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CASE REPORT

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Rapidly growing primitive neuroectodermal tumor of the uterus causing obstructive uropathy in a 13-year-old African girl: A case report

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Abstract

Background: Uterine primitive neuroectodermal tumors (PNETs) are a rare group of embryonal tumors, with less than 100 cases reported, mostly in postmenopausal women. Their rarity poses diagnostic and management challenges, contributing to high mortality.

Case presentation: A 13-year-old presented with a 2-month history of abdominal pain, swelling, and heavy menstruation. Physical examination revealed a firm, fixed, non-tender pelvic mass. She underwent a panhysterectomy and nodal resection. Histopathology confirmed PNET. The tumor recurred within a month, and intraoperative spillage occurred during repeat surgery. She developed obstructive uropathy and succumbed approximately 11 months after initial presentation.

Conclusion: Uterine PNETs are aggressive tumors that rapidly infiltrate and obstruct pelvic structures. Owing to their rarity, diagnosis and management remain challenging, with high mortality.

Keywords: embryonal tumors, obstructive uropathy, primitive neuroectodermal tumor, uterus

Introduction

Primitive neuroectodermal tumors (PNETs) are highly malignant embryonal tumors composed of small, round cells of neuroectodermal origin, first described in 1973 (1). They rarely arise in the female genital tract, with fewer than 100 cases of uterine PNETs reported globally, predominantly in postmenopausal women (2). Patients often present at an advanced disease stage with abnormal uterine bleeding, an aggressive clinical course, and a poor prognosis (3). Because of the rarity in the female reproductive tract, there is a paucity of data (4). This is a

case of an aggressive uterine PNET in a 13-year-old African girl.

Case presentation

A 13-year-old African girl presented to the gynecological oncology ward at Kenyatta National Hospital as a referral. She had a 2-month history of right inguinal region swelling, lower abdominal pain, and heavy menstrual bleeding necessitating multiple blood transfusions. At the referring facility, she had misdiagnoses of hormonal imbalance initially, then endometrial lymphoma following the finding of a magnetic resonance imaging (MRI). On examination, the patient was mildly pale, with inguinal lymphadenopathy, mild ascites, a firm, fixed, nontender right iliac fossa mass (6 x 8 cm), and a bulky uterus corresponding to the size of 14 weeks. No hepatosplenomegaly was noted.

Laboratory results showed anemia (hemoglobin 7.6 g/dL; reference range 12-15.5 g/dL). Liver function tests revealed elevated γ-glutamyl transferase (112 U/L; reference 11-52 U/L). The renal function test was normal. She was HIV seronegative. Ultrasound-guided biopsy of the pelvic mass revealed a tumor of small, round to oval, blue cells with scant cytoplasm, pseudorosette formation, and high mitotic activity. Immunohistochemistry demonstrated undifferentiated neoplastic cells with strong p16 and CD99 (Golgi positivity) and patchy synaptophysin expression (**Figure 1**).

She received 3 cycles of neoadjuvant chemotherapy (vincristine, doxorubicin, cyclophosphamide/ifosfamide, and etoposide (VDC/IE)), followed by panhysterectomy with inguinal node dissection. Intraoperatively, a uterine tumor extending to the cervix, upper vagina, and pouch of Douglas with necrosis invading the sigmoid colon was observed. Postoperatively, she received adjuvant chemotherapy (VDC/IE). However, a positron emission tomography scan revealed recurrent disease at the surgical bed and inguinal nodes. MRI confirmed a cystic pelvic mass (81 x 75 x 92 mm) and a solitary inguinal node (25 mm, short axis).

Repeat surgery at five months revealed a cystic mass adherent to the posterior wall of the bladder and anterior vaginal wall. Tumor spillage occurred during debulking, while the inguinal node was resected successfully. Cytology confirmed recurrent metastatic PNET. Five weeks later, she developed obstructive uropathy with acute kidney injury (creatinine 692.9 mmol/L (reference 45-86 mmol/L); urea >40 mmol/L (reference 1.8-5.2 mmol/L); and estimated glomerular filtration rate 7 ml/min/1.73m² (reference ≥90 ml/min/1.73m²). She also developed bowel obstruction. Chemotherapy was withheld owing to renal impairment. Despite supportive care, she deteriorated and succumbed approximately 11 months after the initial presentation.

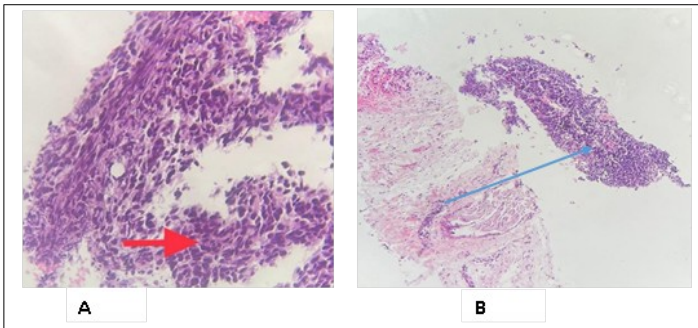

Figure 1. **A)** Immunohistochemistry images showing undifferentiated neoplastic cells with strong p16 in K4 expression and CD99 Golgi positivity; **B)** Patchy synaptophysin expression

Discussion

This is the first case of uterine PNET reported at the Kenyatta National Hospital, the largest teaching and referral hospital in East Africa. Similar to the findings by Park et.al (3), uterine PNETs occur in adolescents and postmenopausal women, often presenting with abnormal vaginal bleeding and uterine masses that mimic common gynecologic conditions. In this case, the patient presented with heavy vaginal bleeding that was initially attributed to a hormonal imbalance. Ultrasound findings suggested disseminated endometriosis, and she was treated with combined estrogen and progesterone without improvement. Subsequent MRI suggested endometrial lymphoma, and by the

time a histological diagnosis was confirmed, the disease had significantly progressed.

Even after diagnosis, treatment remained challenging owing to the lack of consensus on optimal chemotherapy regimens, although cisplatin and etoposide have been used in some cases. Targeted therapies have been suggested to improve survival and quality of life (5), and when detected before metastasis, there may be relatively favorable responses to an intensive multimodality treatment (6). Currently, however, no standardized diagnostic, therapeutic, and follow-up protocols exist (2). The management of this patient was guided by published case reports, including a report of pulmonary PNET successfully treated with three cycles of neoadjuvant chemotherapy followed by surgical resection, with disease-free survival at 3 years (7). Adjuvant chemotherapy was also administered based on a multidisciplinary consensus, consistent with recommendations for multimodal treatment approaches.

Despite neoadjuvant chemotherapy, extensive surgery, and adjuvant therapy, the disease recurred aggressively, leading to obstructive uropathy with acute kidney injury that precluded further chemotherapy. Dialysis was deemed futile given the persistent obstructive uropathy and the absence of evidence supporting clinical benefit. The patient deteriorated and succumbed approximately five months after repeat surgery and 11 months after initial diagnosis. This tendency to recur with a more aggressive disease has been documented in published cases. Elizalde et.al (6) described a case of a 60-year-old postmenopausal woman who developed rapidly progressive disease after primary surgery and died four months later (7 months from diagnosis) despite aggressive treatment.

Conclusion

Uterine PNETs are aggressive tumors that rapidly infiltrate and obstruct pelvic structures. Owing to their rarity, diagnosis and management remain challenging, with high mortality.

Consent for Publication

Informed consent for publication was obtained from the patient's next of kin.

Conflicts of interest

The authors have conflicts of interest to declare.

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