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Case Report

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BEYOND THE USUAL SUSPECTS: A CASE OF PARA TESTICULARSCHWANNOMA

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ABSTRACT

Schwannomas are usually benign encapsulated tumors that commonly arises from cranial, spinal and peripheral nerves, mostly effecting the head and neck region along the nerve sheath. Although schwannoma is the most common peripheral nerve sheath tumor, it is extremely rare in Para testicular region. This is a case of 53-year-old male with left scrotal non tender swelling; there is no regional lymphadenopathy and no regional skin involvement. USG revealed a well encapsulated heterogeneously echogenic extra testicular lesion in left testis. MRI revealed a left para-testicular well encapsulated lobulated T1 isointense, T2 heterogeneous appearing mass measuring 5.2x5x5.3cm (APXTSXCC) in the scrotum. Surgical excision was done involving left inguinal orchidectomy with spermatic cord. Histopathological analysis of the specimenconfirms the diagnosis of schwannoma.

INTRODUCTION

Schwannoma is a benign encapsulated neoplasm that usually arises from cranial, spinal, and peripheral nerves. They mostly affect the head and neck region along a nerve sheath. [2] These tumors consist of clonal population of Schwann cells which undergo cystic and degenerative changes. Most cases are sporadic; some are associated with NF2, schwannomatosis or Carney's complex.[9] Although schwannoma is the most common peripheral nerve sheath tumor, it is extremely rare in the Para testicular area.

Histologically, schwannoma is an encapsulated tumor with biphasic architectural pattern composed of Antoni A and Antoni B areas. Antoni A areas correspond to compacted spindle cells that are often arranged in palisades or adopt an organoid arrangement (Verocay bodies), whereas Antoni B areas are characterized by loose- textured tissue in a myxomatous matrix that may appear microcystic.[9]

Immunohistochemistry studies show uniform positive staining for S-100 protein.[2]

The diagnosis of schwannoma is challenging, and the clinical and radiographic findingsof scrotal schwannoma are non-specific, which means the tumor can be easily misdiagnosed as another solid tumor. Patients usually present with an asymptomatic, slowly growing scrotal mass with duration of several months to a year.[3,4,5,6]

The most common extra testicular tumor is adenomatoid tumor, which occurs at a peakage of between 20 and 50 years. These patients usually present with an asymptomatic scrotal tumor. Adenomatoid tumors are smooth, round, well-defined, and varying in size. On ultrasound, they are typically homogeneous and hyperechogenic in nature.[7] Leiomyoma is the second most common tumor of the epididymis. This tumor most commonly manifests as a slow-growing, nontender scrotal mass, which occurs at a peakage during the fifth decade of life. Leiomyomas usually present as well-demarcated tumor surrounded by a gray-white fibrous capsule and would range from 1 to 4 cm in size. This tumor has a variable sonographic appearance whether it is predominantly solid or cystic, and it may contain calcifications. Leiomyomas can be associated with a hydrocele in half of the cases.[7,8] So, radiologists and urologists should keep Schwannoma in mind as a differential diagnosis in the presence of an intra-scrotal extratesticular mass.

The only gold standard diagnostic investigation is histology of either the biopsy or excised specimen. Surgical excision is a mainstay of treatment. Recurrence aftercomplete surgical excision is uncommon. Similarly, malignant transformation in a schwannoma is extremely rare [1]. However, if the diagnosis of extra testicular benign lesion is made preoperatively, less invasive surgery might have been considered as a treatment option.

Case Report: A 53 – year - old man presented with an approximately 8 - to 10 - year history of a painless,slowly growing left scrotal mass. On physical examination, a firm, mobile, non-tender mass was palpated in the left hemiscrotum. No overlying skin changes were noted. There is no significant medical or family history. Complete

blood picture, urine analysis, LFTs, RFTs, TFTs were normal. Serum LDH levels are elevated 304 U/L. ABP and β- hCG were negative. Incidentally HBsAg was positive.

