

Intrascrotal Extratesticular Neurofibroma: A Rare Case Report

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ABSTRACT

Solitary neurofibroma of the scrotum is an extremely rare benign tumor, particularly when it is not associated with neurofibromatosis type I. To the best of our knowledge, less than 10 cases have been reported in the English literature.

We report a case of 77 year old male who presented with large right scrotal mass which was surgically excised and after histopathological examination diagnosis of intrascrotal extratesticular neurofibroma with degenerative atypia was made.

Keywords: Extratesticular, Intrascrotal, Neurofibroma

Introduction

Neurofibroma is a benign tumor of the nerve sheath, which results from an abnormal overgrowth of Schwann cells. It can be encountered anywhere within the central or peripheral nervous system, especially in the neck, thorax, cranium, retroperitoneum, and flexor surfaces of the extremities.^[1] Neurofibroma can be solitary or multiple. Solitary scrotal neurofibromas unassociated with neurofibromatosis type I (NF I) are extremely rare. Scrotal tumors are mostly extratesticular, originating from spermatic cord and epididymis. To the best of our knowledge, less than 10 cases have been reported in the English literature.^[2]

Herein, we report a case of 77-year-old man with the diagnosis of intrascrotal extratesticular neurofibroma with degenerative atypia.

Case Report

A 77-year-old man admitted to our hospital with a ten year history of gradually enlarging mass and scrotal discomfort in the right side of scrotum. There was no history of trauma, voiding complaints, or signs and symptoms of genitor-urinary disease.

On physical examination a firm, non-tender, mobile, and non-trans illuminating mass of size 23x21x10 cm and separately palpable from the testis was detected in right side of scrotum. There was no evidence of inguinal lymphadenopathy. Overlying skin was unremarkable. Laboratory investigations including testicular tumour markers were within normal range. Ultrasonography demonstrated a heterogeneous, hypo echoic, solitary mass of minimal vascularity, along the margins of right testis.

The other urinary structures were normal. With clinical diagnosis of testicular tumor, orchidectomy was done and the mass was sent for histopathological examination.

Grossly it was a mass of size 22x21x9 cm, already cut open, external surface capsulated, glistening. Cut section showed fish flesh like appearance & soft consistency. Testicular tissue was identified separate from the mass at one end. (Fig 1 & 2)

Microscopically it showed partially encapsulated tumour mass, with low to moderately cellularity. The individual tumour cells were slender, spindle shaped, wavy with oblong nuclei on the background of neurofibrillary network. There were areas of hyalinization and myxoid change. Part of the tumour showed few bizarre cells with nuclear inclusions and smudging of nuclei suggestive of ancient changes. No evidence of mitosis, hemorrhage or coagulative necrosis were seen. Part of the section showed normal architecture of testis with unremarkable seminiferous tubules.

The histopathological diagnosis of intrascrotal extratesticular neurofibroma with degenerative atypia (ancient changes) was made. (Fig 3 & 4)

Discussion

Benign intrascrotal lesions are common findings in the male population. Unlike testicular lesions, which are 95% malignant, paratesticular lesions are mostly benign. Benign tumors commonly seen in the scrotum are leiomyomas, lipomas, fibromas, hemangiomas.¹ Neurofibroma is a benign tumor of the nerve sheath originating from the Schwann cells.² Neurofibroma can be solitary or multiple and are commonly seen in between of age 8 and 77 years. Most of them occur in paratesticular tissue such

