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Ganglioneuroma of the bladder: Case report and literature review

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Abstract Ganglioneuromas originate from neural crest sympathogonia of the autonomic nervous system. These tumors are benign, fully differentiated tumors that contain mature Schwann cells, ganglion cells, fibrous tissue and nerve fibers. Ganglioneuromas commonly localize in the abdominal and mediastinum regions, such as the adrenal gland, paraspinal retroperitoneum and posterior mediastinum. However, these tumors are rarely localized to the urinary bladder and, herein, we present a brief review of the literature and a ganglioneuroma of a 16-year-old female patient as the second case reported in the urinary bladder.

Key Words Ganglioneuroma; pediatric bladder tumor; benign bladder tumor.

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INTRODUCTION

Ganglioneuromas are benign tumors arising from neuroectodermal tissues composed of ganglion cells and Schwann cells. These tumors most commonly present in the younger population between ages 10-29 years old, but their location is largely limited to the adrenal glands (21%), mediastinum (40%), and retroperitoneum (30%) [1-3]. We report only the second case in the literature of a ganglioneuroma of the bladder in the

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pediatric population. Herein, we present a 16 year-old female with a history of recurrent suprapubic pain evaluated my multiple practitioners prior to presentation in our Emergency Department and found to have a ganglioneuroma arising from the bladder.

CASE REPORT

The patient is a 16-year-old female who presented initially to Gastroenterology with per-umbilical abdominal pain. After a work-up, the etiology was thought to be functional abdominal pain and behavioral recommendations were provided. No imaging was performed at that time. Her pain gradually worsened with localization to her suprapubic abdomen. She was evaluated by a gynecologist for the pain and a pelvic ultrasound was obtained which identified a tumor in the wall of the bladder suspicious for leiomyoma. She subsequently presented to the Emergency Department for continued pain. She had no history of fever, diarrhea, vomiting, urinary tract infections, dysuria, hematuria, or hematochezia. Her bowel movements were regular and soft. She was not sexually active and denied vaginal discharge.

A complete urinary tract work-up was performed including an ultrasound of her kidneys and bladder which further characterized the suspicious lesion as a 3.6cm bladder dome mass (Fig. 1).

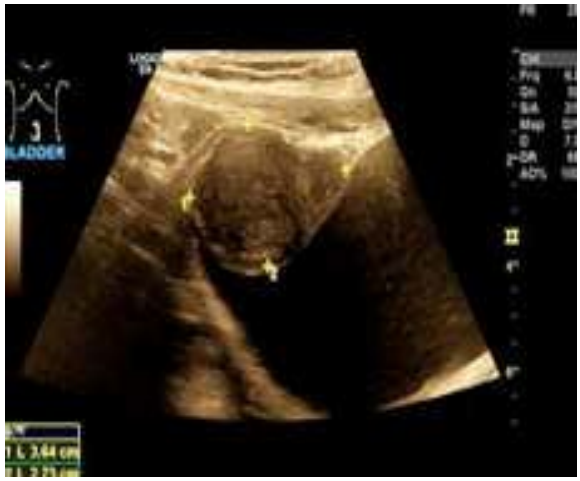
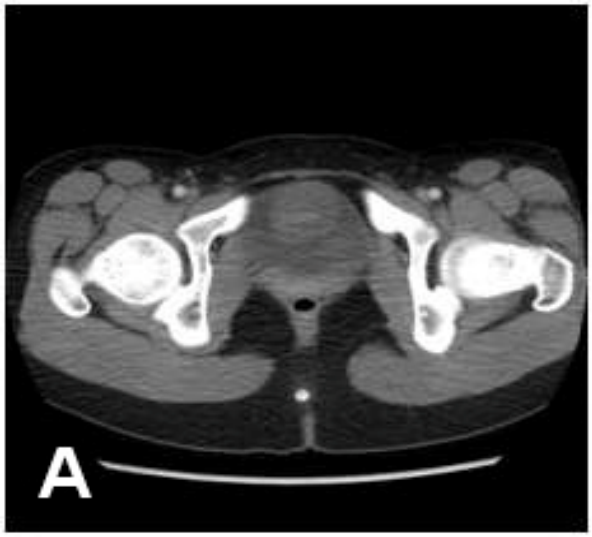


Fig. 1. Ultrasound images of the bladder.

A computed tomography (CT) scan was obtained to further delineate the mass and evaluate for an invasive process. This again demonstrated a 3.8 cm bladder dome mass extending into the lumen with preservation of surrounding tissue planes and no evidence of further invasive process (Fig. 2A and B).



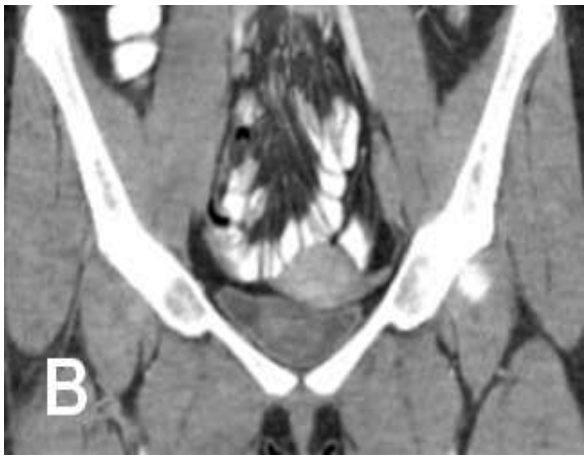


Fig. 2A, B. CT images of the bladder mass

The remainder of the CT scan did not show any signs of lymphadenopathy or metastasis. She was taken to the operating room the following day and underwent a partial cystectomy.

