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Case Report

Pudendal Schwannoma; A Rare Reason for Scrotal and Penile Pain

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Abstract: Schwannomas are benign nerve sheath tumors that usually arise from cranial and periferal nerves. Pelvic region schwannomas are rare tumors and only a few cases have been reported in literature. Pudendal nerve schwannomas usually present as a cause of ipsilateral scrotal, penile and perineal region pain. We report the case of a 66-year-old man referred for worsening pain of 6 years duration involving the right scrotal and right penile shaft. After a series of repeated scrotal and penile ultrasonographies (USG), lumbar and brain magnetic resonance (MR) imaging and also blood tests, MR imaging of the pelvic region detected a well-circumscribed small pelvic wall tumor on the pathway of pudendal nerve. Based on the location, imaging characteristics and comparing to the cases reported in literature, we considered this tumor as a pudendal schwannoma. The most commonly described surgical approach for pelvic schwannomas has been open median laparotomy with transperitoneal dissection. The patient refused surgery, for this reason for treatment and control of the pain he underwent nerve block at anesthesiology clinic. At the 4-week follow-up, the patient described a significant decrease in pain.

Keywords: Magnetic resonance imaging, nerve sheath tumor, pain, pudental nerve, schwannoma.

INTRODUCTION

Schwannomas are benign nerve sheath tumors composed of Schwann cells. They comprise 5% of all benign soft tissue tumours [1, 2]. They have a tendency to involve the cranial nerves and the peripheral nervous system of the neck, mediastinum and extremities. Benign pelvic floor schwannomas are rare and particularly unusual. There are 25 reported cases [3]. Pudendal nerve schwannomas usually present as a cause of ipsilateral scrotal, penile and perineal region pain. On Magnetic Resonance (MR) imaging, neurogenic tumours are seen as a fusiform-shaped mass with tapered ends that are low-to-intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images. The diagnostic confidence is increased if the mass arises from the anatomic location of a known nerve. The signal intensity on T2-weighted images may be either homogeneously hyperintense or show a characteristic MR target sign that consists of high signal intensity in the periphery and low signal intensity in the central region of the lesion [4]. Deep location in an anatomically complex region raises issues concerning the strategy of treatment approach. To date the most commonly described surgical approach for pelvic schwannomas has been open median laparotomy with transperitoneal dissection [3].

We report a case of a pudendal schwannoma detected with MR imaging.

CASE REPORT

A 66-year-old male was referred in 2015 to our department for worsening right-sided perineal, scrotal and penile pain with a 6-year history of pain in these regions. Presumed reasons for this pain were testicular or scrotal pathologies. To rule out these pathologies scrotal and penile gray scale and doppler ultrasonography (USG) were performed and no pathologies were detected. Blood and urine tests were

normal. The other predominant symptoms were paresthesias involving the right testicle and right penile shaft. His medical history revealed no significant illnesses. He had no previous history of surgery, no history of malignancy, diabetes or other disease. On physical examination, there was no evidence of cutaneous lesions or palpable lymphadenopathy and his abdomen was soft, non-tender and without any palpable masses. The neurological examination highlighted a hypoesthesia of the right penile shaft. The rest of the neurological exam was normal. To rule out possible pelvic pathologies pelvic MR imaging was done.

Magnetic resonance (MR) imaging of the pelvis revealed an extra-peritoneal 14x14x9mm soft tissue mass along the right posterior pelvic side wall above the muscular pelvic floor. The lesion was well-circumscribed; oval shaped and was located in the neighboring of the sacral plexus branches as they exited through the sciatic notch. It was also located lateral to the rectum. The mass was iso-intense to muscle on T1 and hyper-intense on Proton Density Weighted-Spectral Attenuated Inversion Recovery (PDW-SPAIR) sequences (Fig. 1). There was a slight heterogeneous enhancement on fat saturated enhanced T1 sequences (Fig. 2). No lymphadenopathies were detected near this region. There was no previous pelvic imaging to compare the size of the lesion. Based on the location, imaging characteristics and comparing to the cases reported in literature, we considered this tumor as a pudendal schwannoma. Also no evidence of metastasis was found on the other radiologic evaluations.

