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Case Report

Accessory Scrotum: A Note on Nomenclature, Diagnostic Criteria and Classification

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Accessory scrotum
Scrotal malformation
Perineal hamartoma
Perianal lipoma

Abbreviation
LSF - Labio-scrotal fold

Abstract

A newborn with pedunculated perianal lipoma and accessory scrotum is reported. By reviewing the 60 cases of accessory scrotum documented in the literature, we propose a standardized definition, diagnostic criteria and clinico-embryological classification of this rare malformation.

CLINICAL DESCRIPTION

A full-term male newborn presented at birth with asymptomatic soft pedunculated mass hanging from the anal margin. The mass was bilobed; one of which was a 2x3 cm soft tissue mass covered with smooth normal colored skin, while the other lobe was an empty sac of dark pigmented wrinkled skin resembling that of scrotum.(Fig 1) There were no other congenital anomalies as revealed by clinical examination and screening ultrasonography. The mass was completely excised under caudal analgesia. Post-operative recovery and healing was uneventful.

Histologically, the two lobes differed significantly; one of which showed smooth muscle bundles (dartos) dispersed in dermal collagen (Fig 2) while the other lobe showed abundant mature adipose tissue in deep dermis.

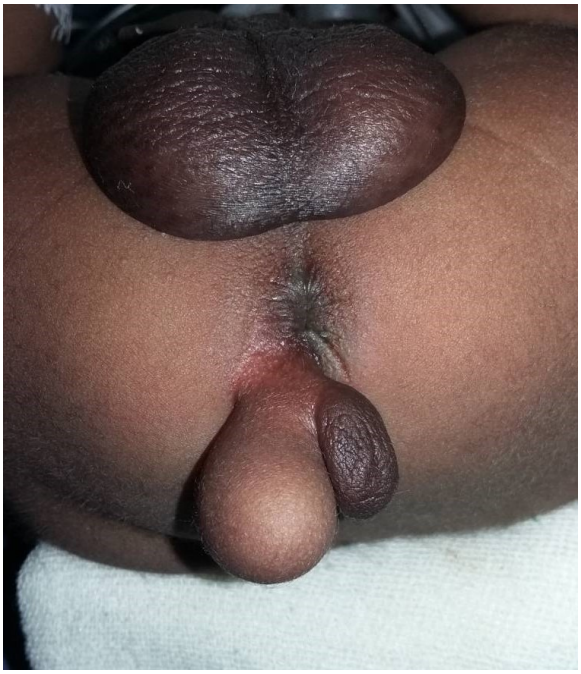

Fig 1. *Clinical photograph showing pedunculated peri-anal mass. The bilobed mass shows lipomatous and scrotal components. Normal scrotum is also seen in addition to accessory scrotum.*

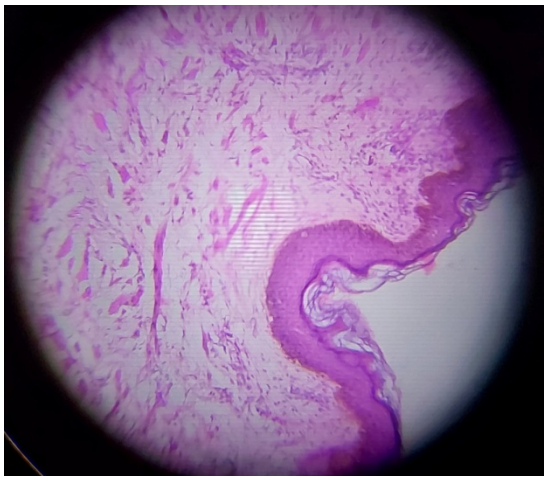

Fig 2. *Histopathology of the accessory scrotum showing skin layers with dartos muscle underneath. (Hematoxylin and Eosin staining; 100X magnification)*

DISCUSSION

Accessory scrotum is an extremely rare malformation with fewer than 60 cases been recorded in the literature.[1,2] It is defined as ectopically located supernumerary scrotal sac sans testis. It is usually located posterior to the normally formed orthotopic scrotum (e.g. perineum or perianal margin).[3] Only 3 cases of accessory scrotum at other locations (e.g. groin, pubis and penile shaft) have been described.[4] Nearly 80% of them are associated with lipomatous perineal hamartoma.[2,5]

'Ectopic scrotum' is yet another rare anomaly that is defined as ectopically located hemi-scrotum containing ipsilateral testis.[3] Absence of hemi scrotum at orthotopic location is characteristic of this condition. Unlike accessory scrotum, they are located anterior to native scrotum (e.g. groin, pubis, upper thigh)[6] and are not associated with lipomatous hamartoma.

