

*Case Report*

## Intrascrotal Solitary Plexiform Neurofibroma: A Case Report

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### ABSTRACT

Neurofibroma is a benign peripheral nerve sheath tumor that can occur in a variety of sites. They are classified into cutaneous, subcutaneous and plexiform subtypes. Plexiform neurofibromas are the least common variant and are pathognomonic for Neurofibromatosis -1. The following is a case report of 53yr old man with solitary plexiform neurofibroma of the scrotum. This rare benign tumor has the potential for malignant transformation and the present case is unique as there were no other manifestations or family history of NF-1.

**Key words:** neurofibroma, plexiform neurofibroma, scrotum.

### INTRODUCTION

Plexiform neurofibroma is a benign peripheral nerve sheath tumor arising from proliferation of all neural elements and surrounds multiple nerve fascicles. It is usually associated with other features of neurofibromatosis 1 (NF-1) an autosomal dominant disorder with an incidence of 1 in 2500-3500 birth. [1] It may also occur as an isolated lesion without definitive evidence of NF-1. [2] Plexiform NF arise in various parts of the body including skin, subcutaneous, cranio facial region, paraspinal, mediastinum, viscera and retroperitoneum. Plexiform NF involving the genito urinary tract is rare, with bladder, upper urinary tract and genital involvement reported in decreasing order of frequency. [3] They are associated with 5 % risk of malignant transformation. [4] The following is a case report of 53 year old man having

isolated plexiform neurofibroma of the scrotum.

### CASE REPORT

A 53-year-old man was admitted to our hospital in the surgery department with complaints of gradually enlarging mass and discomfort in both scrotums for six years. . He gave a history of on and off pain and fever for the past 2 years. There was no history of trauma, voiding complaints, or signs and symptoms related to genitourinary disease. He had no significant past medical history nor family history. Systemic examination was unremarkable. Local examination showed a large firm painless, non-transilluminating, and diffuse mass beneath the scrotal wall. Both the testis is normal; however, because of the presence of large mass, relation of the mass with epididymis and spermatic cord could not be exactly located. No hernia was detected.

Laboratory investigations were within normal limits. Clinically the case was diagnosed as bilateral scrotal lymphedema. The patient was scheduled for reduction surgery. The lesion was dissected from the surrounding tissues along with the excess scrotal wall, and it was noted that the mass did not involve the other surrounding structures. The specimen was sent for histopathological examination.

We received a skin covered soft tissue mass measuring 21 x 18 x 9 cm with multiple warty growth of the skin. Cut surface showed diffuse grey white mass which was soft and firm in consistency. Microscopic examination showed warty hyperplasia of the epidermis overlying a benign neoplasm composed of enlarged nerve fascicles of spindle shaped cells with elongated wavy nuclei in a stroma of fibrillary collagen and large foci of myxomatous change. In between plexiform masses were focal areas of inflammatory cell collections and small and medium sized blood vessels with perivasculitis. Immunohistochemical study of the spindled Schwannian cells showed positivity for S 100 and also positivity for epithelial membrane antigen (EMA) in perineural cells. A final histopathological diagnosis of intrascrotal solitary plexiform neurofibroma was given.



Figure 1. Excised scrotal wall mass



Figure 2. Cut surface showing diffuse grey white mass.

