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(CASE REPORT)

A challenging diagnostic and therapeutic case report: Ewing sarcoma / primitive neuroectodermal tumor of the bladder

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Abstract

Background: Bladder ES/PNET is a rare entity with a few cases described in the literature. This article will focus on an atypical vesical presentation of this tumor.

Summary: We report a case of Ewing's sarcoma arising in the bladder in a 37-year-old patient with type 2 diabetes who presented with clotting hematuria and irritative symptoms of the lower urinary tract. On clinical examination, a hard and mobile hypogastric mass was palpated. CT scan revealed an abdominopelvic mass with predominantly extravesical development. Cystoscopy showed apparent displacement of the vesical dome without clear lesions within it. Given this perplexing presentation, we performed mass excision with partial cystectomy. Anatomopathological, immunohistochemical, and molecular biology studies supported the diagnosis of Ewing's sarcoma of vesical origin. Unfortunately, the patient passed away one month later due to complications related to a digestive obstruction.

Conclusion; The objective of our presentation was to describe an additional case that presented both diagnostic and therapeutic challenges, in order to stimulate further consideration and derive clear recommendations.

Keywords: Case report; Bladder; ES/PNET; Exophytic

1. Introduction

ES/PNET is a rare malignant primary tumor characterized by small round cells [1]. In the literature, only 22 cases of bladder ES/PNET have been documented [3-24]. Its preoperative diagnosis is still complicated due to the absence of specific imaging features. Definitive diagnosis relies on postoperative pathology, immunohistochemistry, and genetic analysis. Due to the minuscule number of reported cases, establishing clear guidelines for its management and treatment is challenging. Therefore, early diagnosis and treatment of ES/PNET patients are highly challenging. This article describes an unusual case of PNET in the bladder in an adult.

2. Case Presentation

We present the case of a 37-year-old patient with a history of type II diabetes treated with oral antidiabetic agents. The patient presented with intermittent clotting hematuria for three months, accompanied by irritative symptoms of the lower urinary tract, including pollakiuria and urgency. On clinical examination, a hard abdominal mass was palpable in the hypogastric region. Abdominal CT scan revealed a necrotic abdominopelvic mass invading the vesical dome with

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irregular contours, showing heterogeneous enhancement after contrast administration. The mass measured 15 cm x 10 cm in transverse diameter and 14 cm in height, exerting a mass effect on the right iliac ureter and causing slight infiltration of the perivesical fat without visible iliac lymphadenopathy. Cystoscopy showed displacement of the vesical dome by the described mass, with a healthy mucosa.

The patient underwent mass excision with partial cystectomy. Microscopic examination revealed a poorly differentiated malignant tumor proliferation composed of small round blue cells. Frequent mitotic figures and the presence of Homer-Wright rosettes in some regions were observed (Fig. 2a). Immunohistochemical analysis supported the diagnosis of Ewing's sarcoma, with focal positivity for anti-NSE, anti-synaptophysin, and anti-CD99 antibodies (Fig. 2b). However, anti-CD20, anti-CD3, anti-PS100, anti-desmin, anti-myogenin, anti-pancytokeratin, anti-ERG, and R MaB antibodies showed negative staining. The patient was referred to the oncology center for additional chemotherapy. Unfortunately, one month later, the patient passed away due to complications related to a digestive obstruction.

