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A rare entity: Ganglioneuroma of the prostate

Ryan J. Cecchi, Jonathan B. McHugh, Eman Abdulfatah, Madelyn Lew *

Department of Pathology, University of Michigan Health System, Ann Arbor, MI, USA

A B S T R A C T

Ganglioneuromas are benign tumors arising from the neural crest. Histologically, they are composed of mature Schwann cells and ganglion cells admixed with fibrous tissue. While they frequently are seen in the abdomen and mediastinum, rare reports have highlighted their occurrences in the genitourinary system. The only prior reported prostatic ganglioneuroma arose in a patient with a history of neurofibromatosis type 1. In this report, we highlight the first reported prostatic ganglioneuroma without a known genetic linkage.

1. Introduction

Mesenchymal lesions of the prostate are rare, comprising <1 % of prostatic tumors. As such, descriptions of their occurrences in literature can be scant, leading to diagnostic difficulty in appropriate classification. However, many of these mesenchymal lesions tend to have fibroblastic, myofibroblastic, or smooth muscle origins. Pelvic-based schwannomas have been reported to compress prostate and seminal vesicles. However, prostatic ganglioneuroma has only been reported in a single case, which arose in a patient with neurofibromatosis type 1.[1] Here we describe the first reported case of a prostatic ganglioneuroma without a genetic linkage.

2. Case presentation

A 58-year-old male with no significant past medical or surgical history presents with concerns of intermittent, weak urinary stream, nocturia, slight post-void dribbling, and "blood in semen." The patient notes that his urinary symptoms have gotten progressively worse with time. He also endorses a "tight feeling and pulling" around his testicles when seated. Subsequent imaging performed for workup of a suspected hernia revealed a 6 cm mass situated at the base of the prostate gland and seminal vesicles extending along the peripheral zone. Imaging also showed an asymmetry of the mass, with a larger right side. Cystoscopy showed no prostatic median lobe and no bladder mucosal trabeculations. PSA during this time period was within a normal range (0.60–0.68 ng/mL), and he denied any personal or familial history of prostate cancer or familial syndrome.

The patient underwent subsequent evaluation by thirteen core biopsies from the right and left base, lateral and medial, mid, and apical prostate as well as bilateral seminal vesicles. Each of the core biopsies showed a proliferation of bland spindled cells with wavy nuclei in a loose fibrillary background admixed with scattered ganglion cells. Immature elements, cytologic atypia, mitotic figures, and necrosis were absent. Benign prostatic elements were only identified in one of the thirteen core biopsies. Immunohistochemistry for S100 and SOX10 highlighted the aforementioned lesional cells. Cytokeratin cocktail and CD117 were negative. The overall morphologic and immunohistochemical features were consistent with a ganglioneuroma.

Approximately 17 months after the initial diagnosis, the patient elected to undergo a radical prostatectomy due to ongoing and worsening symptoms. On subsequent radical prostatectomy, the prostate (7.5 x 7.0 × 5.8 cm) was heavily distorted with an asymmetrically enlarged right lateral lobe pushing both seminal vesicles to the left (Fig. 1A). Upon sectioning the prostate, a soft, yellow lesion largely replaced the appearance of normal prostatic parenchyma (Fig. 1B).

On histology, the prostate was stroma-rich with several cells displaying wavy nuclei admixed with large cells displaying abundant amounts of eosinophilic cytoplasm and prominent nucleoli (Fig. 2), morphologically consistent with Schwann cells and ganglion cells. No pleomorphism, necrosis, or small cell changes were noted. The periphery of the specimen had large nerve bundles with perineurium, corresponding to hyperplastic changes in the nerve fibers (Fig. 3), extending to the edges of the resection specimen. Immunohistochemistry for S100 and SOX10 highlighted nerve fibers and ganglion cells while cytokeratin cocktail was negative, supporting the morphologic features of ganglioneuroma.

* Corresponding author. Department of Pathology, University of Michigan Health System, 2800 Plymouth Road, Ann Arbor, MI, 48109-5054, USA.
E-mail address: lewm@med.umich.edu (M. Lew).