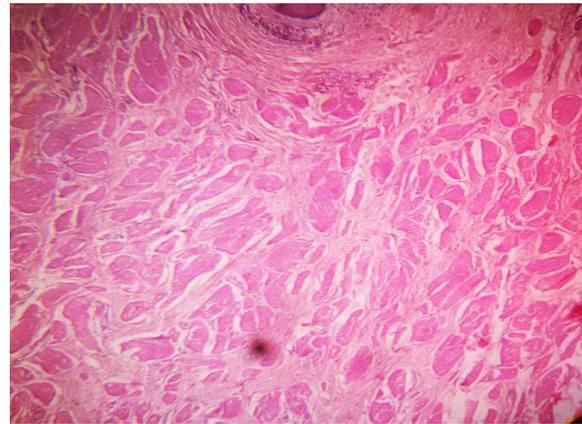


Figure 3. Bundles of nerve fascicles. H&E 10X

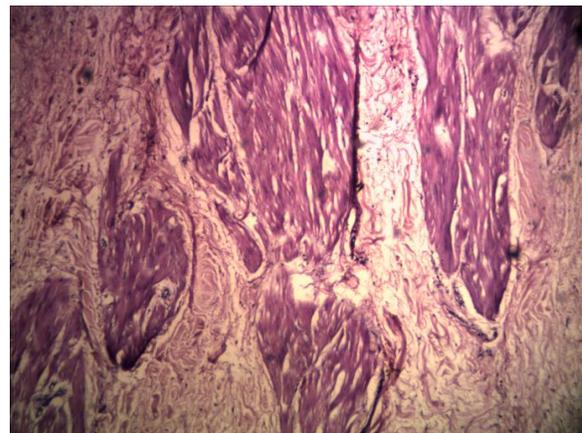


Figure 4. Enlarged nerve fascicles of spindle shaped cells with elongated wavy nuclei. H&E 40X

In our case pathological findings were similar to those reported in the literature. Our case had the largest size and immunohistochemical findings strongly supported our diagnosis.

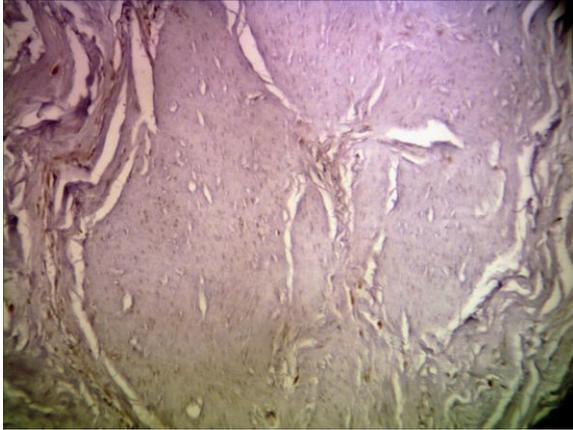


Figure: 5. S-100 Showing positivity

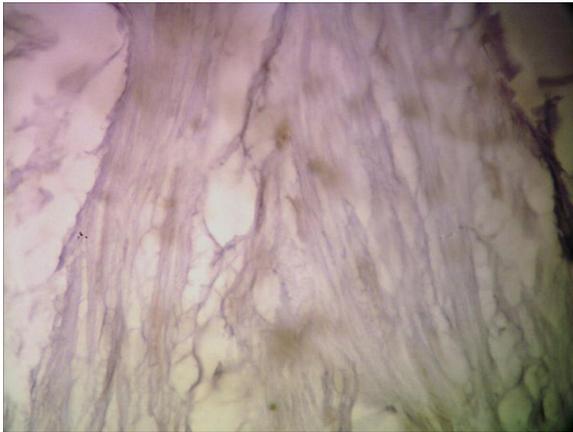


Figure: 6. EMA Showing positivity in perineural cells.

## DISCUSSION

Scrotal tumours were mostly extra testicular arising from spermatic cord and epididymis. Benign mesenchymal scrotal tumours are leiomyoma, fibroma, lipoma, hemangioma and epidermoid cyst. [5] Few cases of isolated scrotal neurofibromas have also been reported. [5,6]

Neurofibromas occur as solitary, diffuse or plexiform lesions with varying clinical and pathological findings. These tumours were well demarcated but unencapsulated consisting of an admixture of Schwann cells, perineurial-like cells and fibroblasts. [7] Plexiform neurofibromas generally occur in early childhood. They occur in any location neck, scalp, tongue, major nerves, sinonasal tract, larynx, salivary gland and inguinal region. [8] Presence of plexiform neurofibroma is

virtually pathognomonic of NF-1. They occur in only 17% of cases of NF I but some patients with small solitary plexiform neurofibromas may not have NF-1. [4] Isolated plexiform neurofibromas have been reported in cauda equina, pedal, penile, skin, bladder, buccal, salivary gland and oral cavity [9-14] Grossly they are ill-defined nodular or multinodular growth with bag of worm appearance. The tumor consists of proliferation of Schwann cells, fibroblasts and axons with associated thick collagen fibres. Hypercellularity, cellular pleomorphism and mitosis may be seen. Presence of increased mitotic activity may be indicative of malignant transformation. The tumor is positive for s100 protein in a small subset of cells and EMA positive perineurial cells are common in plexiform but not in ordinary neurofibromas. [7]

## CONCLUSION

Plexiform neurofibromas should be carefully monitored, because 5% may turn into malignant peripheral nerve sheath tumors. It is necessary to distinguish isolated neurofibroma from neurofibroma associated with NF1, because recurrence and malignant potential are more in neurofibroma associated with NF1.

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