A RARE CASE OF ANCIENT SCHWANNOMA OF SCROTUM

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Abstract
The aim of the article is to present a rare case of ancient variant of scrotal schwannoma in a 26-year old male with immunohistochemical confirmation. Scrotal schwannoma poses a diagnostic challenge to urologists. The "ancient" variant of schwannoma is a rare subtype of a benign encapsulated neoplasm of the nerve sheath. A review of current literature has revealed several reported sites but few in the scrotum.
Keywords: Scrotum, Ancient Schwannoma, Immunohistochemistry

1. Introduction:

Schwannoma is a benign tumour arising from Schwann cells of the peripheral nerve sheath. Histologic predominance of degenerative findings is typical of a rare variant called "ancient" schwannoma. Scrotal localization of these tumours is quite rare.[1] Even more exceptional is development of ancient schwannoma in this region, which we report here.

2. Case summary:

A 26-year old man presented to urology OPD with a six month history of an asymptomatic left sided scrotal swelling **(Fig.1)**. There is no significant medical or family history. The mid-scrotally located mass measured 3 × 2cms and was nodular, hard and clinically separate from the testes. It was not attached to the scrotal skin or other underlying structures. No Lymphadenopathy. Clinically testicular neoplasm was suspected.

Ultrasonography showed extra-testicular, nodular mass in the midline of the scrotum. The nodule was located behind the left testis and it was predominantly solid with few cystic areas. Both testes appeared normal. There was no evidence of distant or nodal metastases. Fine needle aspiration cytology was inconclusive. So, surgical excision with partial scrotectomy was undertaken for removal of the mass and for definitive diagnosis. At resection, the tumour appeared to be superficial to the tunica vaginalis, testes and corpus spongiosum.

3. Pathology:

We received a single nodular partly skin covered mass measuring 2.5x2cms. On cut surface a well encapsulated, well circumscribed, grey-white mass **(Fig. 2)** was seen, which was predominantly solid. Microscopically **(Fig. 3-4),** proliferation of spindled shaped cells with fibrillary cytoplasm was seen, with dense fibrous bands arranging the cells into nodules. In some cells, marked nuclear hyperchromatism and atypia were seen **(Fig. 5)**. Mitoses were not present. Within the lesion, cellular areas were interspersed with looser myxoid and cystic areas. Blood vessels with thickened hyalinized walls **(Fig. 6)** were noted. Staining for S100 protein was positive **(Fig. 7)** in the tumour cells. The margins of this specimen were free from tumour.

So based on location, gross morphology, histopathology and immuno-histochemistry, a diagnosis of ancient schwannoma of scrotum was made. Patient is under follow up and is keeping fine.

4. Discussion:

Schwannoma is a benign encapsulated neoplasm derived from schwann cells of the nerve sheath. The exact incidence of schwannoma is unknown, but they are rare. These benign tumours are found in all age groups but are more common in the first four decades and affect both sexes equally. They have been associated with neurofibromatosis or may arise sporadically. The microscopic appearance of schwannoma is distinctive, with two recognisable patterns.[2] Antoni A areas are composed of compacted spindle cells often arranged in palisades or in an organoid arrangement (Verocay bodies). Antoni B areas consist of tumour cells suspended in a myxomatous matrix that may appear microcystic.[3]

Based on appearances several variants of schwannomas have been observed, including cellular, glandular, epithelioid and ancient types, and all exhibit benign features with benign

progression. Cellular schwannomas are almost exclusively composed of Antoni A areas but lack Verocay bodies. The glandular and epithelioid variants compose of epithelioid areas and glandular component, respectively, to acquire their descriptive names.[3]

Ancient schwannomas show bizarre hyperchromatic nuclei without mitoses. To the unwary, these features can lead to an erroneous diagnosis of malignancy, although the very low mitotic activity should allow these tumours to be distinguished from malignant nerve sheath tumours.[3]

A literature review showed that whilst ancient schwannomas are rare, most cases occur in the head and neck region (trigeminal nerve, facial nerve, vestibular nerve, vagus nerve, parotid, thyroid, vocal cord, floor of mouth, orbit and infra-temporal fossa). Other less common sites include the extremities, mediastinum, thorax , retro- peritoneum , pancreas and pelvis . Scrotal schwannomas have been infrequently described in the medical literature. [4,5,6] However, we could find only one case reference pertaining to the ancient variant in the scrotal region.[3]

Schwannomas pose a difficult diagnostic challenge to urologists because radiological findings are often non-specific [7]. Ultrasonography can differentiate between solid and cystic tumours. Computerized Tomography (CT) can be helpful in determining the size, location, local involvement and distant spread. Magnetic resonance imaging (MRI) provides similarly useful information as CT, but yields better visualisation of the tumour. Fine needle aspirate (FNA) cytology is not often helpful because the tissue architectural information required is not obtainable from cytological specimen [8]. The only gold standard diagnostic investigation is histology of either biopsy or excised specimen. In our case also FNA was inconclusive and histopathology was diagnostic which was confirmed by immunohistochemistry. The expression of high concentrations of S-100 protein in the cytoplasm of tumour spindle cells on immunostaining is useful in the differentiation of a benign schwannoma from a malignant peripheral nerve sheath tumour and from other benign spindle cell tumours. [3]

Surgical excision has remained the mainstay of treatment. Although benign, large and incompletely excised lesions are capable of recurrence; malignant change is exceedingly rare.[6]

In our case, patient is under follow up and keeping fine with no recurrence.

Conclusion:

Ancient schwannoma exhibits pleomorphism without mitosis as the result of cellular degeneration, which can lead to an erroneous diagnosis of malignancy. There is a possibility of malignant transformation of benign schwannoma, so follow-up of the patient is advisable.

