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Case Report

Radiology Section

Multiple Penile and Pelvic Schwannomas in an Adult Male: A Case Report

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ABSTRACT

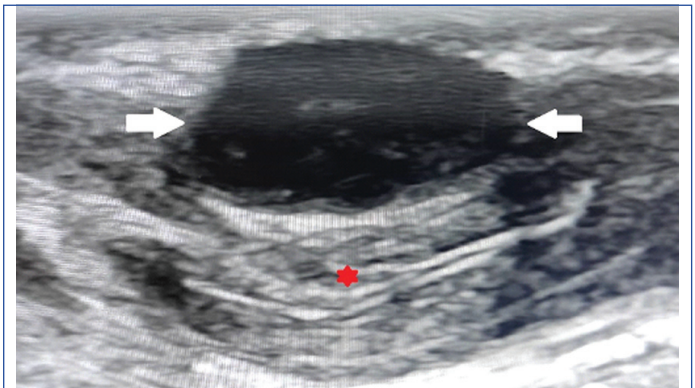
Schwannomas are relatively common benign tumours that gradually increase in size and develop in the peripheral nerve sheaths of adults. However, they are exceptionally rare in the penile and pelvic regions. Hereby, the authors present a case report of an extremely rare case of concomitant multiple schwannomas in the penis and pelvis. A 32-year-old adult male presented with complaints of multiple small swellings on the penile shaft, which were associated with episodes of severe pain. Clinically, there were multiple small, firm to hard, tender nodular swellings on the penile shaft. Ultrasound and Magnetic Resonance Imaging (MRI) revealed six well-marginated lesions in the penile areolar tissue and two lesions along the course of the left sciatic nerve in the left hemipelvis. Surgical excision followed by histopathological evaluation confirmed the diagnosis of multiple schwannomas. Due to their rarity and non specific presentation, most of these lesions are not diagnosed clinically. Therefore, presurgical imaging is necessary to evaluate their anatomical and morphological features. Although imaging findings in schwannomas are not always precise, Histopathological Examination is required for a definitive diagnosis. Histopathology typically shows the mixed presence of Antoni A and Antoni B bodies containing spindle cells arranged tightly and loosely, respectively, in a collagenous matrix. Immunohistochemistry (IHC) typically shows positivity for S-100 protein, which is characteristic of schwannomas. They are also positive for neuron-specific enolase and vimentin, but negative for Cluster of Differentiation (CD) 117 and Smooth Muscle Actin (SMA). Schwannomas are treated by complete surgical excision with preservation of the nerve axon, and they have a good prognosis with a low chance of recurrence.

Keywords: Antoni A and B bodies, Magnetic resonance imaging, Nerve axon, Ultrasound

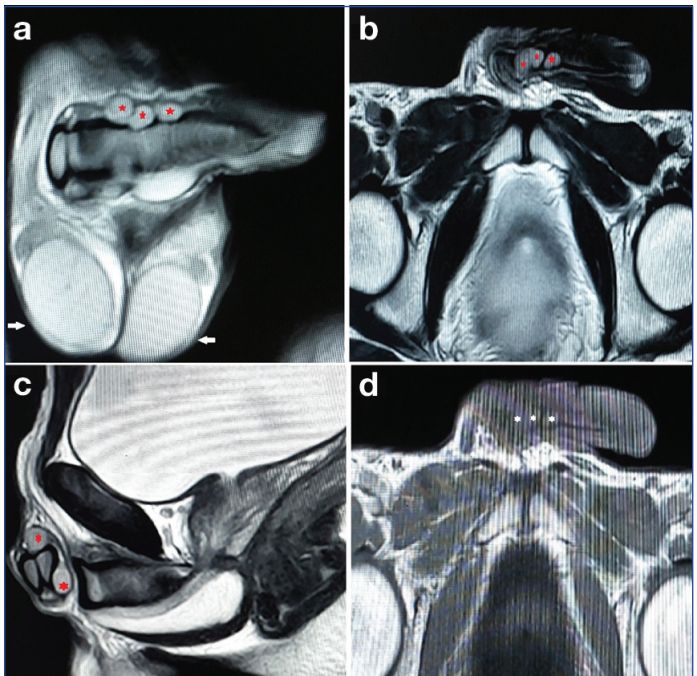
CASE REPORT

A 32-year-old male presented at the outpatient clinic of the Urology Department with complaints of multiple small swellings in the penile shaft that were slowly growing in size and associated with episodes of severe pain at the base of the penis for three months. There was no history of dyspareunia, penile trauma, or instrumentation. Clinical examination revealed multiple small, firm to hard, tender nodular swellings on the shaft of the penis beneath the skin. No skin pigmentation, thickening, or ulceration was observed. There was no evidence of any other palpable lesion or cutaneous markers elsewhere on the body. The patient had no relevant previous medical or surgical history, and there was no familial history of nerve sheath tumours. Routine blood and urine investigations did not reveal any significant findings.

