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Perineo-scrotal schwannoma: A rare case report

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Abstract
Schwannoma is a benign tumor arising from schwann cells. Schwannoma arises frequently from the acoustic nerve. In the peripheral nervous system usually found in relation to main nerves of the limbs like flexor aspect near the elbow, wrist or knee or the head and neck. Perineal and scrotal schwannoma is very rare. We report the rare case of 55-year-old man presented with a 4-5 years history of multiple swelling over perineum and right hemiscrotum. MRI reported multiple enhancing solid cystic component post contrast enhancing lesions in perineal region. The clinical suspicion was a skin appendageal tumor or benign lipomatous tumor. Patient underwent a complete excision of the tumor. The histological features were suggestive of perineo-scrotal schwannoma.

Keywords: Perineum, Scrotum, Schwannoma

Introduction
Schwannoma is a benign tumor arising from schwann cells, which normally produce the insulating myelin sheath covering peripheral nerves. Schwannoma is commonly present in around 20–50 years of age [1]. Clinical features associated with schwannoma are not specific to the disease because schwannoma is usually solitary slow growing, non-aggressive benign tumor, and do not show any symptoms until enlarge and compress the surrounding structures [2]. Schwannoma arises frequently from the acoustic nerve. In the peripheral nervous system usually found in relation to main nerves of the limbs like flexor aspect near the elbow, wrist or knee or the head and neck [3]. Perineal and scrotal schwannoma is very rare [4]. Only few cases are reported in literature. Here, we describe a rare and unusual case of perineal and scrotal schwannoma with a complete case report, including clinical, radiological, intraoperative and histopathological findings.

Case Presentation
Clinical history
A 55 year old man presented with complaint of multiple swelling over perineum and right hemiscrotum since 4-5 years. As per given history by patient swelling gradually increased in size and not associated with pain and no history of fever. Physical examination revealed multiple invariable size of palpable, nontender, soft consistency mobile swelling in the perineum and right hemiscrotum, tumor appeared not to be attached to the skin of the perineum and scrotum or the underlying tissues. The abdominal examination and rectal examination findings were normal. The patient did not report any systemic symptoms.

Imaging
Ultrasonography of inguinoscrotal region was done which revealed a large multinodular mass lesion together measuring around 92x44 mm in right scrotum extending up to lower perineum with increase vascularity within. Both testis were normal. To better examine the patient, MRI pelvis was done and MRI reported T1 isointense, T2 predominantly hyperintense nodular elongated lesion with internal solid component involving perineal region midline. Lesion were 2-3 in number, largest one was approximately 5.4 x 2.1 cm. Post contrast study showed inhomogeneous post contrast enhancement, On STIR sequences lesion appeared bright (Figure-1).

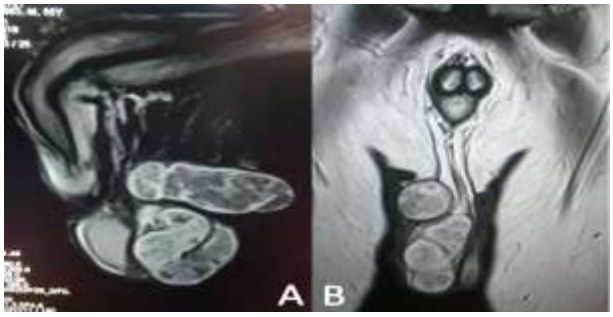

Fig 1A: Post contrast T1 Fatsat sequence showing multinodular lesion in perineal and scrotal region with in homogenous post contrast inhancement. **B:** T2 hetrogenous lesion with multiple hyperintense cystic components within.

Therapeutic approach

Patient underwent a complete excision of the tumor. The patient was placed in a lithotomy position, and surgery was started with an incision over the swelling on the perianal and right scrotal area. Surgical exploration showed a white yellowish encapsulated tumor with elastic consistency, mass was dissected from surrounding tissues, removed in two part measuring 5 × 3.5 cm. and 4 x 3.8 cm (Figure 2). The postoperative course was uneventful.

