

CASE REPORT

Functional urinary bladder paraganglioma in a child presenting with obstructive uropathy: A rare case report

Shameer Tahir | Eisha Sattar | Javeria Khan

Allama Iqbal Medical College, Lahore, Punjab, Pakistan

Correspondence

Shameer Tahir
Email: Shameertahir04@gmail.com

Abstract

Background: Pediatric urinary bladder paraganglioma is an exceptionally rare neuroendocrine tumor arising from chromaffin cells within the detrusor muscle. Due to its rarity and variable presentation, early diagnosis can be challenging, often requiring a high index of clinical suspicion and multi-modal evaluation.

Case Presentation: We report a 12-year-old female who presented with hypogastric pain, urinary frequency, and mild dysuria. Initial imaging revealed a bladder mass associated with left-sided hydroureteronephrosis. Over time, the patient developed episodic hypertension, palpitations, and flushing, suggestive of catecholamine excess. Further evaluation with magnetic resonance imaging (MRI) and positron emission tomography/computed tomography (PET/CT) demonstrated a locally invasive mass involving the bladder wall, vesicoureteric junction, cervix, and upper vagina, without evidence of distant metastasis. Histopathological examination showed nests of polygonal cells arranged in a characteristic Zellballen pattern. Immunohistochemistry was positive for chromogranin, synaptophysin, and S-100 protein, confirming the diagnosis of paraganglioma.

Conclusion: This case underscores the diagnostic challenges associated with pediatric bladder paraganglioma and highlights the importance of early recognition, thorough evaluation, and a multidisciplinary approach to ensure optimal management and outcomes.

KEYWORDS

bladder paraganglioma, catecholamine excess, chromogranin, extra-adrenal pheochromocytoma, hydroureteronephrosis, immunohistochemistry, local invasion, multidisciplinary management, pediatric neuroendocrine tumor, Zellballen pattern

1 | INTRODUCTION

Paraganglioma of the urinary bladder, also termed extra-adrenal pheochromocytoma, is a rare neuroendocrine tumor arising from chromaffin cells in the detrusor muscle^[1,2]. It accounts for less than 1% of all paragangliomas and approximately 0.05% of bladder tumors, with pediatric cases being exceedingly uncommon,

representing nearly 1% of pediatric bladder tumors^[3–5]. The urinary bladder is the least frequent site of occurrence^[6], and cases demonstrating malignant potential through metastasis are exceptionally rare^[7,8]. Functional tumors may present with catecholamine-mediated symptoms, including episodic hypertension, headache, palpitations, and diaphoresis, or with non-specific urinary complaints^[9,10]. We report a 12-year-old female

Shameer Tahir, Eisha Sattar, and Javeria Khan contributed equally to this work.

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with a locally invasive, functional bladder paraganglioma causing obstructive uropathy, illustrating diagnostic complexity, management challenges, and the importance of a multidisciplinary approach.

2 | CASE PRESENTATION

A 12-year-old female presented in early 2021 with hypogastric pain, urinary frequency, and mild dysuria. She denied hematuria, fever, weight loss, or other systemic symptoms. Medical, surgical, and family history were unremarkable. Initial laboratory investigations, including renal function tests and urinalysis, were normal.

Plasma metanephrines tested in January 2021 showed a significantly elevated normetanephrine level of 900 pg/mL (reference: < 190 pg/mL), while metanephrine was 64.4 pg/mL (reference: < 90 pg/mL), confirming the tumor's functional nature.

A dynamic renal scan in February 2021 demonstrated left-sided obstructive uropathy with reduced cortical function^[3–5]. Contrast-enhanced positron emission tomography/computed tomography (PET/CT) in December 2021 revealed a 3.6 cm × 2.9 cm × 3.5 cm heterogeneously enhancing mass along the left posterolateral bladder wall with left hydroureteronephrosis^[6]. Follow-up imaging in October 2022 showed interval growth to 4.9 cm × 4.0 cm × 4.6 cm, without distant metastasis^[7,8]. By December 2022, she developed episodic hypertension, palpitations, and flushing, suggestive of catecholamine excess^[10].

Magnetic resonance imaging (MRI) in February 2025 demonstrated a 5.8 cm × 4.6 cm × 4.7 cm lobulated, enhancing mass arising from the left bladder wall, infiltrating the bladder base, vesicoureteric junction, cervix, lower uterine segment, and upper anterior vaginal wall^[6–8]. An intravesical component of 27 mm and persistent left hydronephrosis were noted.

Histopathology (Figure 1) revealed Zellballen pattern nests of polygonal cells with eosinophilic cytoplasm and round-to-oval nuclei^[1,2]. Immunohistochemistry was positive for chromogranin, synaptophysin, and S-100 protein, and negative for cytokeratin, confirming bladder paraganglioma^[10].