As ectopic location is common to both, 'accessory scrota' have been misreported as 'ectopic scrota' and vice versa in the literature.[7] Diagnostic labeling of an abnormal perineal tissue as 'accessory scrotum' may sometimes be difficult if the skin wrinkles are less prominent or if it is just a tiny skin tag. To avoid confusion in nomenclature and clinical labeling, we propose a set of diagnostic criteria that distinguishes 'accessory scrotum' from 'ectopic scrotum' and other hamartomatous lesions. (Box 1)

High incidence of coexisting perineal tumors (71% perineal lipoma, 6% lipoblastoma, 3% mixed hamartoma) in accessory scrotum is thought to be of embryopathogenic significance.[8] Sule proposed that lipomatous overgrowth (hamar-toma) of caudal mesenchyme in a 4-12-week-old embryo might disrupt the continuity of developing labioscrotal folds (LSF) which are destined to become future scrotum.[9] The detached LSF might then develop into accessory scrotum, while the native LSF continues to develop into normal orthotopic scrotum. Such mechanical disruption is also possible in lower limb malformations wherein fetal heel presses upon the developing LSF.[10]

Interestingly, none of the accessory scrota, that are associated with perineal lipoma or tibial malformations, had coexisting anomalies. In contrast to this, accessory scrota without perineal

Box 1: Diagnostic criteria of accessory scrotum

Major criteria
1. Presence of normal bipartite orthotopic scrotum in addition to ectopic scrotal tissue
2. Absence of testicular tissue (macroscopic or microscopic) within the ectopic scrotal sac
Minor criteria
1. Ectopic tissue with wrinkled dark pigmented skin macroscopically resembling a scrotum
2. Ectopic tissue with concentration of androgen receptors similar to that of scrotum
3. Ectopic tissue being located posterior to the orthotopic scrotum
4. Histological demonstration of dartos smooth muscle within the aberrant tissue
5. Bipartite / bilobed morphology of ectopic tissue analogous to that of normal scrotum
Diagnosis of accessory scrotum necessitates **both** major criteria **plus** any two of the minor criteria.

lipoma often had serious malformations of genitourinary system, anus and other internal organs. Thus, there seems to be three different embyo-pathgenic types of accessory scrota:

Type 1: Typical accessory scrotum associated with perineal tumors (e.g. Perineal lipoma, hamartoma, lipoblastoma) or lower limb deformities (e.g. Tibial aplasia). They are probably due to mechanical disruption of the developing labio-scrotal fold. (Sule's theory)[9]

Type 2: Typical accessory scrotum not associated with perineal mass. They are probably due to defective regional organizer cells of embryonic perineum. The resultant field defects may include malformations of caudal end derivative such as genitourinary tract and anorectum. Any one of the following mechanism may play a role in its embryogenesis: (i) Aborted attempt of caudal duplication (Lamm-Kaplan's theory)[7], (ii) triple primordial anlage of the labioscrotal swelling (Takayasu's theory)[11], (iii) Abnormal division of LSF and posterior migration of the detached segment (Coupris-Bondonny's theory)[12]

Type 3: Atypical accessory scrotum that occurs as a part of syndromic genetic mutations. Distant malformations such as cleft lip and heart disease are common in this type and the accessory tissue may be located anterior to native scrotum.

Several attempts have been made to classify accessory scrota. Park[6] classified associated lipoma into protruding (sessile) or peduncular. He opined that the former type supports Sule's theory while the latter one supports Takayasu's theory. Ratan[13] classified it into 3 types: Type A - accessory scrotum presenting as a tag of rugose skin over a lipoma; Type B - accessory scrotum presenting as a well developed sac; Type C - accessory scrotum associated with pseudo-duplication of external geintalia. Hoar[14] classified it based on anatomical location such as supra-inguinal, femoral, penoscrotal and perineal. He found that femoral type was more associated with distant anomalies like cleft lip; suprainguinal type was more associated with other genitourinary anomalies while perineal type wass an isolated malformation. Amman[15] classified the clinical morphology of accessory scrotal tissue into 3 types: ill defined (faint) rugacity, tiny skin tag (or nodule) and well developed rugose skin. Kumoro[16] classified it into mid-perineum type (satisfying Takayasu's theory) and the lateral type (satisfying Lamm-Kaplan's theory). These classifications lack practical significance, while our classification is clinically applicable. For example, in accessory scrota associated with perineal lipoma (type-1) elaborate investigations are not necessary to rule out coexisting anomalies, while type-2 requires imaging studies of genitourinary tract and type-3 requires detailed genetic work-up.

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