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3. Discussion

Ganglioneuromas are benign tumors of neuroectodermal tissues of the autonomic nervous system and are composed of well-differentiated ganglion and Schwann cells admixed with variable amounts of fibrous stroma and nerve fibers. While they can occur anywhere in the body along the sympathetic chain, they are most frequently identified in the posterior mediastinum, retroperitoneum, and adrenal glands with a predilection for young adults and the female sex. They have also been rarely reported in the genitourinary tract.[1–5] Per these case reports, genitourinary ganglioneuromas are frequently associated with neurofibromatosis type 1.[1,2,5] However, as our patient had no history of neurofibromatosis type 1 and no subsequent lesions identified on imaging, we are reporting, to our knowledge, the first instance of ganglioneuroma involving the prostate without a currently identified genetic linkage.

The rare reports of urinary bladder ganglioneuromas showcase a radiologic appearance of infiltrative masses invading bladder walls. Similarly, our patient presented with a large, infiltrative mass that replaced the large majority of prostatic parenchyma. The neoplastic cells coursed through prostatic stroma and were closely intertwined with adjacent benign elements. Given the extensive infiltration into normal tissues with progressive symptomatology (including urinary obstructive symptoms and pelvic pain) due to mass effect, genitourinary ganglioneuromas can require extensive surgery despite their benign nature. For our patient, the mass effect led to compression of the bladder which significantly impacted his quality of life, warranting more extensive surgical resection. While smaller ganglioneuromas may be encountered and diagnosed on core biopsy (as in our patient) and typically are associated with good prognoses, there have been rare reports of spontaneous malignant transformation and malignant peripheral nerve sheath tumors arising from ganglioneuromas, indicating their uncertain malignant potential. As such, even in asymptomatic patients, close monitoring and potential surgery may be indicated.

In our microscopic evaluation, it was notable that there were several hyperplastic nerve fibers present throughout and at the periphery of the specimen, which are not representative of the actual neoplasm. Initial sampling of these elements by core biopsy may therefore introduce other neural lesions into the list of diagnostic considerations, especially when pre-operative MRI may not present with the traditional "whorled" appearance of ganglioneuromas.

4. Conclusion

Ganglioneuromas are rare benign tumors of neuroectodermal origin that can present as large, invasive masses of the prostate. While our case was histologically characterized by infiltration of ganglion and Schwann cells infiltrating prostatic stroma and extraprostatic fat, hyperplastic nerves were a prominent feature throughout the prostatectomy

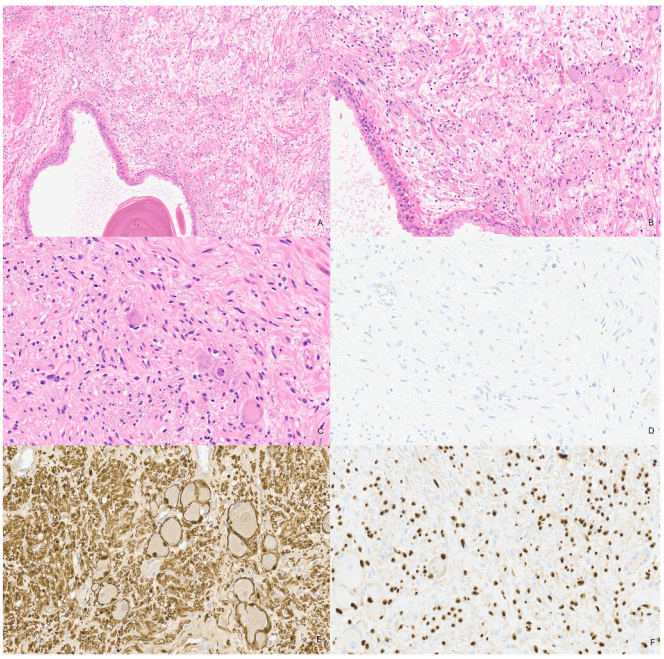

Fig. 2. Representative pictures of the ganglioneuroma and supporting immunohistochemical stains: H&E (A:20x, B:40x, C:40x), cytokeratin cocktail (D:40x), S100 (E:40x), SOX10 (F:40x).