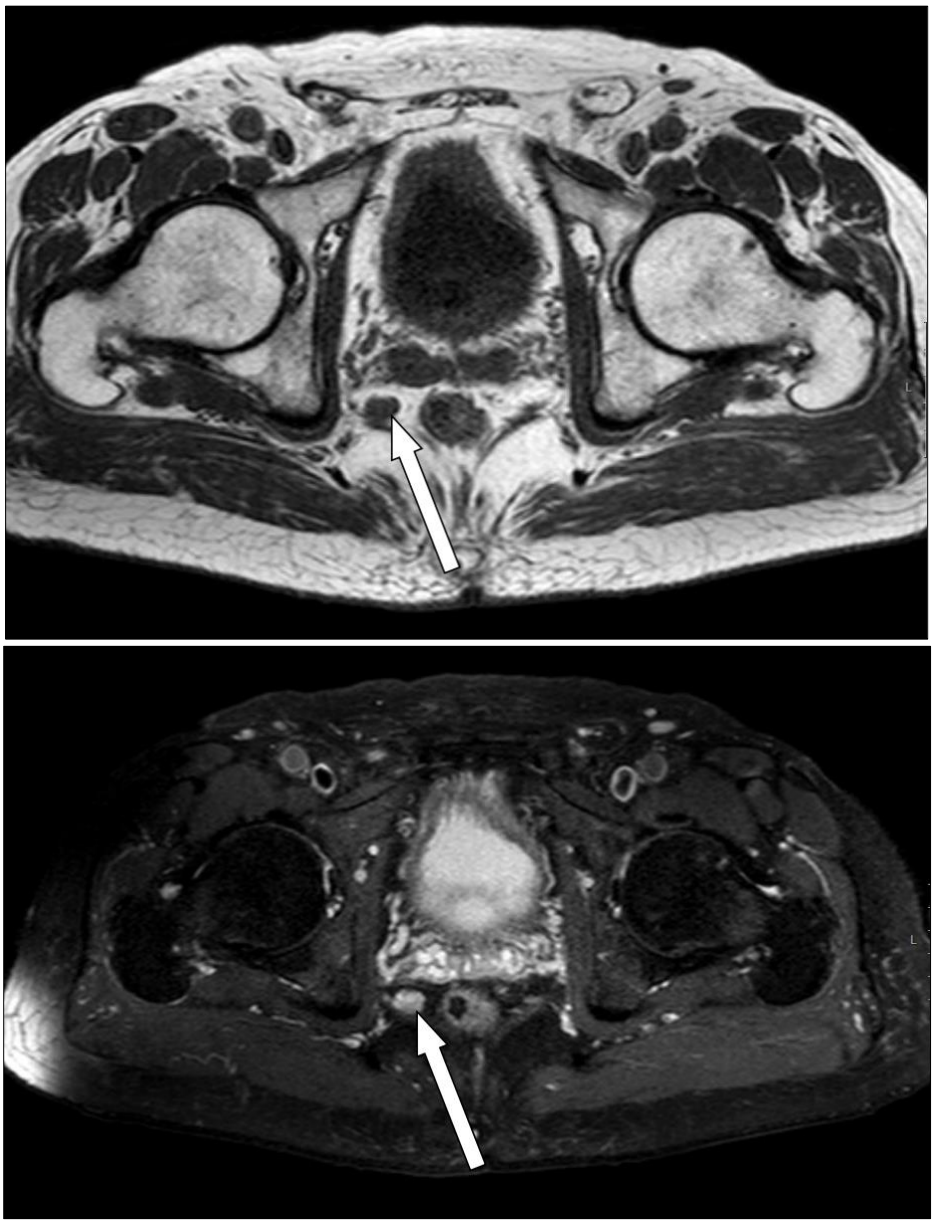

Fig. 1: MR images of the abdomen and pelvis show the extra-peritoneal well-circumscribed soft tissue mass along the right posterior pelvic sidewall above the muscular pelvic floor (white arrows). Above T1 weighted image, below PDW-SPAIR image.

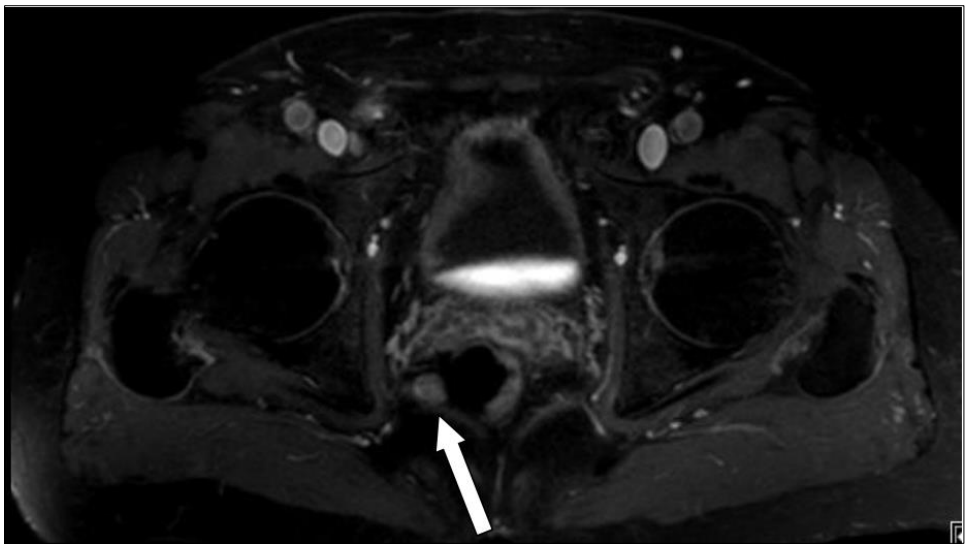

Fig. 2: A MR image of the abdomen and pelvis shows the extra-peritoneal well-circumscribed soft tissue mass along the right posterior pelvic sidewall above the muscular pelvic floor. This figure shows a slight heterogeneous enhancement of the lesion (white arrow)

