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A RARE CASE OF PENO-SCROTAL HYPOSPADIAS WITH PENO-SCROTAL TRANSPOSITION ASSOCIATED WITH ACCESSORY SCROTUM CONTAINING PERIANAL LIPOMA.

Urology

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ABSTRACT

Hypospadias is a common condition in children and is rarely associated with testicular/scrotal anomalies. But a congenital scrotal anomaly associated with accessory scrotum with perineal lipoma is extremely rare. A two year old boy presented to our outdoor patient department with peno-scrotal hypospadias with severe chordee, associated with peno-scrotal transposition and bifid scrotum, and an accessory scrotum with perineal lipoma. After admission other congenital anomalies were ruled out. Single stage surgical correction was undertaken. Skin chordee was corrected by only degloving of the penis. Then Ducket's repair was performed for correction of hypospadias, followed by Glen-Anderson technique for repair of peno-scrotal transposition. Finally wide local excision of the perineal lipoma was done. Post-operative period was uneventful and results were satisfactory.

KEYWORDS

Hypospadias, peno-scrotal transposition, bifid scrotum, accessory scrotum, perineal lipoma

INTRODUCTION

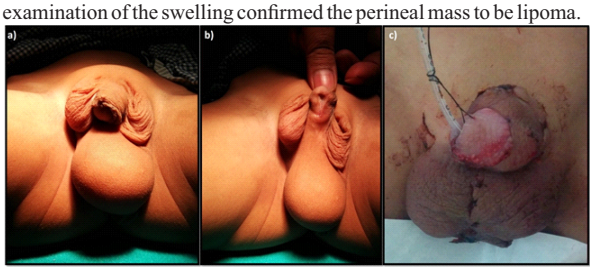
Congenital scrotal anomalies are very rare. Classically, they are of four types: bifid scrotum, peno-scrotal transposition, ectopic scrotum and accessory scrotum. Among these accessory scrotum is the least frequent. So far, only 43 cases of accessory scrotum have been reported in literature [1]. But accessory scrotum associated with penoscrotal hypospadias is even rarer. They are more commonly seen to occur with anorectal malformation.[2] Only 4 cases have been reported so far [1]. We present a very interesting and extremely rare case of penoscrotal hypospadias with associated chordee, bifid scrotum, penoscrotal transposition. The patient was also carrying an accessory scrotum which on further investigation turned out to be a perineal lipoma.

CASE REPORT

A two year old boy was referred in the pediatric surgery department from a local city hospital. The mother complained that her boy had difficulty in micturition since birth and had a swelling in groin. Upon physical examination, the patient had dorsal hood of the penis with severe chordee and peno-scrotal hypospadias. There were two scrotal sacs on either side of penis which was a typical bifid scrotum along with penoscrotal transposition. Both the testis could be palpated in the scrotal sacs. There was also a soft peduncular mass in the perineum. It was approximately 4 x 3 cms in size (figure 1a, b).

The patient was then admitted and further investigated. Ultrasonography of inguinal and perineal region revealed normal testis. The pedunculated mass in perineum gave hyper echoic shadow suggestive of lipoma. Other investigations included ultrasonography of abdomen and echocardiography to rule-out any congenital anomaly. No other associated anomaly was detected and routine blood investigations were within normal ranges.

The pre-operative diagnosis of penoscrotal hypospadias with scrotal transposition with accessory scrotum was made and patient was planned for single stage reconstructive surgery. Transverse island preputial flap repair (Ducket's repair) was done for correction of hypospadias. Glenn Anderson technique was undertaken for correction of penoscrotal transposition (figure 1c). Perineal mass was excised. Post-operative period was uneventful. The histopathological examination of the swelling confirmed the perineal mass to be lipoma.



DISCUSSION

Congenital scrotal anomalies are usually classified into four type's namely bifid scrotum, penoscrotal transposition, ectopic scrotum, and accessory scrotum. Though congenital scrotal anomalies are rare occurrences, bifid scrotum and penoscrotal transposition is relatively more common than accessory scrotum which is very infrequent. Development of scrotum starts at 4th week of gestation. The two labioscrotal swellings fuse to form scrotum. Early division or abnormal migration of labioscrotal swellings can be related to the etiology of congenital anomaly of scrotum[3]. While unilateral abnormal migration of a labioscrotal swelling results in unilateral penoscrotal transposition or ectopic scrotum whereas early division with subsequent abnormal migration of a labioscrotal swelling result in an accessory scrotum. Simultaneous development of a perineal lipoma may result in development of bifid scrotum and penoscrotal transposition by interrupting the normal migration of the labioscrotal swelling[3].

In Bifid scrotum, the hemiscrotum is normally positioned with its partial or complete separation, seen in patients with severe hypospadias or chordee. When a part or whole of scrotum is located superior to the penile shaft it's termed as penoscrotal transposition. Ectopic positioning of the scrotum is called as ectopic scrotum. In most of the cases it is unilateral, with the ectopic tissue lying either suprainguinal (most common), infrainguinal (femoral) or on the thigh (least frequent). The ipsilateral testis is usually present within the ectopic hemiscrotum[4]. If there is additional ectopic scrotal tissue present apart from normally developed scrotum, the entity is termed as accessory scrotum. Number of congenital abnormalities such as hypospadias, diphallia, defects of scrotal position, anorectal anomalies, and the VACTERL (vertebral, anal, cardiac, tracheoesophageal, renal, and limb anomalies) can be associated with accessory perineal scrotum. Other anomalies such as juxtapose subcutaneous tumour has a significant incidence of nearly 72.5% association with assessory scrotum. Histologically, one case of subcutaneous tumour was lipoblastoma[5]. The differential diagnosis of perineal mass includes lipoma, lipoblastoma, infantile hemangioma, hamartoma and choristoma[6]. While in accessory scrotum skin rugosity is present whereas a perineal lipoma lacks the same which easily differentiates them clinically.

The four such reported cases were detected prenatally and most of them were detected after birth.[1] The prognosis of surgically treated patients are good, and only one has died from an associated anomaly before surgery.[7]

The etiology of the case in question seems to be due to interruption of developing labioscrotal swelling by intervening mesenchymal tissue (lipoma). As, all other organ systems are normal viz. musculoskeletal, cardiac, central nervous system implies that the causative factor did not have any adverse effect on differentiation of these organ system which develop at the same embryological time period .

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