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References:

1. Sebastiano Pante et al. Perineal schwannoma. *Can J surg*. 2009; 52(1): E 8-9.
2. Rosai J: Ackerman's surgical pathology. Mosby 8th edition 1995; 2:2264-66.
3. Chan PT, Tripathi S, Low SE and Robinson LQ. Case report – ancient schwannoma of the scrotum. *BMC Urology* 2007; 7(1). PMID:17244372
4. Matsui F, Kobori Y, Takashima H, Amano T, Takemae K: A case of intrascrotal schwannoma. *Acta Urol Jap* 2002; 48:749-51. PMID:12613010
5. Zarate RE, Fernandez GI, Lujan GM, Ortega MP, Berenguer SA: Schwannoma of the scrotum: report of case and review of literature. *Actas Urol Esp* 1997; 21:1012-3. PMID:9494170
6. Safak M, Baltaci S, Ozer G, Turkolmez K, Uluoglu O: Long-term outcome of a patient with intrascrotal extratesticular malignant schwannoma. *Urol Int* 1998; 60:202-4. PMID:9644798
7. Isobe K, Shimizu T, Akahane T, Kato H: Imaging of ancient schwannoma. *AJR Am J Roentgenol* 2004; 183:331-6. PMID:15269020
8. Daneshmand S, Youssefzadeh D, Chamie K, Boswell W, Wu N, Stein JP, Boyd S, Skinner DG: Benign retroperitoneal schwannoma: a case series and review of the literature. *Urology* 2003; 62:993-7. PMID:14665342

Fig – 1) Clinical photograph showing left sided scrotal swelling

Fig – 2) Cut surface – Well encapsulated, soft, grey white mass

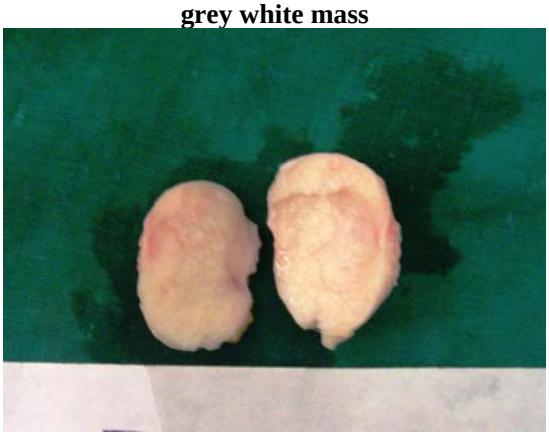

Fig – 3) Antoni 'A' area – Verocay bodies.

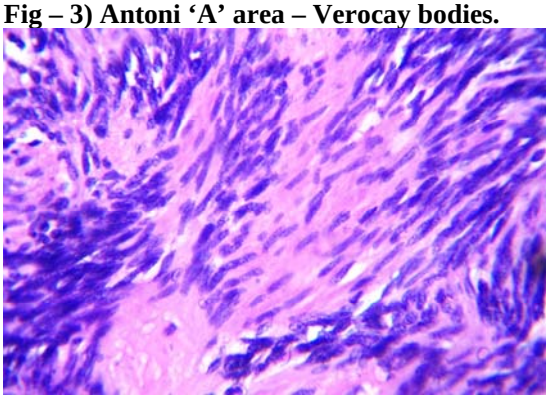

Fig – 4) Antoni 'B' area – Oedematous area

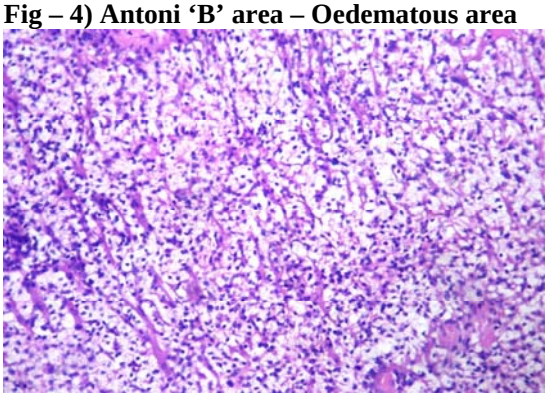

Fig – 5) Cells are Oval to spindle and hyperchromatic – Ancient type

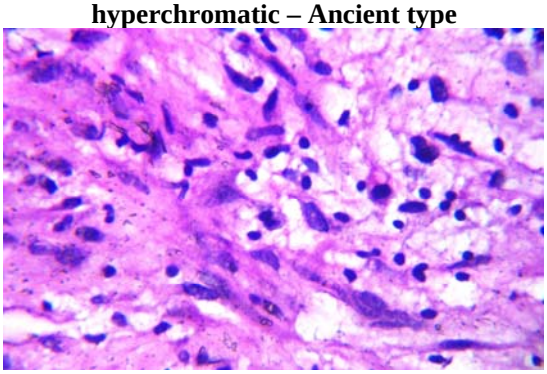

Fig – 6) Hyalinized blood vessel

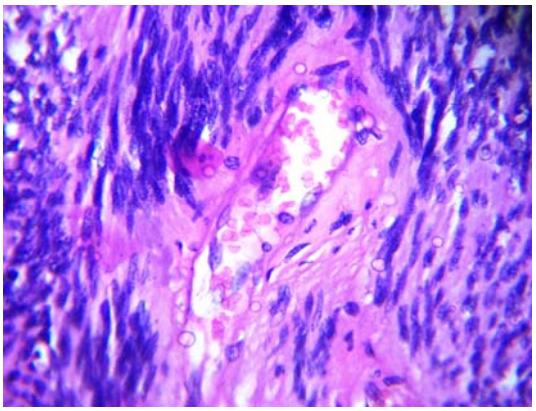

Fig – 7) Immunohistochemistry - Positive for S100