Initial ultrasonography of scrotum and abdomen revealed well encapsulated heterogeneously echogenic extra testicular left scrotal lesion with internal vascularity, closely abutting (fig.1) and lateral to left testicle and cholelithiasis respectively.

MRI revealed a left para-testicular well encapsulated lobulated T1 isointense, T2 heterogeneous appearing mass measuring 5.2x5x5.3 cm (APXTSXCC) in the scrotum; not revealing any obvious diffusion restriction. Lesion closely abutting and displacing the left testis posterosuperiorly. Lesion revealing broad based contact with scrotal inner wall. Both testicles and epididymis otherwise appear unremarkable. No abnormal enlarged inguinal lymph nodes. No abnormal signal intensity in the external scrotal skin.

Possibilities of extra testicular Leiomyoma, Fibroma, Adenomatoid tumor (in that order) have been suggested.

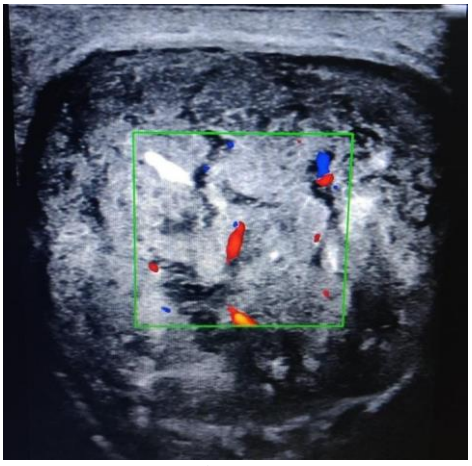

Figure 1

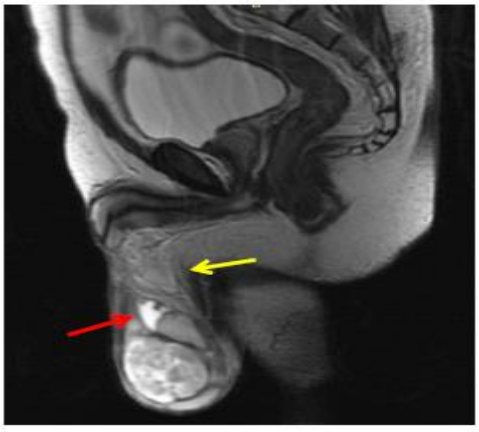

Figure 2

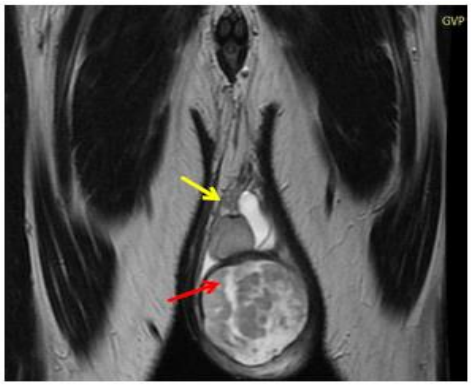

Figure 3

Figure 2 - Sagittal T2 weighted image, revealing the well encapsulated lesion (red arrow) displacing the left testis posterosuperiorly (yellow arrow), with minimal fluid in the left scrotal sac.

Figure 3 - Coronal T2 weighted image revealing relationship between the tumour and left testis (Left to right).