Penile ultrasound showed six small oval-shaped hypoechoic superficial lesions with smooth margins (the largest measuring 6.5×9.5×11 mm) and mild posterior acoustic enhancement in the penile shaft. The ultrasound image of one of these lesions is shown in [Table/Fig-1]. MRI revealed six small well-defined oval-shaped lesions that appeared hyperintense on T2-weighted images and isointense on T1-weighted images in the superficial areolar tissue of the penile shaft [Table/Fig-2a-d]. Based on the imaging features, the possible diagnoses considered were benign tumours of the penis, such as schwannomas, haemangiomas, neurofibromas, and leiomyomas. MRI also revealed two well-marginated lesions that appeared heterogeneously hyperintense on T2-weighted images and isointense on T1-weighted images along the course of the left sciatic nerve. One lesion was located at the left greater sciatic foramen, measuring 21×32×26 mm, and the other was lateral to the

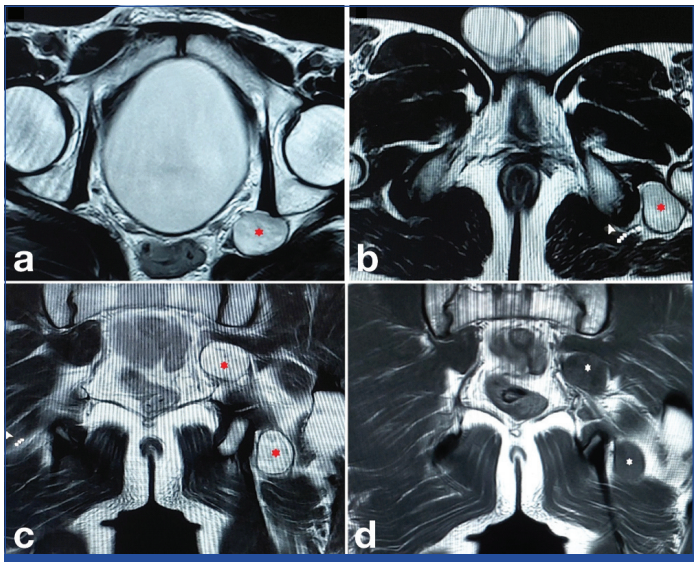


[Table/Fig-1]: Ultrasound image of penis shows a well-defined superficial oval shaped hypoechoic lesion (arrows) on dorsal surface of penile shaft indenting on corpus cavernosum (red asterisk).



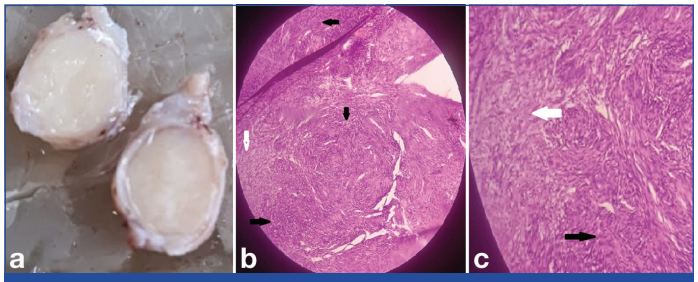
[Table/Fig-2a-d]: MRI images of penis show multiple small schwannomas appearing hyperintense on T2W sequence obtained in multiple planes (red asterisks in a-c) and isointense on T1W image (white asterisks in d).

left ischial tuberosity, indenting on the quadratus femoris muscle, measuring 26×22×32 mm [Table/Fig-3a-d]. No significant inguinal or iliac lymphadenopathy was observed.



[Table/Fig-3a-d]: MRI images of the pelvis show two schwannomas along the course of left sciatic nerve appearing heterogeneously hyperintense on axial T2W image (red asterisks in a,b) and coronal T2W image (red asterisks in c) and isointense on coronal T1W image (white asterisks in d).

Surgical excision of these lesions revealed multiple varying-sized grayish-white nodules in the superficial areolar tissue of the penile shaft and left hemipelvis. Haematoxylin and Eosin (H&E) stain evaluation confirmed the diagnosis of spindle-cell tumours consistent with findings of multiple schwannomas [Table/Fig-4a-c]. Postoperative follow-up after one month revealed a significant resolution of his symptoms.



[Table/Fig-4a-c]: (a) Gross specimen, (b,c) Penile schwannoma (100x magnification image of Haematoxylin and Eosin stain) showing Antoni A regions marked with black arrows and Antoni B regions marked with white arrows.

DISCUSSION

Schwannomas are relatively common benign tumours that gradually increase in size and develop in the peripheral nerve sheaths of adults, although they can occur in any age group and do not have a gender preference [1,2]. Schwannomas originate from Schwann cells in the sheaths of cranial and peripheral nerves [1,2]. In 10% of patients, they are associated with neurofibromatosis, multiple meningiomas, and schwannomatosis [1]. They are linked to Neurofibromatosis type 2 (NF2) gene deletion or alteration and a decrease in Merlin protein [2]. Schwannomas commonly occur in the head, neck, chest, limbs, and retroperitoneum, but rarely involve the genitourinary system (prostate, urinary bladder, testes, seminal vesicles, spermatic cord), and are exceptionally rare in the penile region [2,3]. In present report, we describe an extremely unusual case of multiple schwannomas coexisting in the penis and pelvis, which were detected using ultrasound and MRI, and confirmed through histopathological examination following surgical excision. To the best of authors' knowledge, this is the first reported case of its kind in the literature.

