Turkish Journal of Pediatric Surgery 2025;39(2):99-102
Doi: 10.62114/JTAPS.2025.57

Turkish Journal of Pediatric Surgery

Case Report

OPEN ACCESS

Accessory scrotum: A rare congenital anomaly

Duygu Tatli Ucarci[1], Y. Doruk Bilgili[2], Burçin Acuner[3], Filiz Kutlu[4], B. Haluk Güvenç[1]

The scrotum forms by migration of the labioscrotal folds inferomedial to the genital tubercle. Failure of migration, possibly related to a gubernacular defect, results in scrotal anomalies such as penoscrotal transposition, accessory scrotum, scrotal ectopia, or bifid scrotum. The accessory scrotum clinically presents as an ectopic scrotal tissue, either in the perineum or elsewhere without the presence of a testis within it, in addition to a normally developed scrotum.[1] Associations with hypospadias and chordee are common. It is reported as the rarest congenital anomaly of the scrotum with approximately over 50 cases in the English literature.[2] Although reported as an extremely rare anomaly, we herein describe three consecutive cases treated in a territory hospital in the Western Black Sea region during the last two years.

Abstract

The accessory scrotum is the rarest form among the four types of congenital scrotal anomalies. The clinical presentation outlines the presence of scrotal skin outside its normal location, with no testis in it. Three patients (an eight-day-old, a 20-day-old, and a 111-day -old) attended our clinic with a patch of rugosity (1×1 cm, 6×4 cm, and 2.5×1.5 cm in size, respectively), mimicking scrotal tissue. In two cases, a perineal patch with rugosity was found overlying a mass resembling a lipoma. One patient presented with bilateral inguinal hernia and a patch of rugosity in the left inguinal region, with no underlying palpable swelling. Abdominal and regional ultrasonography did not reveal any abnormalities. Total excision was possible for all lesions, one of which needed an inguinal rotational flap, following bilateral herniotomy and high ligation. The postoperative course was uneventful. Histological investigation revealed rugose epidermidis, rudimentary dartos fibers, and hair follicles, confirming accessory scrotum.

Keywords: Accessory scrotum, congenital anomaly, neonate.

CASE REPORT

Case 1– An eight-day-old infant presented with bilateral inguinal hernia and patch of rugosity (1×1 cm in size), mimicking scrotal tissue in the left inguinal region, with no underlying palpable swelling (Figure 1). The tissue mentioned was completely resected with an inguinal rotational flap, followed by routine bilateral herniotomy and high ligation.

Case 2– A 20-day-old infant with a perineal patch of rugosity (6×4 cm in size) overlying a mass resembling a lipoma and mimicking scrotal tissue on the right side of the lipoma was admitted to our clinic (Figure 2). Total excision was performed during surgery.

Case 3– A 111-day-old infant with a perineal patch of rugosity (2.5×1.5 cm in size) overlying

Received: August 01, 2024
Accepted: January 02, 2025
Published online: August 11, 2025

Correspondence: Y. Doruk Bilgili, MD.

E-mail: y.doruk.bilgili@gmail.com

[1]Department of Pediatric Surgery, Zonguldak Bülent Ecevit University Health Application and Research Center, Zonguldak, Türkiye

[2]Department of Pediatric Surgery, Bandırma Onyedi Eylül University, Balıkesir, Türkiye

[3]Department of Plastic and Reconstructive Surgery, Zonguldak Bülent Ecevit University Health Application and Research Center, Zonguldak, Türkiye

[4]Department of Pathology, Zonguldak Bülent Ecevit University Health Application and Research Center, Zonguldak, Türkiye

• *Presented as a poster at the 7th WOFAPS Congress, held on October 12-15, 2022, in Prague, Czech Republic.*

Citation:

Tatli Ucarci D, Bilgili YD, Acuner B, Kutlu F, Güvenç BH. Accessory scrotum: A rare congenital anomaly. Turkish J Ped Surg 2025;39(2):99-102. doi: 10.62114/JTAPS.2025.57.