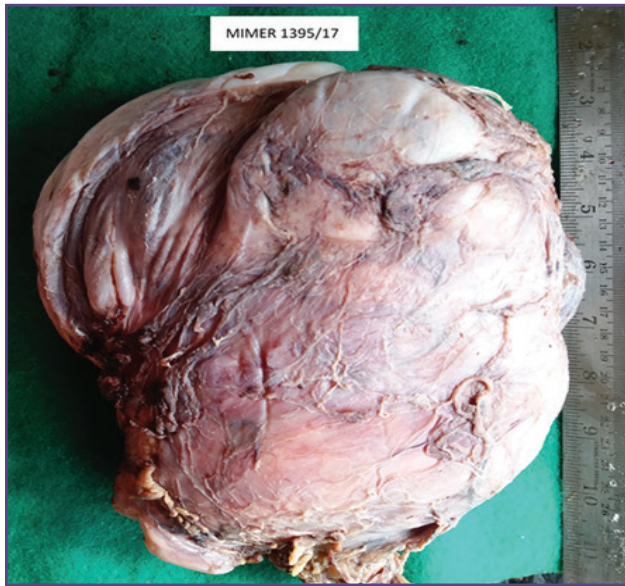


Fig. 1: Photomicrograph showing partially encapsulated tumour mass.



Fig. 2: Photomicrograph showing glistening and fish flesh like cut surface of tumour mass.

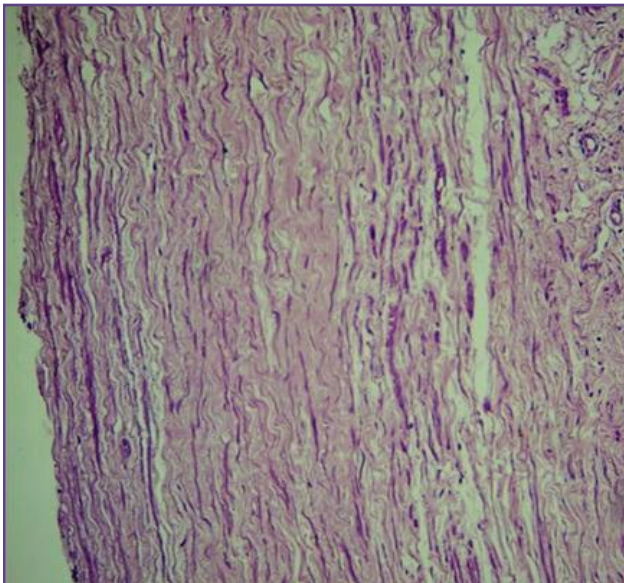


Fig. 3: Photomicrograph showing tumour mass composed of wavy spindle shaped cells with oblong Nuclei on neurifibrillary background (H&E 10X).

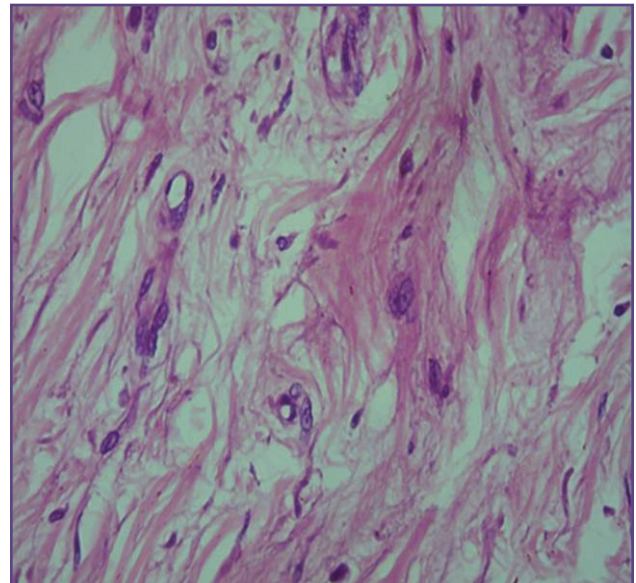


Fig. 4: Photomicrograph of tumor mass showing few bizzare cells (H&E 40X).

as epididymis or spermatic cord. Neurofibromas are well-differentiated nerve sheath tumors of benign origin, which commonly present as a part of systemic disease NF-1 or NF-2.³ In most cases of scrotal neurofibroma, the exact origin of the tumor cannot be determined, but overall the majority of them are extratesticular.² Solitary neurofibroma within the scrotum, which is unassociated with neurofibromatosis, is an extremely rare benign tumor.³ The presenting complaints are usually scrotal discomfort,

painless swelling and hydrocele.^{1,2} The gross appearance of neurofibroma varies from lesion to lesion. As a rule, the tumors are not encapsulated and have a softer consistency.¹