Typically, schwannomas are solitary tumours; however, multiple schwannomas are reported in up to 30% of cases involving the penis [1]. They are usually asymptomatic and only cause neurological symptoms when they become large enough to compress nerves [1]. Schwannomas in the penis are more commonly found in the dorsal aspect, as the penis is supplied by dorsal nerves. Painful schwannomas in the penile region are rare, but may present with symptoms such as dyspareunia, sexual dysfunction, and urinary tract obstruction [2,4-6]. Other benign tumours reported in the penis include neurofibroma, haemangioma, lipoma, Peyronie's disease, leiomyoma, and fibroma [4,6,7]. Neurofibroma can cause penile enlargement, with or without the presence of café-au-lait skin spots [8]. Haemangioma appears isoechoic on ultrasound and shows internal vascularity on Doppler study [7]. Lipoma, arising from adipocytes, appears iso-hyperechoic on ultrasound and is extremely rare in the penis [9]. Peyronie's disease is characterised by fibrotic thickening of the tunica albuginea, with plaques and calcifications visible on imaging [10]. Penile leiomyoma and fibroma appear as well-defined lesions with isoechoic characteristics and no internal colour flow [11]. Malignant tumours in the penile region, such as squamous cell carcinoma, leiomyosarcoma, Kaposi's sarcoma, and epithelioid sarcoma, are extremely rare [4,6,11-13].

Schwannomas occurring in the pelvic cavity are exceptionally rare and typically arise from the sacral nerve or hypogastric nerve plexus [3]. While schwannomas in the extremities are uncommon, they can cause lower limb pain and sciatica due to compression of sciatic nerve axons [14]. These tumours are usually benign and have a low chance of malignant transformation and recurrence [3]. Differentiating pelvic and gynecological tumours, such as ovarian tumours, abscesses, hematomas, cystic tumours, and sarcomas, from schwannomas can be challenging [3]. Due to their rarity and nonspecific presentation, many of these lesions are not diagnosed clinically, highlighting the importance of presurgical imaging to evaluate their anatomical and morphological features. Imaging plays a crucial role in identifying the precise location, margins, presence of solid or cystic components, relationship with adjacent tissues, and local or distant spread of the tumours.

Penile and pelvic schwannomas typically appear as well-defined hypoechoic lesions with increased flow signals on ultrasound imaging [6]. On MRI, they appear hypointense to isointense on T1-weighted sequences and heterogeneously hyperintense on T2-weighted sequences. Smaller tumours show homogeneous contrast enhancement, while larger lesions exhibit heterogeneous enhancement [3,6]. In present case report, the patient presented with multiple small penile lesions and an atypical clinical symptom of severe pain at the base of the penis, necessitating imaging assessment. Ultrasound of the penis did not provide a definitive diagnosis; therefore, MRI was performed. The MRI not only revealed multiple well-defined superficial masses in the penis but also identified two smoothly marginated masses along the course of the left sciatic nerve, suggesting the possibility of nerve sheath tumours.

As imaging findings in schwannomas are not always precise, histopathological examination is required for a definitive diagnosis. Schwannomas are well-demarcated lesions with well-defined capsules. They are characterised by the presence of spindle cells arranged tightly (Antoni A) and loosely (Antoni B) in a collagenous matrix. The tumour cells have curved vesicular nuclei with eosinophilic cytoplasm [14]. Various histological subtypes of schwannomas have been described, including cystic, cellular, plexiform, epithelioid, psammomatous, melanotic, ancient, and those with pseudoglandular components [15]. Immunohistochemistry typically shows positivity for S-100 protein, neuron-specific enolase, and vimentin, while being negative for CD117 and smooth muscle actin (SMA) [3]. Malignant transformation is rare but can be detected on histopathology. Yeh CJ et al., previously reported two cases of painless penile schwannoma, one with a small solitary lesion and the other with multiple lesions [1]. Both patients underwent simple

surgical resection with no postoperative complications. Multiple penile schwannomas were also reported by Song Z et al., in a middle-aged adult male, with no evidence of malignant transformation and no recurrence during a three-year follow-up [2]. Dau MHT et al., previously reported a case of a solitary pelvic schwannoma in a 37-year-old male who presented with left-side abdominal pain and a left abdominal mass [3]. The patient underwent surgical excision, and the mass was confirmed as a schwannoma through pathology and immunohistochemistry. Schwannomas are typically treated with complete surgical excision while preserving the nerve axon. They have a good prognosis and a low risk of recurrence [1].

CONCLUSION(S)

Schwannomas are exceptionally rare in the penile and pelvic regions, making clinical and imaging diagnosis challenging. Imaging plays a crucial role in localising and characterising these tumours before surgery. Complete surgical excision is the preferred treatment, and it offers a favourable prognosis with minimal risk of malignancy and recurrence.

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