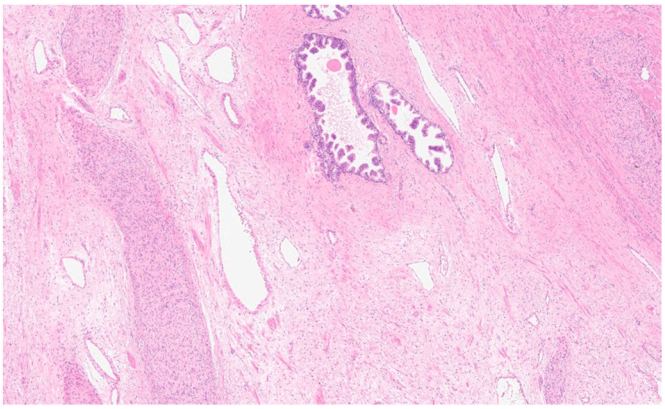

Fig. 3. Hyperplastic nerve with intact perineurium in close proximity to residual normal prostatic parenchyma at the apex of the prostate.

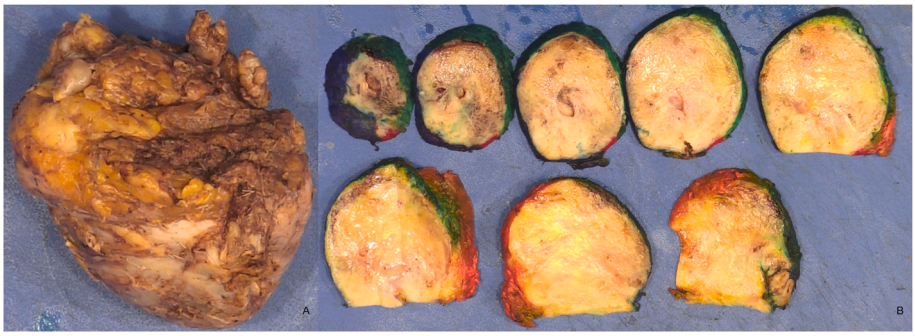

Fig. 1. (A) Anterior view with slight rightward skew of the prostate. Notice the bilateral seminal vesicles located on the left side of the prostate. (B) Cut surfaces of the prostate showing replacement by soft, tan, yellow tissue with minimal normal prostatic parenchyma located towards the apex. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

specimen.

Author contributions

Ryan Cecchi: conceptualization, investigation, writing original draft; Jonathan McHugh: writing – review & editing; Eman Abdulfatah: writing – review & editing; Madelyn Lew: conceptualization, investigation, writing the original draft; writing – review & editing.

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Declaration of competing interest

The Authors declare that there are no competing interests.

References

1. Nassiri M, Ghazi C, Stivers JR, Nadji M. Ganglioneuroma of the prostate. A novel finding in neurofibromatosis. *Arch Pathol Lab Med*. 1994;118(9):938–939.
2. Ţarcă E, Cojocaru E, Trandafir LM, et al. Ganglioneuroma of the bladder in association with neurofibromatosis type 1. *Diagnostics*. 2022 Dec 12;12(12):3126.
3. Ling K, Xiaoping C, Xiaolu Y. A case of ganglioneuroma of the ureter and review of the literature. *J Int Med Res*. 2022 Nov;50(11), 3000605221123671.
4. Fang CW, Wang JS, Wu TT, Lin JT. Occurrence of paratesticular ganglioneuroma 18 years after concurrent adrenal ganglioneuroma and papillary thyroid carcinoma - a case report. *BMC Cancer*. 2019 Dec 30;19(1):1265.
5. Ku S, Balasubramanian A, Kao CS, Eisenberg ML, Skinner EC. Co-manifestations of genital neurofibromatosis in a patient with neurofibromatosis type 1. *Urology*. 2020; 141:e49–e50. https://doi.org/10.1016/j.urology.2020.03.030.