Primary solitary neurofibroma of penis in a child: case report and review

Mohammad Sohail Ahmad1, Santosh Kumar Mahalik1, Biswajit Sahoo2, Mukund Namdev Sable3, Akash Bihari Pati1 and Kanishka Das1*

Abstract
Background: Genital involvement in neurofibromas is rare and can involve both the clitoris and penis; in the latter, it usually accompanies a systemic pathology. Isolated penile neurofibroma is anecdotal. This report presents an unusual solitary penile neurofibroma in a child, discusses its surgical management and reviews the literature.

Case presentation: A 6-year child presented with history of swelling on the undersurface of his penis and obstructive voiding symptoms since 18 months of age. He underwent a partial excision at 5 years of age elsewhere which recurred. The swelling was a 3 × 2 cm, firm, midline, smooth surfaced, longitudinally oblong mass over the ventral penis extending from root to mid shaft with restricted mobility. There was no inguinal lymphadenopathy. Ultrasoundography and cystoscopy characterized it to be a corpus spongiosal soft tissue mass encasing and indenting the contained penile urethra. The mass was completely excised along with involved urethra and corpus spongiosum and a penoscrotal urethrostomy created. Histopathological analysis revealed it to be a benign nerve sheath tumor/neurofibroma. He had no stigmata of neurofibromatosis and the ophthalmologic examination was unremarkable. At 2-year follow-up, he is well, has normal glanular sensation and erectile function and awaits urethral reconstruction.

Conclusion: Primary solitary penile neurofibroma in children is extremely rare. This report details the presentation and management of a ventral penile neurofibroma involving the adjacent urethra/corpora spongiosa.

Keywords: Neurofibroma, Penis, Child, Case report

1 Background
Neurofibromatosis type1 (NF1/Von Recklinghausen disease) is an autosomal dominant neurocutaneous disorder with an incidence of 1 in 3000 live birth [1, 2] and may involve any region of the body. Genitourinary neurofibromas can originate from the pelvis, prostatic and/or vesicular nerve plexus, the urinary bladder being the commonly affected urogenital organ. Genital involvement is rare and can involve both the clitoris and penis [3]; in the latter, it usually accompanies a systemic pathology. They are of perineural and schwannian cell origin and arise from the autonomic cavernous plexus in the corporal bodies. This report presents an unusual solitary penile neurofibroma in a child and discusses its surgical management along with a review of literature.

2 Case presentation
A swelling was first noticed at 18 months of age on the undersurface of the penis and had increased in size progressively thereafter. It was excised at 5-years of age and the histopathology was suggestive of a soft tissue neoplasm. He presented to us after a year with a recurrent swelling. He voided with moderate straining but there was no history of retention of urine/hematuria/urinary tract infection.

There was a 3 × 2 cm, firm, midline, smooth surfaced, longitudinally oblong mass over the ventral penis extending from root to mid shaft. (Fig. 1a, b) It had a
restricted mobility and the inguinal lymph nodes were not palpably enlarged. The urinalysis, complete blood count, renal and liver function tests were normal. Ultrasound showed a mass in the corpus spongiosum encasing the proximal penile urethra (Fig. 1c, d). Cystoscopy (9 Fr scope) showed an extrinsic irregular mass chinking the urethral lumen beginning 3 cm from the meatus proximally for 2 cm (Fig. 2a, b). The bladder was mildly trabeculated and bilateral ureteric orifices were normal. The mass was excised along with the involved urethra and corpus spongiosa with a centimeter margin proximally and distally and a penoscrotal urethrostomy fashioned. Grossly, the 3.4 × 1.8 × 1.7 cm mass was smooth, had greyish white surface and a central bulbous enlargement. Microscopic examination revealed an unencapsulated, highly cellular spindle cell tumor. The cells were arranged in predominantly fascicular and focally storiform pattern, and they showed mild nuclear atypia and focal degeneration. The cytoplasm was positive for S-100 and the vessels positive for CD34, a diagnosis of a benign nerve sheath tumor—localized neurofibroma was made (Fig. 3). He had no stigmata of NF1 and the ophthalmologic examination was unremarkable. There are no signs of recurrence of the tumor after 2 years (Fig. 4a, b), and the ultrasound shows postoperative changes around the urethrostomy with normal adjacent urethra (Fig. 4c, d). The glanular sensation and erectile functions are preserved and he awaits urethral reconstruction.

3 Discussion
Diagnosis of NF1 is clinical with the presence of at least two of the seven criteria—Six or more café-au-lait spots, two or more cutaneous or subcutaneous neurofibroma/a plexiform neurofibroma, axillary or groin freckling, two or more Lisch nodules, optic pathway glioma, bony dysplasia and first degree relative with NF1 [2]. Genitourinary tract involvement in NF 1 has a prevalence of 0.65% with urinary bladder being the most commonly affected organ due to an abundance of autonomic nerves. Common extra vesical sites include renal hilum, ureters, uterus and prostate [3].