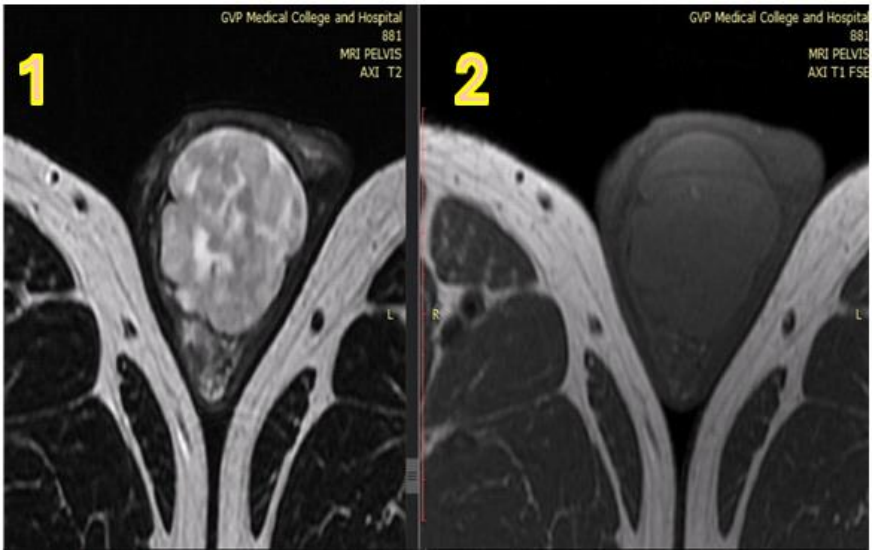

Image 1 (T2 WI) **Image 2 (T1WI)**

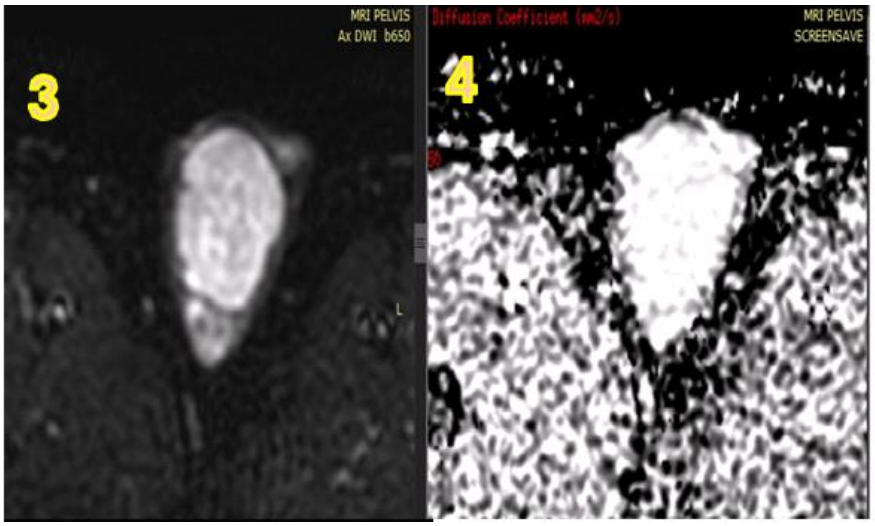

Image 3 (DWI) **Image 4 (ADC)**

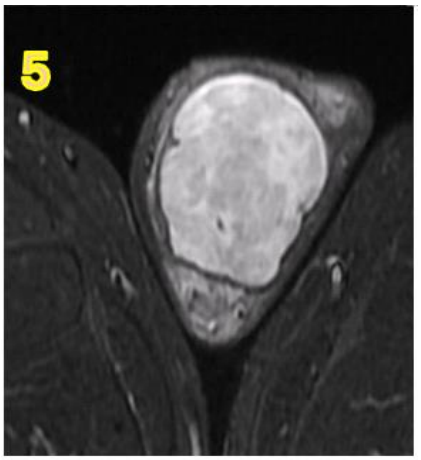

Image 5 (STIR)

T2 heterogeneously hyperintense appearing, T1 isointense, hyperintense signal on ADC (no true DiBusion Restriction).

Left inguinal orchidectomy with spermatic cord and scrotal resection has been done; due to high suspicion of testicular tumor. Gross specimen of the tumor revealed a smooth glistening mass with solid yellow-brown appearance and myxoid areas within. Histopathological examination revealed spindle cells with wavy nuclei and fibrillary cytoplasm, consistent with a diagnosis of schwannoma. Testes and spermatic cord appeared normal.

