Andrology and fertility

Intra-scrotal extra-testicular schwannoma: A case report and literature review

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ABSTRACT

Intra-scrotal schwannoma is a rare neoplasm and a few reports were describing this entity in the literature and mostly difficult to be diagnosed pre operatively(1) We recently treated a case of intra-scrotal extra-testicular schwannoma which was discovered in a patient with history of painless scrotal lesion for 5 years, paratesticular lesion excision was done which was result as schwannoma tissue. follow up with US scrotum was unremarkable for the patient. surgical excision will provide diagnostic and therapeutic goals. Even tough recurrence is rare a urologist should take care to ensure complete surgical resection.

Background

Para-testicular tumors are unusual neoplasms, the anatomical structures of para-testicular areas include spermatic cord, ductus deferens, epididymis, tunica vaginalis, lymphatics, vessels and other supportive tissues. The most common tumors that arise from the para-testicular tissue are benign neoplasms, such as leiomyoma, lipoma and adenomatoid tumor.

Intrascrotal schwannoma is a rare neoplasm and a few reports were describing this entity in the literature and mostly difficult to be diagnosed pre operatively. We recently treated a case of intra-scrotal extra-testicular schwannoma which was discovered in a patient with history of painless scrotal lesion for 5 years, here we describe the medical history, radiological, and histopathological finding.

Case presentation

A 38 year old male otherwise healthy presented with history of left scrotal swelling since 5 years. It is painless and does not change on size. Physical examination revealed left firm, mobile, non-tender upper scrotal mass without skin changes.

Ultrasound showed normal testicles and epididymis with small left epididymal head cyst measuring 0.8 x 0.5 x 0.8 cm, and an extratesticular scrotal lesion lateral to the left testicle measuring 2.2 x 1.5 x 2.3 cm with mixed echogenicity and internal vascularity (Fig. 1). MRI demonstrated Left para-testicular well defined small mass developed outside the spermatic cord and it measures 2.3 cm which showed low signal intensity in T1 and heterogeneous signal intensity in T2 mostly low signal intensity and showed significant intense heterogeneous enhancement after IV Gadolinium (Fig. 2). The tumor markers were negative.

Surgery was done which was in form of paratesticular lesion excision (testis sparing) and the tumor was sent to pathology department. Specimen consists of a 5.53 g brown white-tan firm lesion measuring 2.5 x 2.5 x 1.3 cm. The outer surface is smooth and glistening. The immunohistochemical studies show that tumor cells are positive for S100 protein, CD34, EMA, ER, PR and PGP9.5 while being negative for Neurofilament protein, GLUT-1, SMA and Desmin (Fig. 3). These findings are consistent with benign peripheral nerve sheath tumor with features of schwannoma. Patient was discharge on the same day of surgery, follow up in the clinic for post operative care was unremarkable. 3 months later US scrotum was unremarkable with no sign
of lesion recurrence.

Discussion

Schwannoma is also known as neurilemoma and neurinoma, it is benign Schwann cells derived-tumor. It can occur in all age groups and affects male and female equally. The majority of schwannoma is sporadic, but it may be associated with neurofibromatosis type 2, schwannomatosis, and meningiomas. More frequently it involves head, neck and limbs. Other frequent sites are posterior mediastinum, retroperitoneum, posterior spinal roots, bone, gastrointestinal tract, pancreas, liver, thyroid, adrenal glands and lymph nodes. Rare sites include penis and vulva. Genital schwannoma lesion usually asymptomatic until they increase which cause mass effect to surrounding tissue, specifically for intrascrotal schwannoma the most common presentation is painless scrotal swelling. As far as our knowledge, less than ten cases of intra scrotal extra testicular schwannoma were described in the literature.

No single radiological technique is diagnostic for schwannoma. On US, schwannoma generally appears as a hypoechoic well-circumscribed. On CT, it appears as well covered homogenous tumor with internal cystic changes which displace adjacent structures without direct invasion. The schwannoma characteristics on MRI include specific signs, target, fascicular and split fat signs. The target sign consists peripherally of high T2 signal, which represent Antoni B, and the low to intermediate T2 signal on the center, which represents Antoni A. Fascicular sign characterized by multiple ring-like structure with hyperintense on periphery, while split fat basically means fat rind surrounds the tumor. However, none of these options have allowed schwannoma to be identified, because there is no pathognomonic finding for this disease.4

Gross appearance, most of schwannomas are circumscribed masses abutting an adjacent nerve. While on histology, it appears as biphasic pattern, composed of Antoni A and Antoni B. Antoni A is a dense, hypercellular area, the cells are arranged in palisades, resulting in alternating parallel bands of nuclear and anuclear areas called Verocay bodies. Whereas Antoni B has more myxoid and hypocellular. The vessels usually are dilated and surrounded by thickened-hyalinized vessels wall. S-100 staining is positive. Based on microscopic appearance, there are several variants include ancient, epithelioid, glandular, and cellular types. Fine needle aspiration is considered not helpful because the cytology specimen cannot obtain the tissue’s architecture that required for diagnosis.2,3,5

The standard therapeutic approach is surgical removal. Incompletely resected lesion are capable of recurrence as mentioned by some authors, although no recurrence was described in the literature. Malignant transformation is rare.

Conclusion

Schwannoma should be considered as one of our differentials on each intrascrotal extra-testicular tumor case. US and MRI are the best radiological modalities for localization of Para testicular lesion. Clinically, para-testicular schwannoma presented as painless progressive swelling and, definitive diagnosis is achieved by histopathological investigation, so surgical excision will provide diagnostic and therapeutic goals. Even

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Fig. 1. Scrotal ultrasound showed an extra-testicular scrotal lesion lateral to the left testicle measuring 2.2 × 1.5 × 2.3 cm with mixed echogenicity.

Fig. 2. MRI demonstrated Left paratesticular well defined small mass developed outside the spermatic cord and it measures 2.3 cm.

Fig. 3. A: Hematoxylin and eosin stained section showing bland spindle cell proliferation with relatively hypercellular and hypocellular areas, blood vessels with vascular wall hyalinization and mild perivascular lymphocytic infiltrate. B: Immunohistochemical study with anti-S100 protein showing positive staining.
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