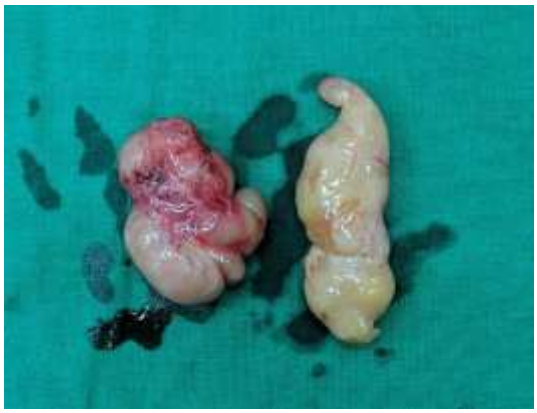

Fig 2: Surgical specimen: white yellowish encapsulated tumor with elastic consistency

Microscopic findings

Well circumscribed encapsulated mass composed of spindle cells. Showed areas of hypercellularity (Antony A) and loose microcytic paucicellular area (Antony B). Varocay bodies were present. Thick hyaline blood vessels were presented within the lesions with no apparent nuclear atypia. Mitosis were negligible (<1/10 HPF). These histological features were suggestive of schwannoma

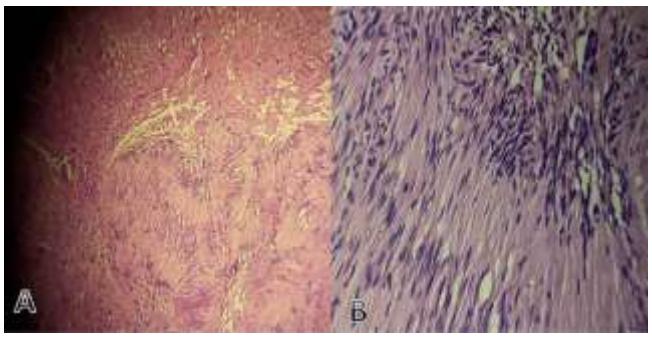

Fig 3: Microscopic appearances- Spindle cells in an Antoni type A area and varocay body

Discussion

A schwannoma is a usually benign nerve sheath tumor composed of Schwann cells, which normally produce the insulating myelin sheath covering peripheral nerves.[5]. Schwannomas may arise from the cranial, spinal and the peripheral nerves of the body. Schwannoma of the head and neck are a fairly common occurrence, most common of these is a vestibular schwannoma. Outside the cranial nerves, schwannomas may present on the flexor surfaces of the limbs [4] and the rare sites reported being hands, tongue, palate and the larynx, among all the perineal and scrotal schwannomas are the rarest [6, 7] The rate of reported pelvic schwannomas is 1% [8] Schwannomas are found in all age groups but are common in around 20-40 years of age and affect both sexes equally [5].

Schwannomas are typically slow growing and nonaggressive benign tumor. Therefore, they are usually asymptomatic and discovered as large perineal masses. When symptomatic, patients may present with non-specific pain, palpable mass or rectal dysfunction. Malignant schwannomas are usually large, infiltrating and fast growing tumors [9]. In our case, the clinical suspicion was that of a skin appendageal tumour or benign lipomatous tumor. The differential diagnosis of benign perineal and scrotal tumor includes leiomyoma, lipoma, fibroma or haemangiomas [10]

Pre-operative diagnosis of schwannomas is very difficult and challenging. Radiological findings are often non-specific. Ultrasonography can differentiate between solid and cystic tumours. CT can be helpful in determining the size, location, and local involvement. Magnetic resonance imaging (MRI) provides similarly useful information as CT, but yields better visualisation of the tumour [11]. The MRI characteristics of peripheral and scrotal schwannomas typically include hypointensity or isointensity on T1-weighted images and hyperintensity on T2-weighted images [9]. MRI is also useful for analysing the tumor's relationships with adjacent structures in the perineum. Fine needle aspirate (FNA) cytology is not often helpful because the tissue architectural information required is not obtainable from cytological specimen. The only gold standard diagnostic investigation is histology of either biopsy or excised specimen [11]

The microscopic features of schwannoma have two patterns. 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. Several variants of schwannomas based on appearances have been reported, including *cellular*, *glandular*, *epithelioid* and *ancient* types, and all exhibit benign progression. Cellular schwannomas are almost exclusively composed of Antoni A areas but lack Verocay bodies. The glandular and epitheloid variants composed of epitheloid areas and glandular component, respectively. *Ancient schwannomas* show bizarre hyperchromatic nuclei without mitoses[5] Immunohistochemical analysis is very useful to differentiate schwannomas from other perineal masses, by showing a positive and uniform S-100 staining. Malignant schwannomas are characterized histologically by perineural and intraneural spreads, herniation into the lumina of the vessels and nuclear palisading [9]

Surgical is the standard therapeutic approach. Recurrence after complete surgical excision is rare. Similarly, malignant transformation in a schwannoma is extremely uncommon [12]

Conclusions

Perineal schwannoma is very rare tumors, usually asymptomatic, presents as large masses. MRI is the best modality with which to confirm the localisation of the tumour and local involvement, before surgical removal allows histopathological investigation and definitive diagnosis. Surgery is the standard therapeutic approach. To prevent recurrence, particular care should be taken to ensure complete excision, especially in cases that involve large or multiple masses

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