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Case Report

Perineal lipoma mimicking an accessory penis with scrotum

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ABSTRACT

A case of accessory penis with scrotum in a 4 months old boy is reported because of its rarity. The infant presented with a tumour mimicking an accessory penis with scrotum between the normal sited scrotum and anus. Both testes had descended into the scrotum. After complete evaluation, there was no other urological anomaly. The tumour was excised and the histo-pathological findings of the tumor indicated a perineal lipoma. An overview of normal development of male external genitalia has been provided and the deranged mechanism resulting in this anomaly has been reviewed with hypothesis regarding etiology of accessory scrotum.

Keywords: Accessory penis, Accessory scrotum, Congenital urogenital deformities, Perineal lipoma

INTRODUCTION

Accessory scrotum is considered the rarest of all congenital scrotal abnormalities.[1] In accessory scrotum, in addition to a normally developed scrotum, ectopic scrotal tissue is present either in the perineum or elsewhere, without the presence of testis within it.[2,3] Accessory scrotum has been observed in isolation or in conjunction with other anorectal/urogenital abnormalities.[1] An interesting case of accessory penis with scrotum in the perineum with a perineal lipoma is reported with the relevant embryological basis for this condition, and hypotheses for the development of an accessory penis with scrotum have been discussed.

CASE REPORT

We report a case of a 4 months old male infant, born to a 28 year old mother from non-consanguineous marriage at full term. The boy presented with an abnormal mass in the perineum which was present since birth. Physical examination of the genitalia showed a normal penis with scrotum containing both testis appearing normal on palpation in each hemi-scrotum. Another mass of size 3 cm x 1.5 cm was situated between the normally sited scrotum and the anal orifice. Attached to the mass superiorly was an appendage of size 1.5 x 1 cm covered with skin showing rugosity mimicking an accessory penis with scrotum. The mass and its appendage were soft in consistency and were freely mobile over the perineum. The infant underwent radiological evaluation preoperatively and no urinary or ano-rectal abnormality was demonstrated (Figure 1).

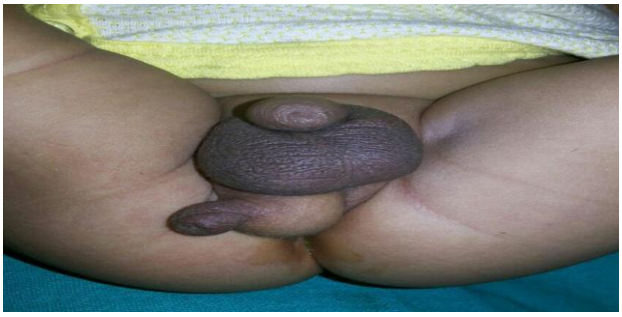

Figure 1: Presentation.

The mass was excised under general anaesthesia. Post-operative period was uneventful. The histological examination of the excised specimen was suggestive of a lipoma. The patient recovered well and was discharged with no clinical complaints. On Follow up after 3 months, patient was asymptomatic with a healthy scar and normal bladder and bowel habits (Figure 2-6).

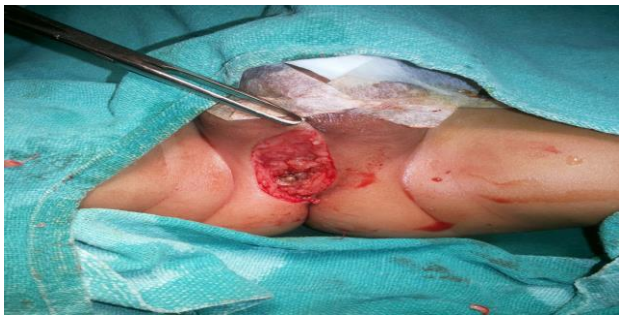

Figure 2: After excision.

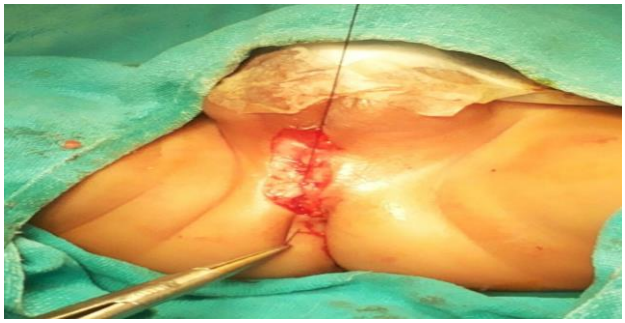

Figure 3: Subcutaneous closure using absorbable sutures.