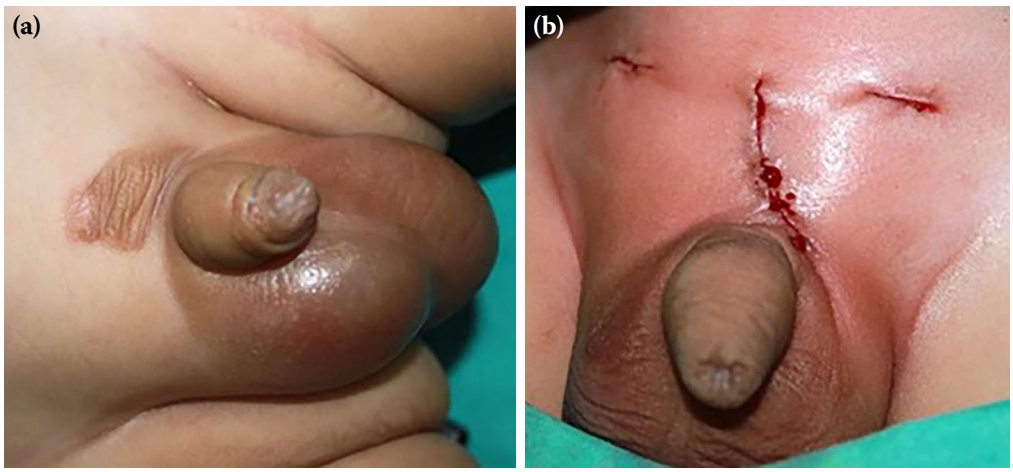

Figure 1. Preoperative **(a)** and postoperative **(b)** photos of Case 1.

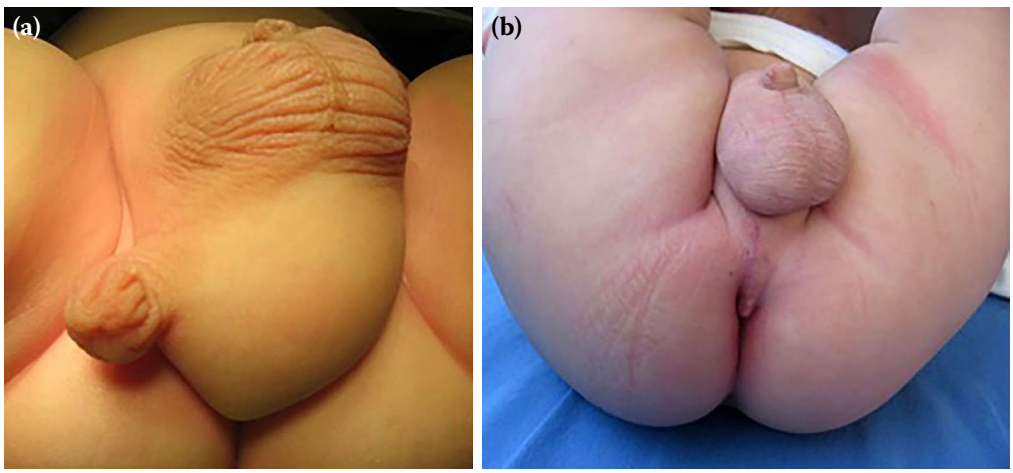

Figure 2. Preoperative **(a)** and postoperative **(b)** photos of Case 2.

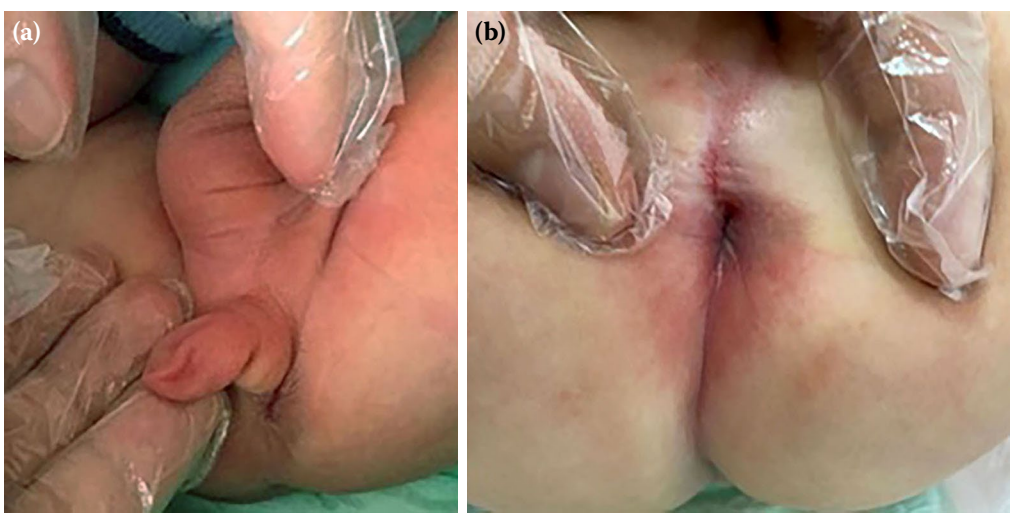

Figure 3. Preoperative **(a)** and postoperative **(b)** photos of Case 3.

