 O’clock in the midline between the fused scrotal folds. The vertical incision was stopped at a point where the penis has to be transposed. A racket shaped incision was made laterally, either side from 12 O’clock position to outline the scrotal folds. Thereafter these incisions were deepened to release the abnormal
soft tissue bands. Once fully mobilised, the scrotal folds are moved caudally and penis move cranially to its normal position. Soft tissue and skin approximation were made thereafter [9]. Complications after surgery include urethral and testicular injury, urinary fistula, flap necrosis and penile oedema [10].

This case being extremely rare is projected to add on to the existing literature and to emphasize that a search for other anomalies should be undertaken before subjecting them to corrective surgeries.

References: