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CASE REPORT

Large Solitary Plexiform Neurofibroma of the Penis Without Erectile Dysfunction: a Case Report from Kerala

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Abstract
Case Report Solitary plexiform neurofibroma of the penis is an extremely rare peripheral nerve lesion with only about 20 cases reported all around the world. Here, we are reporting a case of solitary penile plexiform neurofibroma with sexual dysfunction due to difficulty in penetration. This 47-year-old gentleman presented with a swelling on the dorsal side of the penis. He was aware of the swelling for the last 15 years. The swelling was slow growing and there was no associated loss of sensation or sexual dysfunction. For the last 2 months, he noticed that the tumour is rapidly increasing in size. He was also having difficulty in penetration during sexual intercourse which forced him to seek medical attention. He underwent excision of the swelling from our department. After 18 months of follow-up following excision, there has been no evidence of recurrence of the tumour.
Conclusion Penile plexiform neurofibromas are a rare type of lesions and are usually associated with neurofibromatosis. Solitary penile plexiform neurofibromas are extremely rare. Excision, when done properly, gives good results, without erectile dysfunction.

Keywords Plexiform neurofibroma · Penis · Solitary · Schwannoma

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Case Report

This 47-year-old gentleman from Kerala presented to surgery OPD with a swelling on the penile shaft, which he noticed 15 years back; it was initially smaller and gradually increasing in size till it reached the present size of 5.5 × 5 × 3.5 cm^3. He never experienced pain, altered sensation or erectile dysfunction due to the swelling. He is married and has fathered two children. Recently, he noticed increase in the size of the swelling, due to which he had difficulty in mechanical penetration during sexual intercourse. His International Index of Erectile Function-5 score was 18.

He had no comorbid illness or relevant family history. His general examination revealed no neurocutaneous markers, or axillary/inguinal freckles or opthalmologic involvement. Clinical examination showed 5.5 × 5 × 3.5 cm^3 sized ovoid, firm swelling on the dorsal aspect of the penis with fixity to fascia and without fixity to corpus cavernosum/spongiosum (Fig. 1a). There was no inguinal lymphadenopathy. Detailed opthalmologic examination revealed no glioma or Lisch nodules. Routine laboratory investigations were within normal limits. MRI showed a lesion of size 5.5 × 5 × 3.5 cm which was hypointense in T1-weighted images and heterointense in T2-weighted images (Fig. 1b, c).

Excision was performed under spinal anaesthesia. The swelling was arising from the dorsal nerve of the penis and fixed to Buck's fascia. There was no extension to corpus cavernosum/spongiosum. There was no neurovascular injury during the procedure. Gross appearance of the resected specimen showed light brown nodular tissue measuring of 5.5 × 5 × 3.5 cm with

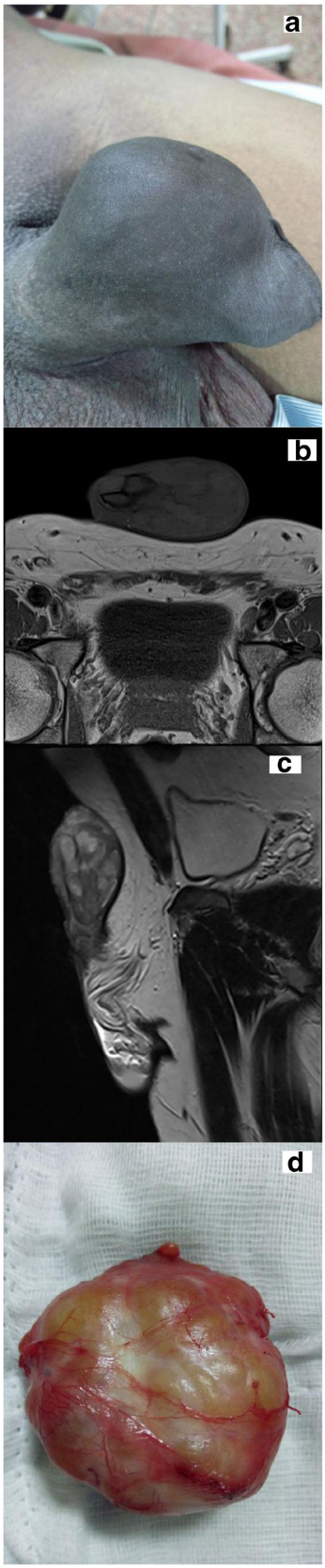

Fig. 1 **a**–**c** Penile shaft tumor preoperative (external view (**a**) and MRI—T1-weighted (**b**), T2-weighted images (**c**)) and **d** postoperative images

bosselated surface (Fig. 1d). Cut section showed fatty and fibrotic area with lobulation. Microscopy showed a neoplasm composed of spindle-shaped cells with elongated nuclei arranged in a lobulated pattern. Immunohistochemistry showed that the cells with mild positivity for S100. Ki-67 proliferation index was 2–3%. Histopathology was suggestive of penile plexiform neurofibroma (Fig. 2).

After surgery, he had a normal sensation over the penis and no pain. He was followed up for the last 18 months in our OPD and showed no evidence of recurrence. His postoperative International Index of Erectile Function-5 score was 24.

Discussion

Neurofibromas are benign peripheral nerve sheath tumours having Schwann cells, perineurial-like cells and fibroblasts, mixed with nerve fibres, collagen and myxoid matrix [1]. They are usually found in young individuals. The cell of origin is the Schwann cell.