Microscopically, neurofibromas are formed by combined proliferation of all the elements of peripheral nerve axons, Schwann's cells, fibroblasts, and perineural cells. Schwann's cells are usually the predominant cellular elements in the tumor. These lesions are immunoreactive for S-100 protein.¹

Some neurofibromas show unusual features such as degenerative cytological atypia (neurofibroma with ancient change, atypical neurofibroma) and/or increased cellularity (cellular neurofibroma), often raising the differential diagnosis with Malignant Peripheral Nerve Sheath Tumour (MPNST). Cellular neurofibromas may show moderate cellularity and a more pronounced fascicular growth pattern, but lack the “monotonous” generalized cytological atypia, chromatin abnormalities and mitotic activity seen in MPNST. Neurofibromas with ancient change show degenerative nuclear atypia, containing scattered cells with markedly enlarged, hyperchromatic nuclei, often with “smudgy” chromatin; however, they lack increased cellularity, fascicular growth, or mitotic activity. Similar changes may be seen in so-called “ancient schwannomas”. Other less common morphological findings in neurofibroma include the presence of melanin pigment, metaplastic bone and glandular differentiation.^{4,5,6}

The distinction of atypical or cellular neurofibroma from low grade MPNST change is perhaps the most difficult challenge in the pathology of peripheral nerve sheath neoplasms, particularly in the setting of an NF1 patient. Some authors use the term “atypical neurofibroma” to denote neurofibromas with degenerative nuclear changes, analogous to ancient change in schwannoma. These tumors are of little concern. Others, however, have reserved this term for nerve sheath tumors showing worrisome histologic features (e.g., high cellularity, scattered mitotic figures, monotonous cytology or fascicular growth), but not fully meeting criteria for malignancy. Clinically, atypical changes usually develop in large, slowly growing neurofibromas, and pain may be a feature. “Atypical neurofibromas” have generally been regarded as benign.⁴

Our case was a 77 yr old man who presented with gradually enlarging mass and scrotal discomfort in the right side of scrotum since 10 years. On physical examination, it was a firm, non-tender, mobile, non-trans illuminating, mass of size 23x21x10 cm and separately palpable from the testis in the right side of scrotum. Grossly mass was of size 22x21x9 cm, with partially capsulated & glistening external surface. Cut section showed fish flesh like appearance & soft consistency. Microscopically it showed partially encapsulated tumour mass, with low to moderately cellularity. The individual tumour cells were slender, spindle shaped, wavy with oblong nuclei on the background of neurofibrillary network. Part of the tumour

showed few bizarre cells with nuclear inclusions and smudging of nuclei suggestive of ancient changes. No evidence of mitosis, hemorrhage or coagulative necrosis were seen. Thus, absence of mitotic activity, hemorrhage or coagulative necrosis ruled out malignancy, however the presence of low to moderate cellularity, few bizarre cells with nuclear inclusions and smudging of nuclei pointed towards atypia.

Exact origin of the tumor was unknown in the reported cases¹. In our patient, we also could not determine the exact anatomic structural origin of the tumor. But we recognized that this mass was separate from the testis, vas deferens, and epididymis.

In neurofibroma, the treatment is surgical excision of tumor.¹The tumor has a good prognosis, and its complete excision has yielded good results with no recurrence.⁴ In cases, where the tumor involves the testicle, orchidectomy is inevitable.¹

Conclusion

Although extremely rare, intrascrotal extratesticular neurofibroma should be considered in the differential diagnosis of testicular and paratesticular tumors. Neurofibroma with ancient changes showing degenerative atypia should be differentiated from low grade MPNST by lack of increased cellularity, mitotic activity and coagulative necrosis.

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