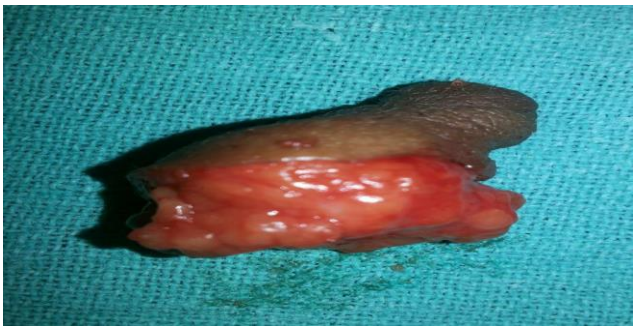

Figure 4: Excised specimen.

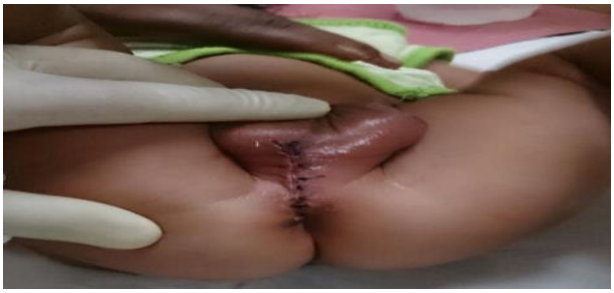

Figure 5: Post-op day 5: suture removal.

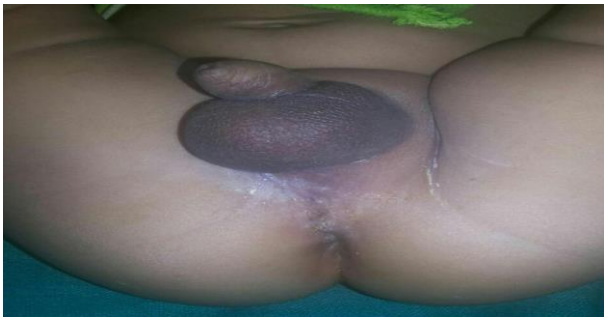

Figure 6: Scar on follow-up after 2 weeks.

DISCUSSION

Isolated congenital perineal lipomas are rare lesions that can lead to a misdiagnosis of an accessory scrotum in men, in particular because these two conditions can be associated in over 80% of cases.[4,5] The differential diagnosis is mainly an accessory scrotum, sacrococcygeal teratoma, fetus in fetu, haemangioma, haemartoma or lipoblastoma.[5]

A complete evaluation of the urogenital and anorectal tract is recommended, taking into account related anomalies described such as renal agenesis, anorectal malformations, scrotum and penile anomalies.[4-6]

About 30 cases have been reported in the literature either being solitary or in association with other urogenital or nonurogenital abnormalities.[7]

Accessory scrotum is an extremely rare abnormality. Congenital scrotal anomalies are conventionally classified into four types: bifid scrotum, penoscrotal transposition, ectopic scrotum, and accessory scrotum. Bifid scrotum is a partial or complete separation of otherwise normally positioned hemiscrotum in patients with severe hypospadias or chordee. In cases of penoscrotal transposition, part or whole of the scrotum is located superior to the penile shaft. Ectopic scrotum is ectopic positioning of the scrotum which is usually unilateral, with the ectopic tissue usually suprainguinal, but in some cases infrainguinal (femoral) or on the thigh. The ipsilateral testis is usually present within the ectopic hemiscrotum. In accessory scrotum, in addition to a normally developed scrotum, ectopic scrotal tissue is present either in the perineum or elsewhere, without the presence of testis within it.[7]

In the absence of a perineal lipoma, accessory perineal scrota are usually associated with other anomalies, including hypospadias, diphallia, defects of scrotal position, anorectal anomalies, and the VACTERL (vertebral, anal, cardiac, tracheoesophageal, renal, and limb anomalies) association.[8]

Male external genital development depends on the conversion of testosterone to the more active dihydrotestosterone and its subsequent action via tissue

receptors. The genital tubercle enlarges into the penis. The scrotum forms from the labioscrotal swellings at fourth week of gestation. The labioscrotal swellings migrate inferomedially and merge at 12 weeks of gestation to form the scrotum, with the line of fusion being the scrotal raphe.[9]

The etiology for an accessory scrotum is not known. There are two common hypotheses for the development of an accessory scrotum. Lamm and Kaplan postulated that one labioscrotal swelling may embryologically divide into two portions with the inferior portion migrating incompletely to form an accessory scrotum.[10] According to Sule et al the accessory labioscrotal fold usually develops due to intervening mesenchymal tissue disrupting the continuity of developing labioscrotal swelling.[11]

Hence Accessory Penis with Scrotum are essentially isolated lesions, which are usually benign and the standard treatment is local excision.[5]

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