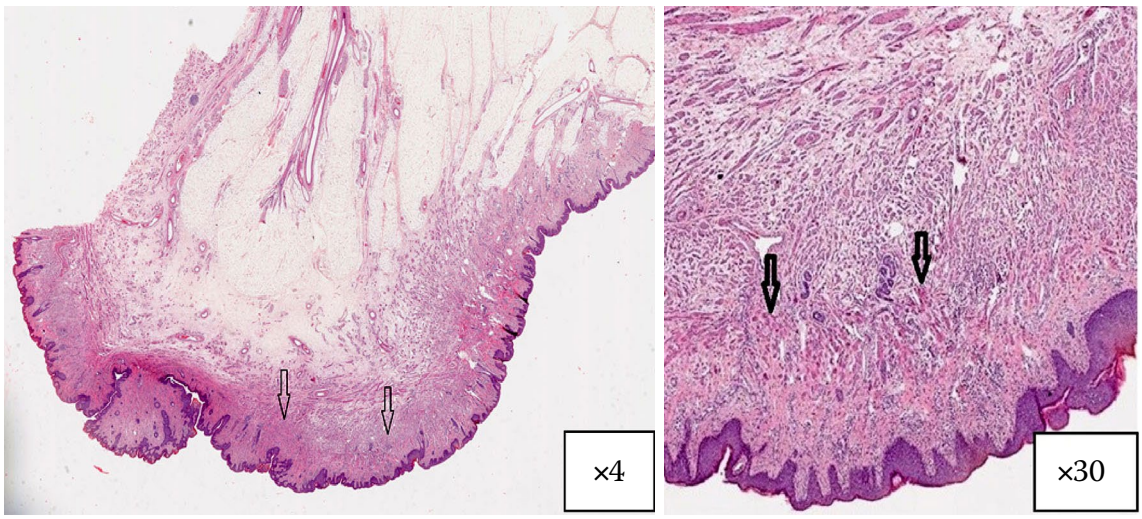

Figure 4. The histological examination revealed rugose epidermis, rudimentary dartos muscle fibers, and hair follicles confined within the mass, confirming the diagnosis of accessory scrotum. Slide stained with hematoxylin-eosin. Rudimentary dartos muscle fibers are indicated by the black arrow.

a mass resembling a lipoma was admitted to our clinic (Figure 3). Total excision was performed during elective surgery.

Systemic examinations of these three cases were unremarkable except for this mass. Penile and scrotal development were normal with bilateral descended testicles in all cases. The cases were discharged following the same day of surgery and recovered uneventfully. Histological investigation revealed rugose epidermidis, rudimentary dartos fibers, and hair follicles, confirming accessory scrotum (Figure 4). Written informed consent was obtained from the parents of all patients.

DISCUSSION

The exact prevalence of accessory scrotum remains uncertain, and approximately over 50 case reports have been published to date in the English literature.[2] According to our retrospective analysis, a series of 4,667 genitourinary surgeries (inguinal hernia, undescended testis, hydrocele, hypospadias, circumcision, and penoscrotal web) have been performed in our department since 2012. The reported three consecutive cases with accessory scrotum, however, attended during the previous two years. The scrotum forms from labioscrotal swellings at the fourth week of gestation. The labioscrotal swellings migrate inferomedially and merge at 12 weeks of gestation to form the scrotum, forming a line of fusion known as the scrotal raphe.[1] The exact etiology of an accessory scrotum has not been clearly defined. There are various published hypotheses concerning the etiology of accessory scrotum. Noguchi et al.[3] hypothesized that a lateral accessory scrotum forms from double-segmentation scrotal swelling on the perineal median line as repeated scrotal swelling occurs. Lamm and Kaplan[4] reported that failure of migration of the labioscrotal swellings from the caudal portion to the midportion results in the formation of an accessory scrotum. Takayasu et al.[5] suggested that this result may be due to a triple primitive anlage of the labioscrotal swelling or from a teratoid structure. Sule et al.[6] suggested that the accessory labioscrotal fold usually develops as a consequence of intervening mesenchymal tissue disrupting the continuity of the caudally developing labioscrotal swellings. The latter theory may explain the development of most accessory scrotums with mesenchymal tumors, as in our first two cases. However, this is insufficient to explain why they are not associated with a tumor, such as our last case.