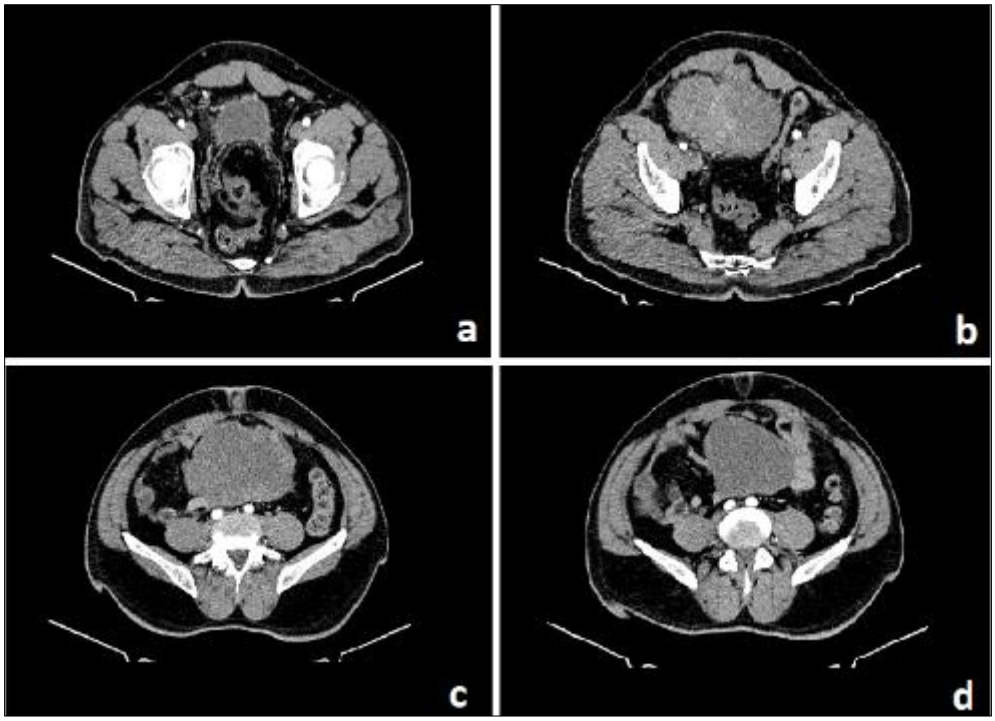

Figure 1 Axial section of an abdominopelvic CT scan showing an exophytic development of PNET

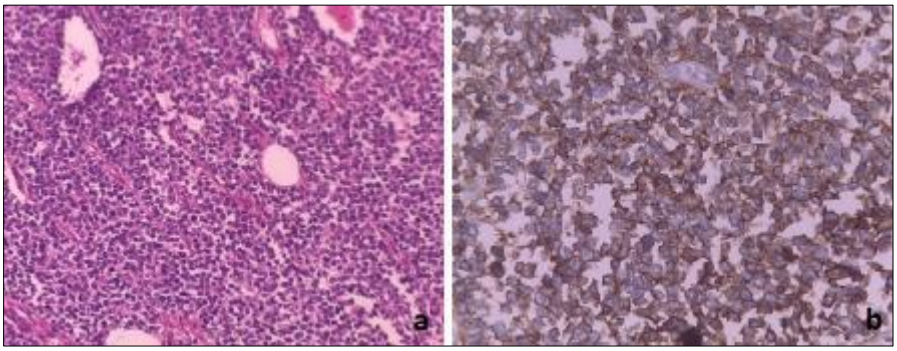

Figure 2 (a) Sheets of monomorphic small round cells with hyperchromatic nuclei and clear cytoplasm. (H&E, 400). (b) Histopathology section exhibiting strong membranous CD99 immunostaining (H&E, 350)

3. Discussion

A primitive neuroectodermal tumor (PNET) is a rare and malignant tumor belonging to the Ewing's family of tumors [1]. It was first described in 1918 [2]. These tumors pose both diagnostic and therapeutic challenges for clinicians and pathologists.

The primary issue encountered with a bladder ES/PNET is the diagnostic problem due to its occurrence at an unusual age, the rarity of its localization in the bladder, and notably its atypical presentation as a predominantly exophytic tumor.

Only 22 cases of bladder ES/PNET have been described in the literature (table 1). The youngest patient was 10 years old [11], the oldest was 81 years old [8], resulting in a mean age of 41.9 years, which is younger than that reported in urothelial carcinomas [25].

Table 1 Published cases of primitive neuroectodermal tumor (PNET) of the bladder to date.

