

Oncology

Periprostatic Perineurioma, Description of a Case With Unusual Localization



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ABSTRACT

Perineurioma is a rare entity, it is a benign peripheral nerve sheath tumor entirely composed of perineurial cells. A 62-year-old male patient was admitted to our hospital, suffering from scrotal and pelvic pain combined with a severe and continuous pain in his right thigh. A transrectal ultrasound revealed a periprostatic oval lesion of about 5 cm in maximum diameter. A sovrappubic laparotomy was performed with a complete tumor excision. The morphological and immunohistochemical data were most consistent with the diagnosis of perineurioma.

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Introduction

Perineurioma is a rare lesion and represents approximately 1% of all soft tissue neoplasms which is nearly always benign peripheral nerve sheath tumor and are entirely composed of perineurial cells. Though intraneural and mucosal types also exist.

About 200 cases have been reported and these tumors are slightly more common in females than males and occur over a wide age range, with a peak in middle-aged adults, with children being rarely affected.

These tumors are mostly sporadic, commonly arising in the lower limbs, followed by the upper limbs and trunk. The head and neck region, visceral organs, and central body areas are almost never affected and periprostatic lesions have never been described.

Case presentation

A 62-year-old male patient was admitted to our hospital suffering from scrotal and pelvic pain combined with a severe and continuous pain in his right thigh, and sometimes in the left thigh.

The clinical examination showed a prostate gland of a normal morphology and size, with negative digital rectal examination and

PSA within normal limits, however, a transrectal ultrasound revealed a periprostatic oval lesion of about 5 cm in maximum diameter (Fig. 1). An abdominal CT and MRN confirmed the presence of a neoplasm, whereas a colonoscopy was completely negative.

The lesion was biopsied twice using a fine needle specifically for prostatic biopsies. Both the first and the second biopsy were characterized by tiny, filamentous and friable fragments; they consisted of small, spindle cells embedded in a myxoid stroma, negative for S100, EMA result is doubtful, atypia absent, proliferative activity absent. They were typical of a myxoid benign lesion with spindle cells.

Surgical intervention was requested by the patient because of persistent pain in his right thigh.

A sovrappubic laparotomy was performed with a complete tumor excision. The surgery consisted of a navel–pubic incision with the opening of the rectus sheath. The external iliac vein and the right obturator nerve were identified after the incision of the right endopelvic fascia. The neoplasm presented a tight-elastic consistency and contracted relationship with the prostatic apex, urethra, the elevator ani muscle and rectum muscles. The mass was extracted without damaging all the structures described above. Evaluation of the integrity of the urethral bladder was done with methylene blue dye from the bladder catheter. Drainage was carried out, suture in layers and 48 months after surgery the patient is still disease free.

On gross examination (Fig. 2), the surgical specimen consisted of a polylobate and pseudoencapsulated lesion; it measured 3 × 2.5 cm

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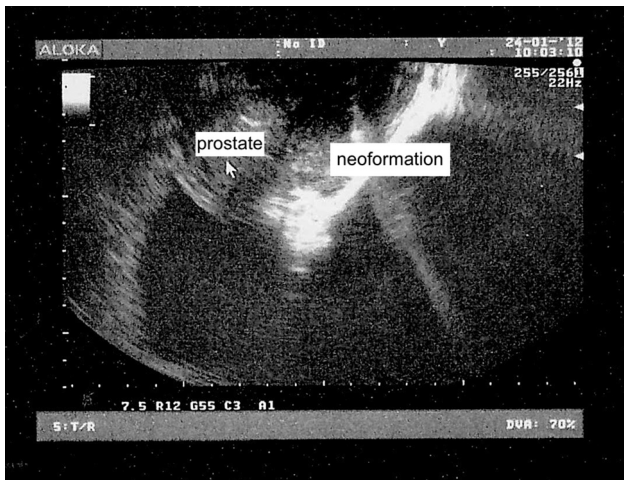


Figure 1. Transrectal prostatic ultrasonography (showing the prostate and the adjacent neoformation).

in size and weighed 55 g. The cut surface was yellowish and elastic in consistency.

Microscopically (Fig. 3) we observed a spindle cell proliferation, consisting of elements with bipolar pale eosinophilic cytoplasm and tapering nuclei, within a myxoid stroma, these cells presented immunoreactivity for EMA and CD34 (supporting perineurial nature), but lacked MSA, S100, CD68, CD117 and β -Catenin. There were no histological signs of malignancy (i.e. absence of mitosis, necrosis and cytologic atypia).

The morphological and immunohistochemical data were most consistent with the diagnosis of perineurioma.

Discussion

Perineurioma of soft tissue are usually benign peripheral nerve sheath tumors which are entirely composed of perineurial cells.

These neoplasms are slightly more common in females than males, occurring in a wide age range, with a peak in middle-aged adults.

These tumors are usually sporadic and the majority of these tumors were situated primarily in subcutaneous tissue, in deep soft tissue, and some other cases were limited to the dermis. This kind of lesion commonly arises on the lower limbs, upper limbs and trunk; on the other hand, visceral organs, central body areas, with the head and neck being more rarely affected.¹

The first case was described by Lazarus and Trombetta who suggested the existence of a pure perineurial cell tumor.²

If we consider the morphological aspect of perineurial cells they closely resemble fibroblasts, hence a fundamental diagnostic step is the evidence of immunohistochemical expression of EMA.