External genital involvement in neurofibromatosis is comparatively infrequent, clitoromegaly being the most...
common [3]. Penile involvement is usually associated with concomitant genitourinary involvement. Isolated penile neurofibroma is extremely rare, and only 17 cases of primary solitary neurofibroma of penis have been documented in children under 16 years since 1950 (Table 1) [1, 4–8]. Features of NF 1 were present in 10, and the rest were histological surprises. Two (Case 4, 14) had significant neurological affliction. The histology was described as plexiform (13/17), solitary (3/17) or left unqualified (2/17). Complete excision of the neurofibroma was curative in ten. In dorsal penile neurofibroma arising close to the neurovascular bundle, a complete resection preserving the bundle is technically impractical and somato-sensory deficit must be weighed against cosmesis. Partial resection is associated with recurrence and possible malignant transformation in 5–15% cases. In seven who had partial excision, five were curative. One had a local recurrence 2 months after surgery and underwent partial penectomy for a definitive cure thereafter. The case with multiple cranial nerve palsies and quadriplegia died later due to neoplasia elsewhere (cerebral ependymoma). Although the short-term follow-up mentions improvement and no recurrence, long-term follow-up data are lacking.

In contrast, the lesion described here was located on the ventral aspect of the penis and indented the urethra. The management aimed at complete resection with a cuff of normal tissue and delayed urethral reconstruction.
4 Conclusions

Primary solitary penile neurofibroma in children is extremely rare. This report details the presentation and management of a ventral penile neurofibroma involving the adjacent urethra/corpora spongiosa. Total excision of neurofibroma is ideal; however, partial excision do give good results with rare instances of recurrence. Long term follow of these cases is warranted to detect any recurrence and malignant transformation.
Abbreviation
NF1: Neurofibromatosis type 1.

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Case conception and design: SKM. Acquisition of data: MSA/BS/MNS, KD. Analysis and/or interpretation of data: SKM, KD. Drafting the article: MSA/SKM. Images design: BS/MNS/KD. Revising it critically for important intellectual content: ABP/KD. All authors contributed to the final approval of the version to be published. All authors read and approved the final manuscript.

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Ethics approval and consent to participate
All procedures performed in this study were in accordance with the ethical standards of our institutional review board with the 1964 Helsinki declaration. Consent for this surgery was obtained from the patient and her father.

Consent for publication
Consent for publication of this report was obtained from the patient and his father.

Availability of data and materials
Not applicable.

Competing interests
None to declare.

Table 1  Pediatric patients (below 16 year) with primary penile neurofibroma

| Case | Author (Year) | Age | Features of NF 1 | NF subtype | Surgical management | Follow up (period) |
|------|---------------|-----|-----------------|------------|---------------------|-------------------|
| 1    | Haines and Garvey (1950) | Neo | Yes | Plexiform | Total excision | Improved (1 year) |
| 2    | Witus et al. (1958) | 27 months | Yes | Plexiform | Partial excision | Improved? |
| 3    | Dehner et al. (1970) | Neo | Yes | Unclassified | Total excision | Improved (1 year) |
| 4    | Fethiere et al. (1974) | Neo | Cranial nerve palsies, cervical syrinx and ependymoma, flaccid right hand, quadriplegia | Plexiform | Partial excision | Improved, demise with other tumors, ? NF-2 |
| 5    | Elliott et al. (1981) | 7 years | No | Solitary | Partial excision | Improved (18 months) |
| 6    | Dwosh et al. (1984) | 8 years | Yes | Plexiform | Total excision | Improved (6 months) |
| 7    | Thompson PD et al. (1992) | 4 years | Yes | Unclassified | Total excision | Improved (5 years) |
| 8    | Mathew R et al. (1996) | 2 years | No | Plexiform | Partial penectomy | Improved? |
| 9    | Rodo J et al (1999) | 3 years | No | Plexiform | Partial excision | Improved (1 year) |
| 10   | Kousseff et al. (1999) | 27 months | Yes | Plexiform | Partial excision | Recurred after 2 months, partial penectomy, improved thereafter (6 years) |
| 11   | Littlejohn et al. (2000) | 9 years | Yes | Plexiform | Partial excision | Improved (1 year) |
| 12   | Madak H et al. (2007) | 5 years | No | Plexiform | Total excision | Improved (13 months) |
| 13   | Ballouhey Q et al. (2013) | 13 years | No | Solitary | Total excision | Improved (8 months) |
| 14   | Douglas et al. (2014) | 8 years | Facial nerve involvement | Plexiform | Total excision | Improved, periodic follow up |
| 15   | Sridutt et al. (2017) | 12 years | No | Solitary | Total excision | Improved? |
| 16   | Collins NC et al. (2018) | 11 years | No | Plexiform | Total excision | Improved (1 year) |
| 17   | Banthia R et al. (2020) | 7 years | Yes | Plexiform | Total excision | Improved (6 month) |

Abbreviation
NF1: Neurofibromatosis type 1.

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Author contributions
Case conception and design: SKM. Acquisition of data: MSA/BS/MNS, KD. Analysis and/or interpretation of data: SKM, KD. Drafting the article: MSA/SKM. Images design: BS/MNS/KD. Revising it critically for important intellectual content: ABP/KD. All authors contributed to the final approval of the version to be published. All authors read and approved the final manuscript.

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