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Case Report

The Almost Invisible Perineurioma in Urinary Bladder

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Abstract

Perineurioma is a rare entity, it is a benign peripheral nerve sheath tumor entirely composed of perineurial cells. An 18-year-old male patient was admitted to our hospital, suffering from hematuria, burning micturition and dysuria for the last one month. Computed tomography of abdomen and pelvis shows asymmetrical bladder wall thickening. TURBT was performed along with cold cup biopsy. The morphological and immunohistochemically data were most consistent with the diagnosis of perineurioma.

Keywords: *Perineurioma; TURBT; Urinary Bladder*

Introduction

Perineurioma is a tumor composed entirely of neoplastic perineurial cells. They are generally slow-growing benign tumors. They represent approximately 1% of all soft tissue neoplasms [1]. They usually present as a painless nodule in the deep soft tissue. Its diagnostic features include spindle cell proliferation, wavy or tapering nuclei and perivascular whorls. 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 [1].

In the current case report, we describe our experience in a patient with urinary bladder mass which shows perineuroma and benign spindle cell tumor on repeated transurethral resection of bladder tumor (TURBT).

Case Report

A 18 years old man presents in urology outpatient department with complain of off and on episodes of hematuria, burning micturition and dysuria for the last one month. Symptoms did not settle by using broad-spectrum oral antibiotics prescribed by the general practitioner. He had no known comorbid. He denied using any antiplatelet medications and history of trauma. Past history reveals TURBT which shows benign spindle cell neoplasm on histopathology.

The hematological profile shows hemoglobin 11.7 g/dl, white cell count 6.3 * 10^3/uL, platelet 245 * 10^3/uL, serum creatinine 0.7 mg/dl, urinalysis shows numerous pus cells, RBCs 8 - 10/HPF and normal clotting profile. His ultrasound KUB shows thickening of urinary bladder wall. Contrast enhanced computed tomography of the abdomen and pelvis shows asymmetrical bladder wall thickening with maximum caliber on antero-superior wall measuring 1.9 cm. This enhancing area is seen extending into the urachus with resultant thickening of medial umbilical ligament suggestive of its involvement.

The case was discussed in the multidisciplinary team meeting. Cystoscopy along with TURBT was planned which shows multiple nodular lesions in various parts of urinary bladder mainly trigone, posterior wall and dome. Cold cup biopsy taken and sent for histopathology.

Discussion

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

It is an uncommon benign tumor of peripheral nerves composed primarily of peripheral nerves. It is more common in adults especially females. Cytogenetics description reveals a monosomy 22, deletion of 22q11-13.1. Perineuriomas include soft tissue, extra-neural, intraneural and sclerosing variant.

Microscopy reveals bland, elongated cells in parallel bundles resembling neurofibroma or pacinian neurofibroma; may have storiform growth. It may have a collagenous stroma with pericellular cracking with no atypia and rare mitotic figures.

The sections in this case shows fragments of urinary tract mucosa. There is a benign neoplasm in sub epithelial region. It is composed of whorls and fascicles of spindle-shaped cells. Hypocellular areas are also seen at places, where these cells are disposed in parallel bundles. In other foci, they exhibit a storiform pattern. Ganglion cells are also seen. Stroma is collagenous.

Immunohistochemistry shows smooth muscle actin negative, CD34 negative, S-100 protein positive and desmin positive suggestive of perineurioma urinary bladder.

Perineural cells have close resemblance with fibroblast therefore a fundamental diagnostic step is the evidence of immunohistochemically expression of epithelial membrane antigen (EMA).

Conclusion

Mucosal perineuromas affects adults over a wide age range, with a peak during middle age. Mucosal perineuriomas usually occur throughout the colon. Similar lesions very rarely arise in the stomach or small intestine. Occasionally, multiple mucosal perineuriomas may be detected. The aim of this case report is to develop a predictive model in the form of guidelines that shows the risks and complication in patients diagnosed with perineuroma urinary bladder. The model can be used to identify patients with perineuroma by physicians and surgeons which will help in the management.

Bibliography

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