We know four different types of perineurioma: soft tissue (extraneural), intraneural, sclerosing and reticular. The first type³ is usually a subcutaneous lesion, the second^{4,5} is a benign neoplasm characterized clinically by the fusiform swelling of a major nerve and usually characterized by a neurologic deficit. The third type is a benign, non-recurring, lesion which is a small solitary skin lesion. The last one represents an unusual morphologic variant within the perineurioma group, and it has been recently described.

Conclusion

Our case is undoubtedly a (soft tissue) extraneural perineurioma. This kind of tumor gives rise to an ovoidal lesion without any connections to a nerve and, in the case examined, there were no anatomical relations with nerve fibers. Moreover, the classical architectural features of intraneural lesions, such as characteristic pseudo-onion bulbs, were not present and were composed of spindled, wavy, cells with remarkably thin cytoplasmic processes arranged in lamellae and embedded in collagen fibers, typical of soft tissue perineuriomas.

Both urologist and pathologists should be made aware of this type of periprostatic lesion, and an immunohistochemical

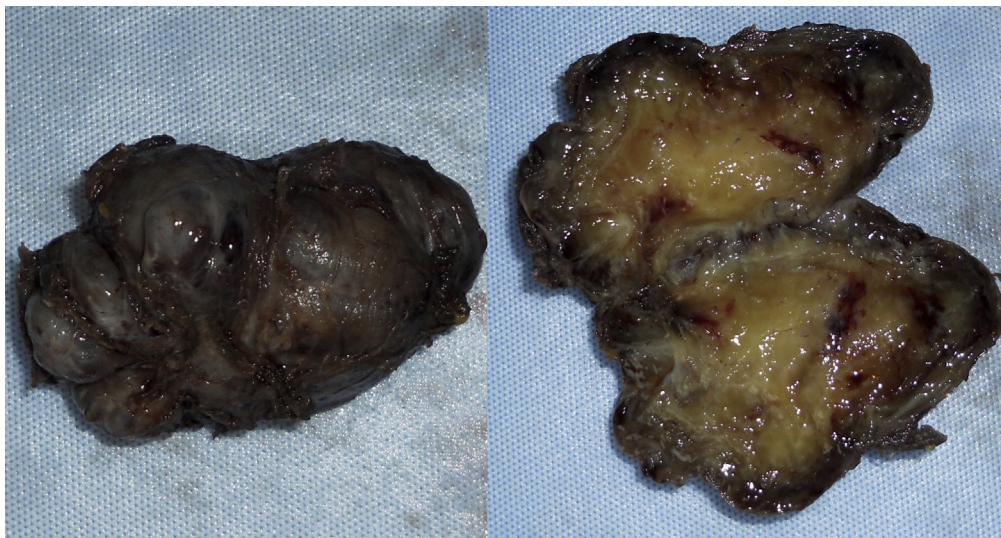


Figure 2. Gross examination of the lesion (of the external surface of the lesion, is evident the polylobate and pseudoencapsulate lesion. The cut surface of the lesion showing the yellowish color and elastic consistency).

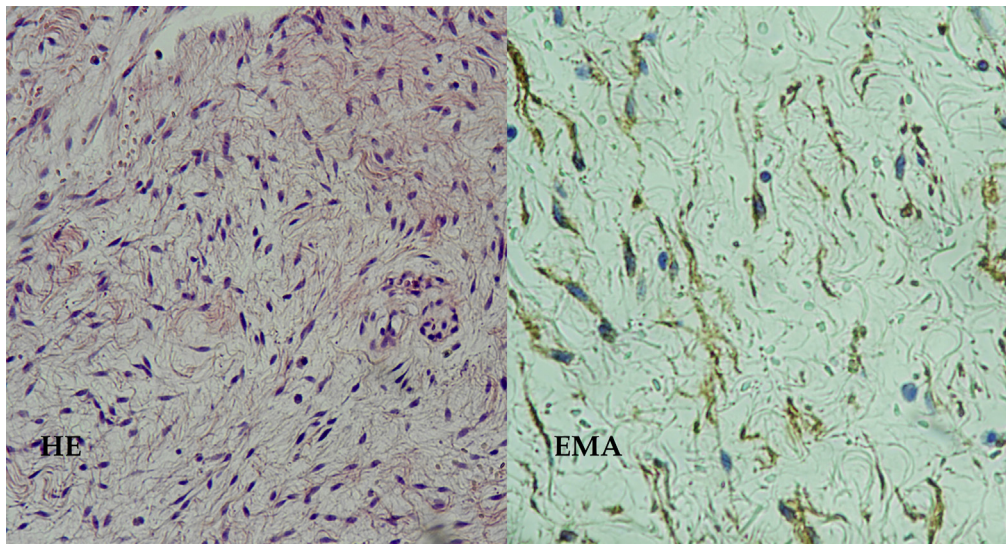


Figure 3. Histological aspect of the lesion.

evaluation is highly recommended, when an unusual mesenchymal tumor is found.

Conflict of interest statement

There is no conflict of interest.

Acknowledgments

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