Duplicated urinary tract - A rare cause of perineal lesion in newborn. Case report.

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Introduction

Congenital perineal masses are rare entities that may be associated with anorectal malformation and urogenital anomaly. We present a case of a term baby boy with a duplicated urinary tract presenting as a perineal mass. He also has a bifid scrotum and penoscrotal hypospadias

Case Summary

This is a term, non-syndromic baby boy. He was referred to us upon birth for abnormal genitalia and perineal mass (Figure A). He passed large amount of meconium and urine shortly after birth. On examination, he has penoscrotal hypospadias with chordae and a complete penoscrotal transposition. He has a bifid scrotum and bilateral palpable descended testes. There is a midline perineal mass measuring 3cm x 3cm inferior to the hypospadic meatus, covered by mucosa-like tissue. The anus was located anteriorly and appears stenotic.

An ultrasonogram (USG) of the urinary system and MRI spine-pelvis was done, suggesting a perineal mass with small extension into the rectovesical space, low lying spinal cord and mild syringomyelia. He has normal bilateral kidneys and bladder otherwise. A complete excision of the perineal mass was done during neonatal period. Both intra-operative frozen section and formal histopathology shows urothelial epithelium, thus confirming the diagnosis of a duplicated urinary tract.

Discussion

Most reported cases of perineal mass for newborns are hamartomatous lesions. Differential diagnosis includes duplicated male genitalia, enteric duplication, vascular tumour or exstrophied bladder duplication. Embryologically, the urorectal septum develops into the perineum. It is hypothesized that an abnormal shape and development of the urorectal septum underlies the condition.

Radiological examinations (USG and MRI) are essential to delineate the extension of the lesion in relation to the adjacent structures (urethra, bladder, rectum),and to screen for other associated anomalies.

Excision is the definitive treatment and can be planned electively as long as child remains well. We have successfully achieved complete excision during neonatal period in this child

Conclusion

Duplicated urinary tract are rare perineal hamartomas. They are associated with other renal anomalies and anorectal malformation. The treatment goals include excision of the mass, reconstruction of the genitourinary tract and preservation of reproductive system, with early detection of other vesicorenal abnormalities.

Figure A: Male genitalia with a perineal mass

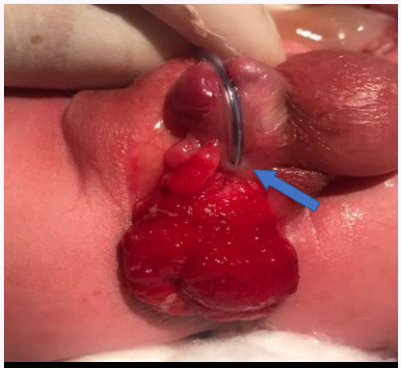
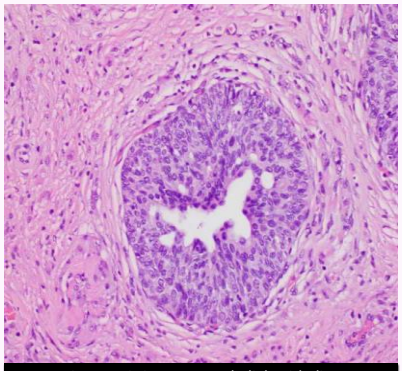
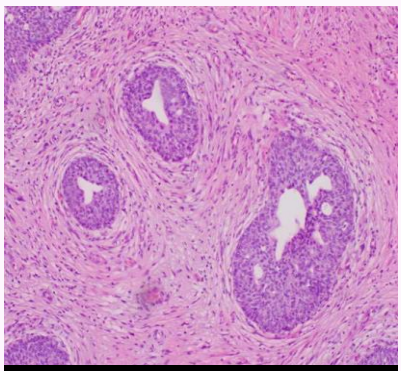


Figure B: Urethra meatus just above the mass (Arrow)



Microscopic images: Urothelial epithelium



Microscopic images: Urothelial epithelium

References:

1. Massoumi RL, Parsons LN, Jarzembowski JA, Wagner AJ. Pediatric pedunculated perianal problems. Journal of Pediatric Surgery Case Reports. 2016 Jun 1;9:45-8.
2. Sun J, Vongphet S, Zhang Z, Mo J. Perineal mass protrusion with rectal mucosa: a rectal duplication that underwent exstrophy. Journal of Pediatric Surgery. 2011 Aug 1;46(8):e5-8.