Author	Year of publication	Sex/age	Tumor size (mm)	Growth patern	Treatment
Banerjee et al. [3]	1997	M/21	80×60×40	Endophytic	Cystectomy+chemotherapy
Gousse et al. [4]	1997	F/15	30×20×20	Endophytic	TURBT+ Partial cystectomy
Mentzel et al. [5]	1998	M /21	140×100×100	Endophytic and exophytic	TURBT + nephrostomy
Desai. [6]	1998	F/38	120×70×35	Endophytic and exophytic	Cystectomy
Colecchia et al. [7]	2002	F/61	-	Endophytic and exophytic	-
Krüger et al. [8]	2003	M/81	-	Endophytic and exophytic	TURBT + nephrostomy
Lopez-Beltran et al. [9]	2006	F/21	90×80×60	Endophytic	cystectomy+chemotherapy
Ellinger et al. [10]	2006	M/72	-	Endophytic and exophytic	TURBT+ ileal conduit
Osone et al. [11]	2007	M/10	10	Endophytic	TURBT
Al Meshaan et al. [12]	2009	F/67	30×25×10	Endophytic	TURBT+ Partial cystectomy
Busato et al. [13]	2011	F/52	33×15×22	Endophytic	TURBT+ chemotherapy
Okada et al. [14]	2011	M/65	50	Endophytic	TURBT+ cystectomy+ chemotherapy
Zheng et al. [15]	2011	M /74	-	Endophytic and exophytic	TURBT+ palliative surgery+ chemotherapy
Rao et al. [16]	2011	F/14	150×120×75	Endophytic and exophytic	Partial cystectomy
Sueyoshi et al. [17]	2014	M/10	135×131×129	Endophytic and exophytic	Double J tube + partial cystectomy+ chemotherapy
Lam et al. [18]	2016	F/30	64×94×77	Endophytic	TURBT+ chemotherapy+ cystectomy
Vallonthaiel et al. [19]	2016	F/27	103×98×47	Endophytic	TURBT+ chemotherapy
Tonyalı et al. [20]	2016	F/38	40×26×25	Endophytic	cystectomy + chemotherapy
Zhang et al. [21]	2020	F/78	63×44	Endophytic	TURBT
Gao et al. [22]	2020	F/45	30	Endophytic	TURBT + cystectomy + chemotherapy

Liu et al. [23]	2020	M/64	60×50	Exophytic	Partial cystectomy+ radiotherapy
Jingyi et al. [24]	2023	F/19	55×36	Endophytic	TURBT+ chemotherapy
Present case	2025	M/37	150 × 100 × 140	Exophytic	Partial cystectomy

These tumors have a preference for the central nervous system in children and young adults, although they can occasionally develop in other locations such as the thoracic wall, head and neck, kidneys, adrenal glands, female genital tract, extremities, and scrotal sac [23]. The presence of a PNET in the bladder is extremely rare.

The majority of the described cases (21/22) exhibit endophytic growth, while 8 of them are accompanied by exophytic growth. However, a tumor of purely exophytic development has only been reported in one case [23], and our case is the second. Cystoscopy confirmed this presentation by revealing a completely healthy vesical mucosa, which raises suspicions of an alternative diagnosis, such as a tumor originating from the digestive tract.

Given the rarity of these tumors, it is difficult to consider them initially. A definitive diagnosis relies on anatomopathological study, immunohistochemistry, and molecular biology [1].

Apart from the diagnostic challenge, the second issue that arises is the treatment. Due to the limited number of patients with PNET, no recommendations can be established, and most knowledge about treatment has been derived from studies on the Ewing's sarcoma family and the reported cases in the literature.

Seven patients underwent total cystectomy [3,6,9,14,18,20,22], five underwent partial cystectomy [4,12,16,17,23], and eight underwent only transurethral resection of the bladder [5,8,10,11,13,19,21,24]. Chemotherapy was associated in most cases, with a favorable response observed in some instances [19,24].

In our case, we performed a partial cystectomy with excision of the mass, and postoperative chemotherapy was planned. However, the patient's general condition rapidly deteriorated, preventing them from receiving the chemotherapy.

Successful treatment necessitates a multimodal approach [3,9,20,22]. Most patients with apparently localized disease have occult metastases, thereby warranting additional therapy, such as chemotherapy [10]. Poor prognosis has been reported for those with existing metastases [12,13].

Abbreviations

- ES/PNET: Ewing Sarcoma / Primitive Neuroectodermal Tumor of the Bladder
- CT: computed tomography.

4. Conclusion

In conclusion, clinicians and pathologists should be aware of this rare entity during diagnosis and management. Surgery supported by chemotherapy should be considered as an option, especially in advanced disease cases. With the description of new cases and long-term follow-up results, our knowledge and experience will continue to improve.

Compliance with ethical standards

Disclosure of conflict of interest

The authors report no competing personal or financial interest related to this work.

Statement of informed consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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