

Case Report

Rare presentation of nephrogenic adenoma at the meatus of penoscrotal hypospadias: A case report

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ABSTRACT

Background: Nephrogenic adenoma is a rare benign condition that arises in the urothelium but can occur anywhere along the urinary tract. Mainly it occurs in Caucasian males in the age range of (25-77years) with a ratio of 2:1 (male/female). The condition mainly occurs in adults; however, 10% of children are also affected. The lesion may occur as a metaplastic response to urothelium, or it may occur congenitally due to the proliferation of a metanephrogenic nest.

Case Presentation: We have presented a case of a three-year-old patient with penoscrotal hypospadias and a polypoidal urethral mass protruding out at the meatus of the phallus. Biopsy and immunohistochemistry report of the urethral polypoidal mass favored nephrogenic adenoma. The lesion was treated with transurethral resection, while penoscrotal hypospadias was planned to be treated by two-stage hypospadias repair.

Conclusion: In this case report, we have discussed a rare presentation of congenital nephrogenic adenoma at the meatus of penoscrotal hypospadias. The congenital origin of nephrogenic adenoma without any pre-existing cause has not been documented yet.

Keywords: Congenital nephrogenic adenoma, Penoscrotal hypospadias, Child, Urethral tumor.

INTRODUCTION

A nephrogenic adenoma is a rare benign lesion of the urinary collecting system in which cuboidal cells lining small tubules and cysts proliferate into the lamina propria with or without polypoid and papillary lesions in the lumen. [1] The bladder and different urinary tract sites comprise 80% of lesions, along with the urethra, which contains 15% of lesions, the ureter, 5%, and rarely in the renal pelvis, which is about <1%. [2] The exact etiology is unknown, various speculations include: it can be a metaplastic lesion, or it can emanate from embryonic tissue, or it could be metaplasia that on occasion co-occurs with multifocal urothelial carcinoma. [3]

In this case, we report the concurrence of nephrogenic adenoma originating from the embryonic Meso-nephrogenic nest, indicating its congenital origin without

any pre-existing cause at the meatus of penoscrotal hypospadias.

CASE REPORT

A 03-year-old male patient was presented with complaints of urine passage from the meatus at the penoscrotal level. On physical examination, the patient had a bifid scrotum (Fig. 1), bilateral testis palpable in the scrotum, short phallus, severe chordee, and urethral opening at the penoscrotal level, and a polypoid mass at the meatus of penoscrotal level (Fig. 1). On the digital rectal examination, no mass was palpable. Other investigations showed normal CP, Biochemistry, and Serology. Karyotyping showed 46XY, 17Hydroxy-progesterone levels were normal, Testosterone level = 0.2, HCG/DHT ratio = decrease 5-Alpha-Reductase. Ultrasound of the renal system appeared normal. Antegrade urethrogram and cystogram findings showed normal posterior urethra and bladder. Examination

under anesthesia showed severe chordee with penoscrotal hypospadias, bilaterally descended testes, a polypoidal mass protruding from penoscrotal meatus, and no inguinoscrotal swelling. On cystoscopy, the bladder looked normal. The polypoidal mass was resected and sent for histopathology. In a histopathology report, microscopic examination revealed solid proliferation of small round to oval cells with occasional tubular structures surrounded by a thickened hyalinized membrane. The individual cells were bland looking with scant cytoplasm. The tubules' lining showed hobnailing at a few foci with abundant eosinophilic cytoplasm. No granuloma, dysplasia, or malignancy was seen in the section examined. Histopathology was indicative of nephrogenic adenoma.

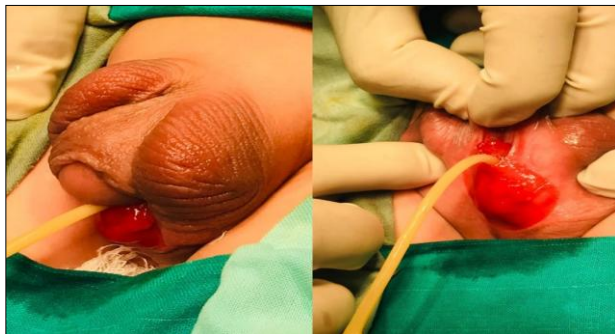


Figure 1: Polypoidal mass protruding out from penoscrotal hypospadias (right) and Bifid scrotum (left).

DISCUSSION

In 1949, Devi's described the very first case of nephrogenic adenoma. He coined the term "Hamartoma of bladder" for this kind of lesion. The name "Nephrogenic adenoma" was given by Friedman and Kuhlenbeck later in 1950, considering its histological appearance similar to renal tubules. [4]

The pathogenesis of nephrogenic adenoma remains controversial. The congenital origin of nephrogenic adenoma is supported by histological similarities of both renal epithelial and nephrogenic adenoma cells and the theories on the presence of common lectin receptors, i.e., Glycine max(SBA) and Arachis hypogea(PNA) on mesonephric and metanephric tubules and nephrogenic adenomas, suggests a structural courting of these entities. Any explanation of the pathogenesis of nephrogenic adenoma must account for its relative rarity, appearance after irritation or trauma, and

histologic, ultrastructural, and histochemical similarity to primitive renal tubules. [5]

Nephrogenic adenoma can also occur as a complication after hypospadias repair if a urethral bladder graft is used. [6] However, nephrogenic adenoma occurs congenitally in this case due to the proliferation of metanephrogenic nests in hypospadias patients. The lesion occurs congenitally, not because of hypospadias or any other chronic condition of the urinary tract.

Most children present with nonspecific signs and symptoms of gross hematuria, bladder instability, and dysuria, and in all cases, the final diagnosis can be established after ultrasound KUB, cystoscopy, and histopathologic evaluation of a biopsy specimen. [7] Histological findings are helpful in the diagnosis of congenital nephrogenic adenoma due to its epithelial resemblance with embryonic meta-nephrogenic rest.

Treatment of nephrogenic adenoma is transurethral resection. [8] The recurrence rate of these lesions varies from 28% to 90%. [9] Since the recurrence rate is very high, these patients should be followed for a long time. [10]

In conclusion, this case report discusses a rare presentation of nephrogenic adenoma through an abnormal meatus at the phallus in penoscrotal hypospadias. We review the pathogenesis and theories on its congenital origin. Usually, the congenital origin of the lesion is supported by any pre-existing associated condition of the urinary tract. However, in this report, the occurrence of congenital Nephrogenic adenoma without any underlying pathology and its protrusion through an abnormal meatus in a patient with penoscrotal hypospadias has never been presented before.

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