The patient referred to neurosurgery clinic. For histopathologic diagnosis, department of neurosurgery offered surgery to the patient but he refused surgery. For this reason patient referred to algology clinic at anesthesiology department for symptomatic treatment of the pain. At the patient's request, and with his informed consent, algology department performed nerve block to the patient. At the 4-week follow-up, the patient described a significant decrease in pain.

One year later the follow-up, pelvic MR imaging showed no change in the size and signal characteristics of the lesion.

DISCUSSION

The pudendal nerve arises from sacral nerves S2–S4 and, after passing between the piriformis and coccygeus muscles, leaves the pelvis through the distal part of the greater sciatic foramen. This nerve accompanies the pudendal vessels along the lateral wall of the ischiorectal fossa within Alcock's canal. This canal, also called the pudendal canal, is formed by the obturator internus fascia. Along its course, the pudendal nerve gives rise to the inferior rectal nerve, the perineal nerve, and the dorsal nerve to the penis or clitoris. Accordingly, it supplies the somatic anal and urethral sphincters, as well as penile and clitoral musculature [2-6].

Functionally, the pudendal nerve ensures the sensitivity of the perineum teguments (the glans penis, clitoris, scrotum, labia majora, skin of the central fibrous perineal body, and anus). It also gives the motor innervation of the erector muscles and the striated sphincters, therefore controlling erection and the voluntary actions of the sphincters. It is mostly involved in sexual activity, micturition and defecation [2-6].

Schwannomas most commonly affect patients 20–40 years of age and constitute about 5% of benign soft-tissue neoplasms [7,8]. Males and females are affected equally. The commonly involved sites include the spinal and sympathetic roots of the head and neck, as well as nerves in the flexor surfaces of the upper and lower extremities (in particular, the ulnar and peroneal nerves). On MR imaging, schwannomas show low-to-intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images. The diagnostic confidence is increased if the mass arises from the anatomic location of a known nerve. The signal intensity on T2-weighted images may be either homogeneously hyperintense or show a characteristic MR target sign that consists of high signal intensity in the periphery and low signal intensity in the central region of the lesion [4,7,8]. The primary treatment is surgical excision. Various laparoscopic and open surgical techniques have been reported in literature [2,5,9-12].

Pelvic region schwannomas are extremely rare benign regions. In our case, the patient had history of worsened pain in the ride side of penile shaft, scrotum and perineal region which is innervated by pudendal nerve. On MR imaging a well circumscribed extraperitoneal 14x14x9mm soft tissue mass along the right posterior pelvic side wall above the muscular pelvic floor was detected. Knowing the symptoms of the patient and due to the location of the mass that corresponds to the pudendal nerve pathway and also signal characteristics we considered this lesion as a pudendal schwannoma. But the patient refused surgery thus we couldn't prove this diagnosis histopathologicaly. But the improvement of clinical symptoms after nerve block treatment and characteristic radiological findings were the evidences fort this prediction. Also the appearance of a well-

circumscribed, encapsulated, non-locally invasive, slow-growing tumor with no regional lymphadenopathy allowed us to confidently differentiate it from the malignant neoplasms.

A retrospective study of 25 cases of retroperitoneal tumors found a significant correlation between irregular margins and malignancy [3]. The authors developed a retroperitoneal tumor scoring system to distinguish primary retroperitoneal benign tumors from their malignant counterparts based on 5 criteria: (1) maximum diameter equal to or larger than 5.5 cm, (2) presence of symptoms, (3) absence of calcification, (4) presence of irregular margins and (5) presence of cystic degeneration or necrosis. They found a significant correlation between the incidence of malignant tumors and the total retroperitoneal tumor score [13].

For this patient, our diagnosis was a pudendal schwanomma. Other benign tumors that could have been suspected include retroperitoneal pseudocyst, abscess and lymphocele, in which case the tumor would have been almost entirely cystic, which is rarely the case for schwannomas and was not the case here. And in the case of malignancy the mass would be fast-growing with irregular margins and would show local invasion or metastasis. If there was associated lymphadenopathy, we would have considered neurofibroma and lymphoma as possible differential diagnoses.

We aimed to report this case of pudendal schwannoma to make contribution to the radiology literature about this rarely seen periferal nerve sheath tumor, which manifests as a reason of penile, scrotal and perineal region pain.

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