The patient was evaluated by a multidisciplinary team including uro-oncology, endocrinology, radiology, and pediatric surgery. Conservative measures maintained renal function and symptom control. Given the extensive local invasion, radical cystectomy was planned, with long-term surveillance recommended.

3 | DISCUSSION

Bladder paragangliomas are exceptionally rare in the pediatric population, accounting for less than 1% of all bladder tumors^[3–5]. These tumors may be functional, producing catecholamines that lead to characteristic

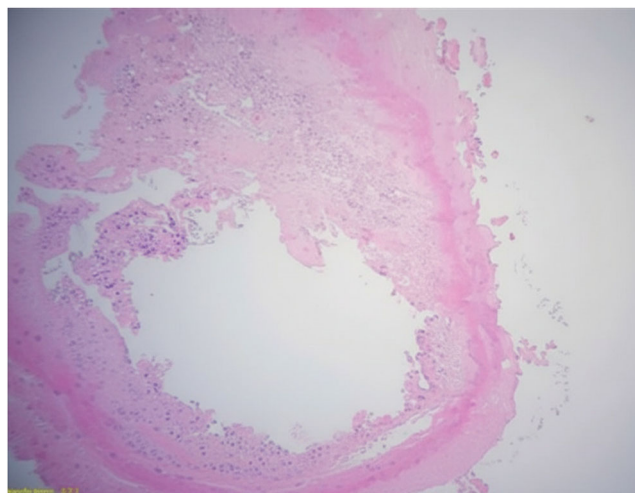


FIGURE 1 Histopathology of urinary bladder paraganglioma shows nests of polygonal cells (Zellballen pattern) separated by vascular stroma. Immunohistochemistry demonstrates positivity for chromogranin and synaptophysin, confirming neuroendocrine differentiation.

symptoms such as episodic hypertension, palpitations, and flushing. However, when these features are subtle or unrecognized, diagnosis may be delayed, increasing the risk of disease progression and complicating management^[9,10].

In the present case, the patient initially exhibited non-specific urinary symptoms, with features of catecholamine excess developing later in the clinical course^[1,2]. This atypical sequence contributed to diagnostic delay. Notably, progressive local invasion led to involvement of the vesicoureteric junction, resulting in obstructive uropathy—an uncommon manifestation that typically reflects advanced or strategically located disease^[6–8].

Definitive diagnosis was established through histopathological examination demonstrating the classic Zellballen architecture, supported by immunohistochemical positivity for neuroendocrine markers. This highlights the critical role of tissue diagnosis in confirming rare neuroendocrine tumors and differentiating them from other bladder neoplasms^[10].

This case emphasizes the importance of maintaining a high index of suspicion in pediatric patients presenting with unexplained urinary symptoms, particularly when accompanied by intermittent sympathetic features^[3–5]. Early recognition is essential, as delayed diagnosis may allow tumor growth, local invasion, and increased surgical complexity.

A multidisciplinary approach is crucial for optimal management. Preoperative recognition of functional status is especially important to prevent perioperative complications related to catecholamine release.

Reporting such rare cases contributes to the growing body of literature on pediatric bladder paragangliomas,

enhancing understanding of their clinical behavior, diagnostic challenges, and management strategies^[7,8]. Clinicians should remain vigilant for catecholamine-mediated symptoms in atypical urinary presentations to facilitate timely diagnosis and improve outcomes^[1,2,10].

4 | CONCLUSION

Pediatric urinary bladder paraganglioma is an exceptionally rare and potentially life-threatening tumor, particularly when functional and locally invasive. Early recognition, prompt evaluation, and multidisciplinary management are crucial to optimize outcomes. Long-term surveillance is essential due to risks of recurrence and progression. Awareness of sympathetic symptoms and obstructive uropathy in children can facilitate timely diagnosis, allowing appropriate surgical and medical intervention while minimizing morbidity.

AUTHOR CONTRIBUTIONS

All authors were involved in the conception and design of the study, data collection, literature review, drafting of the manuscript, and critical revision of the article for important intellectual content. All authors approved the final version of the manuscript and agreed to be accountable for all aspects of the work.

CONFLICT OF INTEREST STATEMENT

The authors declare no conflicts of interest.

DATA AVAILABILITY STATEMENT

The data analyzed during this current study are available from the corresponding author on reasonable request.

ETHICS STATEMENT

Written informed consent was obtained from the patient's guardians for publication of this case report and any accompanying images. The patient was informed about the purpose of publication, and all efforts have been made to ensure confidentiality and anonymity.

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