Neurofibromas are usually classified based on (a) number of lesions, (b) the morphology and (c) location.

Neurofibromas may be classified into solitary or isolated and multiple based on the number of neurofibromas present in the body. Most of these are solitary and not associated with neurofibromatosis type 1, while multiple neurofibromas usually occur in neurofibromatosis.

Based on morphology, neurofibromas are classified into discrete and non-discrete lesions [2]. Discrete lesions include cutaneous and intraneural neurofibromas and non-discrete lesions include plexiform variety. Majority of the neurofibromas in routine clinical practice include discrete lesions.

Cutaneous neurofibromas arise from small cutaneous nerves. They are small, nodular tumours of the skin and subcutaneous tissue. They may be associated with local pain or bleeding but usually do not cause neurologic deficits.

Intraneural neurofibromas arise from nerve roots, nerve trunks, nerve plexuses or peripheral nerves. They are deeper, focal, well-circumscribed, fusiform lesions. These tumours usually present as a mass, with local or radicular pain.

Plexiform neurofibromas are multinodular elongated masses affecting large nerves and are typically associated with neurofibromatosis type 1. Solitary plexiform neurofibromas are rare. Microscopy shows myxoid background that contains Schwann cells, fibroblasts and mast cells with occasional nuclear palisading. Typically, mild S100 positivity will be present. Excision of the lesion is more difficult in plexiform neurofibroma compared to other types of neurofibroma.

Excision remains the treatment of choice for all symptomatic neurofibromas while asymptomatic cutaneous

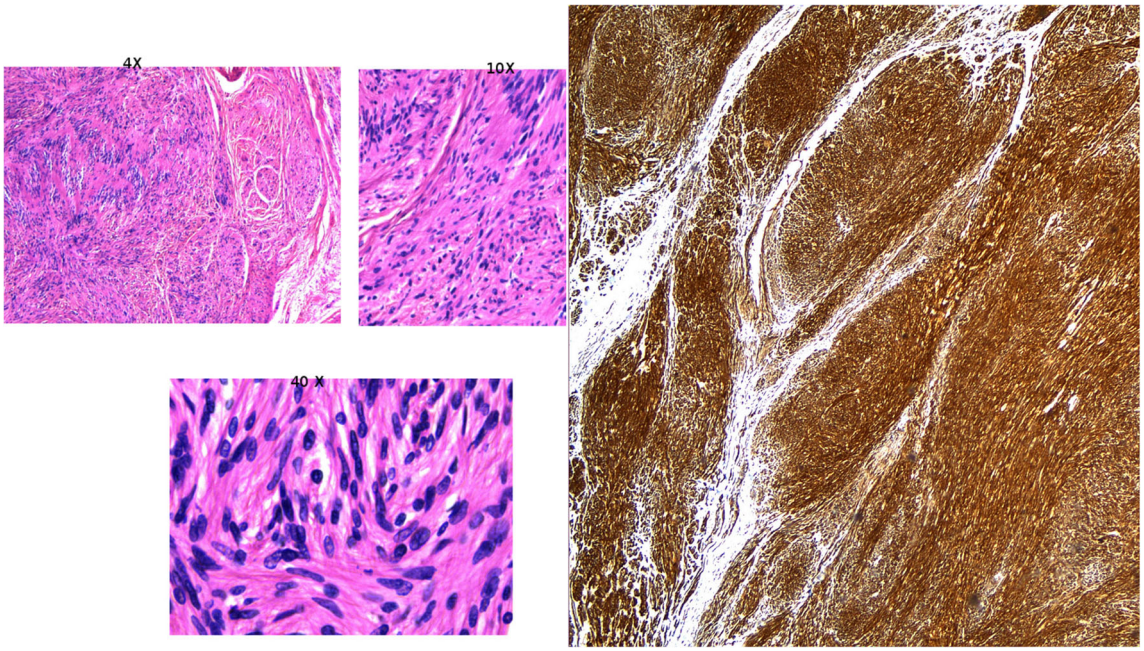

Fig. 2 Histopathology showing plexiform neurofibroma (4×, 10×, 40×) and S100 staining

neurofibromas/intraneural neurofibromas may be managed conservatively on regular follow-up. Plexiform neurofibromas have a higher chance of malignant transformation, and complete excision without compromising the parent nerve function is the treatment of choice [3, 4].

Neurofibromas are more common in the head and trunk, however may involve the bladder, the spermatic cord, the colon, the stomach and extremities [5, 6].

Neurofibromas of the penis, although very rare, are usually associated with neurofibromatosis type 1. Primary solitary neurofibromas of the penis are extremely rare [7], and are often associated with neurofibromatosis type 1. To our knowledge, only about 20 patients with primary neurofibromas associated with penile neurofibromatosis have been described in the medical literature [3, 8].

Penile neurofibroma involves continuity of the dorsal neurovascular bundle with the mass. However, excision can be done safely. Complete resection is necessary to prevent recurrence and to prevent malignant transformation.

Conclusion

Penile plexiform neurofibromas are a rare type of lesions and are usually associated with neurofibromatosis. Solitary penile plexiform neurofibromas are extremely rare. Excision, when done properly, gives good results, without erectile dysfunction.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

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