Scrotal extratesticular schwannoma: A common tumor at an uncommon location

Schwannoma is a benign encapsulated neoplasm of nerve sheath developing from Schwann cells. Most schwannomas occur in the head and neck region, mediastinum, and retroperitoneum. Rarely, have they been reported in hand, tongue, palate, and larynx. Schwannomas may occur sporadically or in association with neurofibromatosis.\(^1\)\(^2\) The scrotum is an extremely unusual site for developing a schwannoma with less than ten reports in the literature.\(^1\)\(^4\)

A 23-year-old man presented with a history of slowly growing, left scrotal swelling of 2 months duration. Examination revealed an oval, firm mass, measuring 2 cm × 1.8 cm in size in the left scrotum not attached to the testis. There was no inguinal lymphadenopathy. Ultrasonography (USG) revealed bilateral mild hydrocele and a well-defined hypoechoic lesion [Figure 1] measuring approximately 1.49 × 1.51 × 1.86 cm in size in the left hemiscrotal sac wall outside tunica vaginalis showing flow on color Doppler. A clinical differential diagnosis of adnexal tumor of skin or tuberculosis was considered. Fine needle aspiration cytology (FNAC) was advised, however, repeated attempts yielded scant acellular material. Excision biopsy was performed under local anesthesia.

Grossly, there was a globular, encapsulated, gray brown soft tissue mass measuring 2 × 1.6 × 1.5 cm with a homogenous, gray white, solid cut surface [Figure 1, inset]. Histopathologically, the tumor was encapsulated and composed of spindle cells with poorly-defined cytoplasm and elongated, wavy nuclei. The nuclei were palisading and arranged in parallel rows with intervening eosinophilic cytoplasm giving a typical appearance of Verocay bodies [Figure 2a]. The classical cellular (Antoni A) areas interspersed with myxomatous hypocellular (Antoni B) areas were seen. Positive immunohistochemistry for S100 further confirmed the diagnosis of intrascrotal extratesticular schwannoma [Figure 2b].

Scrotal masses need to be accurately diagnosed to prevent therapeutic mismanagement. Among various intrascrotal extratesticular lesions, schwannomas have been reported by few authors. Scrotal schwannoma always pose a diagnostic challenge for clinicians as the list of differential diagnosis is long including adenomatoid tumor, leiomyoma, tubercular lesion, and epididymal cyst.\(^3\) Radiological investigations (USG/CT/MRI) play a role in establishing the size, extratesticular location, local involvement, and distant spread of the lesion; however, they lack specific findings which can aid in diagnosing a schwannoma.\(^4\) The present case was clinically suspected to be a skin appendageal tumor while the radiologic diagnosis was that of a paratesticular tumor.

FNAC is a very useful preoperative diagnostic tool. Well-documented cytological features for the diagnosis of a schwannoma include spindle, wavy nuclei, and Verocay bodies.\(^5\)\(^6\) Bhanvadia \textit{et al.}\(^5\) reported cytological features of a case of scrotal schwannoma where smears revealed tightly cohesive tissue fragments with variable cellularity, exhibiting slender spindle cells showing dark, wavy nuclei along with few clusters showing palisades of nuclei at the margin of the pinkish material simulating Verocay bodies. Domanski \textit{et al.}\(^6\) evaluated 116 cases of schwannomas using FNAC and described

\[\text{Letter}\]

\[\text{Figure 1: (a) Ultrasound revealed mild hydrocele and a well-defined hypoechoic lesion (black arrow) in left hemiscrotal sac wall outside tunica vaginalis, (b) a globular, encapsulated, gray brown soft tissue mass with a homogenous, gray white, solid cut surface}\]

\[\text{Figure 2: Photomicrographs showed (a) Histopathologically, the tumor was encapsulated and composed of spindle cells (H&E, 40X), (b) Tumor composed of spindle cells with elongated, wavy nuclei. The nuclei were palisaded and arranged in parallel rows with intervening eosinophilic cytoplasm in a typical appearance as Verocay bodies (black arrows) (H&E, 400X), inset-Positive Immunostaining for S100}\]
the classical cytomorphological features – spindle, wavy nuclei and Verocay bodies, hypocellular myxomatous material (corresponding to type B areas), and cellular areas with nuclear palisades along the fibrillary material (Antoni A areas). Several authors[6‑8] have also observed small round cells with variable amount of cytoplasm arranged in compact nests or short cords. The diagnosis of the usual type of schwannoma is easy. Histopathology remains the gold standard for definitive diagnosis of schwannoma. The microscopic appearance of schwannoma is relatively characteristic, with two specific patterns – Antoni A areas (hypercellular, Verocay bodies) and Antoni B areas (hypocellular). Immunohistochemistry plays an adjunctive role with positive immunostaining for S‑100 protein and negative staining for CD‑34 favoring schwannoma.[2‑4]

Unfortunately, in the present case, FNAC did not provide diagnostic material. The diagnosis was established by the characteristic histopathological features as well as positive S‑100 immunostaining. Surgical excision is the treatment of choice. Although schwannoma is benign, recurrence may occur in large and incompletely excised lesions. Malignant transformation is exceedingly rare.[2‑4]

To conclude, schwannoma, albeit rare, should be considered in the differential diagnosis of intrascrotal and extratesticular masses. Neither radiology (it can only confirm the paratesticular localization of the tumor) nor serum markers aid in diagnosis. The diagnosis can only be confirmed by histopathology and immunohistochemistry.

Declaration of patient consent
The authors certify that appropriate patient consent was obtained.

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Conflicts of interest
There are no conflicts of interest.

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