Histological analysis (Fig. 3) revealed mature Schwannian stroma and neuritic processes arranged in fascicular and cordlike pattern. These are accompanied by perineural cells with the presence of ganglion cells.

The Schwannian stroma was extensively positive for immunohistochemical marker S-100 and synaptophysin (Fig. 4). The perineural cells and capillary endothelial cells showed diffuse positivity for cluster of differentiation - 34. There was a bundle of smooth muscle cells, separate from the lesion, positive for smooth muscle antibody (SMA).

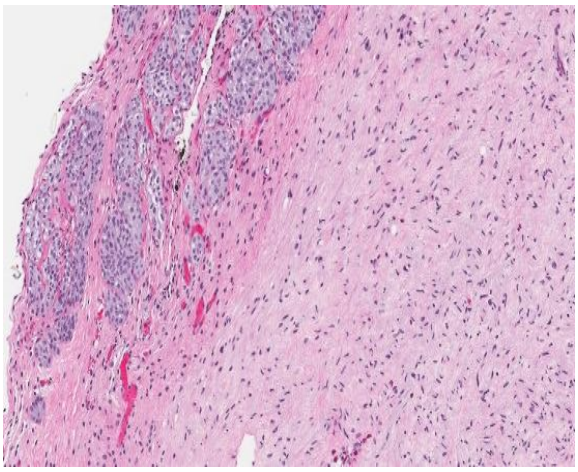


Fig. 3. Hematoxylin-eosin staining demostrating (original magnifications X 100).

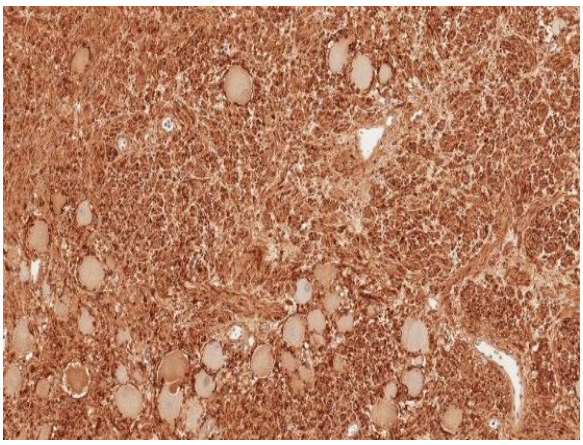


Fig. 4. Immunohistochemistry showing positive staining for S-100 (original magnifications X 100).

DISCUSSION

This report identifies a patient in the pediatric population with very rare pathology and location for this type of tumor. Ganglioneuromas comprise only

<0.5% of all bladder tumors and are usually identified in the adult population once they becomes large enough to cause symptoms of obstruction, urgency, frequency, etc [4]. Looking at previous literature on the topic, we identified a single case of ganglioneuroma of the bladder in a pediatric patient which was treated successfully with resection via transurethral approach. This patient presented with hematuria and recurrent urinary tract infections which led to imaging studies identifying the mass. The prior report does not mention if complete resection was achieved, but intraoperative frozen sections favored a benign tumor, likely ganglioneuroma. This was later confirmed with permanent staining [5]. Our approach does represent another management style, but as these tumors are benign in origin and recurrence is rare, either method is plausible and effective.

Due to our patient being older, we felt it was more appropriate to proceed with an open operation as the risk of transitional cell carcinoma increases with age [6]. We did not think adequate resection would be possible endoscopically due to location and size. The open technique offered the ability to completely resect the tumor without violating it as well as ability to evaluate her urachus more closely for any evidence of invasion or extension. We were also unsure of the pathology and did not want to risk spread if this was a malignant process. With the standard of care for an urachal adenocarcinoma being partial cystectomy, we felt this would be an appropriate way to approach this mass as well. During the procedure, we did not identify any association between the mass and her urachus, so we did not resect her umbilicus and only performed a partial cystectomy.

Ganglioneuromas represent the most mature form of a neuroblastoma [7]. In the older pediatric population, ganglioneuromas are most often identified in the posterior mediastinum and retroperitoneum [3]. The tumors are usually identified due to large growth and compression symptoms, either cause respiratory compromise or spinal cord deficits. A retrospective review of 227 cases of ganglioneuroma which were treated with surgical resection concluded that unfavorable events (Horner syndrome, scoliosis, bowel obstruction) were occasionally reported, but since the diagnosis of ganglioneuroma cannot be confirmed without removal of the mass, this should still be the standard of care [8,9].

Treatment of these lesions is achieved by complete surgical resection. Han PP describes a case series of 7 patients with large intrathoracic neurogenic tumors which were all entirely resected via endoscopic

thoracic approaches [10]. Another report from Li states successful complete resection of 15 ganlgioneuromas of adrenal glands in adults without significant morbidity or mortality [11]. These tumors have excellent prognosis with rare chance of recurrence following resection [12].

Ganglioneuromas originate from neuroectodermal cells and contain Schwann cells, ganglions, and nerves. These tumors account for <0.5% of all bladder tumors in all populations. There are reports of the tumors presenting with urinary obstructive symptoms or urinary urgency if located at bladder neck, but for the most part they are asymptomatic. The most common single location for ganglioneuromas is the adrenal gland which represents 21% [1-3]. Imaging of a ganglioneuroma on CT shows round, lobulated mass of low density, and uniform appearance. These tumors do not enhance with contrast administration.

We report a very rare pathology of ganglioneuroma in the pediatric population. Treatment in both the adult and pediatric population is complete resection of the mass. Further analysis of emerging data will be necessary to standardize a treatment plan.

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