Associated deformities include perineal mesenchymal tumors (including lipoma, lipoblastoma, and hamartoma), bifid scrotum, anorectal malformation, and hypospadias as in our series. The most common associated lesion is mesenchymal tumor, with an incidence rate of

83%, whereas anorectal malformation accounts for 18.6% of associated anomalies.[7] The clinical picture may show extreme manifestations, such as pseudodiphallia, skeletal abnormalities, renal dysplasia, spina bifida, retrocerebellar arachnoid cysts, and vertebral abnormalities.[8] Our third case was distinguished from the other two cases, as it was located in the inguinal region without an accompanying lipoma, and bilateral inguinal hernia was considered a related anomaly.

Three cases of accessory scrotum in the inguinal region have been previously reported.[9-11] Two of these cases had skeletal system anomalies,[9,10] while the third one was free of any anomalies. Apart from the literature, the absence of any other anomalies related to the musculoskeletal system or central nervous system in our patients may suggest that the factors causing accessory scrotum may not always affect these structures that differentiate in the early prenatal period. An increased number of reported patients may aid in understanding the extent and impact of factors triggering differentiation into an accessory scrotum during the early prenatal period.

In conclusion, the accessory scrotum remains a rare and poorly understood congenital anomaly. It is noteworthy that throughout the past two years, three cases have been reported in the Western Black Sea region. Future research should focus on expanding the case registry to understand better the developmental mechanisms and genetic factors involved in formation of accessory scrotum. By doing so, we can enhance clinical awareness. A multidisciplinary approach involving pediatric surgeons and pathologists will be essential in unraveling the complexities of this anomaly and providing optimal care to patients.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

Author Contributions: Methodology, conceptualization, surgery: D.T.U.; Validation and writing-original draft preparation: Y.D.B.; Surgery: B.A.; Data curation: F.K.; Writing-reviewing and editing: B.H.G. All authors read and approved the final manuscript.

Conflict of Interest: The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding: The authors received no financial support for the research and/or authorship of this article.

REFERENCES

1. Chatterjee S, Gajbhiye V, Nath S, Ghosh D, Chattopadhyay S, Das SK. Perineal accessory scrotum with congenital lipoma: A rare case report. Case Rep Pediatr 2012;2012:757120. doi: 10.1155/2012/757120.
2. Ibuka S, Saka R, Sonobe H, Tsukada R, Iwasaki S, Omote R. A case of dumbbell-shaped accessory scrotum with concomitant lipoma. Surg Case Rep 2024;10:106. doi: 10.1186/s40792-024-01906-w.
3. Noguchi M, Matsuoka K, Noda S, Etoh K. Accessory scrotum: a case report. 1984;75:1154-60. Japanese. doi: 10.5980/jpnjurol1928.75.7_1154.
4. Lamm DL, Kaplan GW. Accessory and ectopic scrota. Urology 1977;9:149-53. doi: 10.1016/0090-4295(77)90185-6.
5. Takayasu H, Ueno A, Tsukada O. Acdessory scrotum: A case report. J Urol 1974;112:826-7. doi: 10.1016/s0022-5347(17)59861-4.
6. Sule JD, Skoog SJ, Tank ES. Perineal lipoma and the accessory labioscrotal fold: An etiological relationship. J Urol 1994;151:475-7. doi: 10.1016/s0022-5347(17)34996-0.
7. Wang YF, Chou HC, Chen CY, Tsao PN. A neonate with imperforate anus and accessory scrotum with scrotal bifida. J Formos Med Assoc 2020;119:1331-2. doi: 10.1016/j.jfma.2019.11.022.
8. Murase N, Uchida H, Hiramatsu K. Accessory scrotum with perineal lipoma diagnosed prenatally: Case report and review of the literature. Nagoya J Med Sci 2015;77:501-6.
9. Gucev Z, Castori M, Tasic V, Popjordanova N, Hasani A. A patient with unilateral tibial aplasia and accessory scrotum: A pure coincidence or nonfortuitous association? Case Rep Med 2010;2010:898636. doi: 10.1155/2010/898636.
10. Kendirci M, Horasanli K, Miroglu C. Accessory scrotum with multiple skeletal abnormalities. Int J Urol 2006;13:648-50. doi: 10.1111/j.1442-2042.2006.01362.x.
11. Fitouri F, Chebil N, Ben Ammar S, Sahli S, Hamzaoui M. Accessory scrotum. Fetal Pediatr Pathol 2020;39:90-1. doi: 10.1080/15513815.2019.1629133.