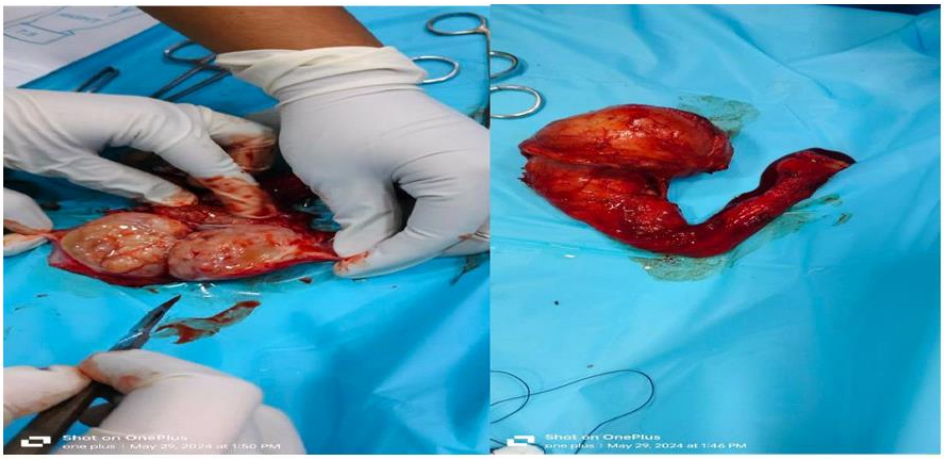

Figure 4

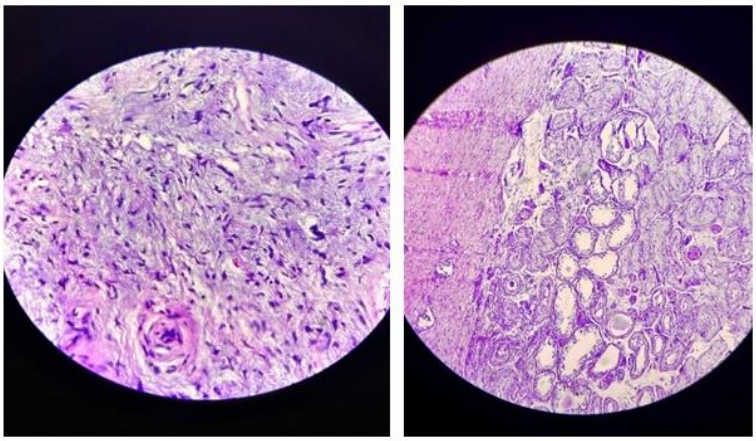

Figure 5

DISCUSSION

Schwannomas typically manifest during the first four decades of life, affecting both genders equally. Most common locations include the head, neck, mediastinum, and retroperitoneum.[11] Less frequently it may occur in sites that encompass the face, neck, scalp, hands, tongue, palate, and larynx. The scrotum is an exceedingly rare site for schwannoma to develop. To date, only a handful of intra-scrotal schwannomas have been documented in the literature.[10,12,13] Hence when a patient presents with a scrotal swelling, schwanomma isn't the leading differential diagnosis in the physician's mind. The non-specific radiological findings also pose a challenge in reaching to this diagnosis. MRI and CT investigations provide better insight into location and spread of the tumor. As discussed above the gold standard in diagnosis is histology of the biopsy or excised specimen. The mainstay of the treatment is surgical excision of the tumor. Although malignancy is rare incomplete excisions may cause recurrence.[14] Regular follow up visits are necessary in cases of large tumors and suspicion of incomplete excision. In a study published by Jiang et al. 5 cases of genital schwanommas were reviewed. Four cases were cured by simple excision and 1 patient with malignant testis schwannoma died of recurrence 1 year after surgery.[15] The follow up period ranged from 2 to 6 years and the mean was 4.5 years.[15] Hence regular follow up is necessary to identify any recurrence. This patient has been followed up for a period of one year, and no indication of recurrence has been observed.

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