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A Case Report and Literature Insight on Primary Urachal Adenocarcinoma

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Abstract **Case Report**

Primary urachal adenocarcinoma is an uncommon malignancy arising from the urachus, constituting fewer than 1% of bladder malignancies and posing significant diagnostic and treatment challenges. We present the case of a 41-year-old female patient diagnosed with poorly differentiated tubulovillous adenocarcinoma of the urachus, who had recurrent non-clotting hematuria and urinary tract symptoms. She had surgery and got adjunctive chemotherapy. In November 2021, she received a diagnosis of peritoneal carcinomatosis. She began first-line chemotherapy and maintenance therapy. Regrettably, the patient was not reachable for further evaluation. She was readmitted to the hospital with declining health and passed away five days after admittance. Primary urachal adenocarcinoma (PUA) is an uncommon neoplasm, constituting 0.2% of urinary tract malignancies and 0.01% of all cancers. It manifests in adults, with the peak incidence occurring in the fifth and sixth decades of life. The pathophysiology entails the malignant transformation of urachal leftovers, which are postnatal embryonic structures. Diagnosis is complex and requires a multidisciplinary approach, including imaging, histological evaluation, and radiation. Treatment includes surgical intervention, chemotherapy, radiation, and palliative care. The prognosis is often unfavorable, with a five-year survival rate between 40% and 60%. In conclusion, primary urachal adenocarcinoma (PUA) is an uncommon, aggressive malignancy with a dismal prognosis. Timely identification enhances results; yet, treatment protocols are still insufficient. Future advancement relies on research, early detection techniques, individualized therapies, and heightened clinician awareness.

Keywords: Urachal, chemotherapy, adenocarcinoma, oncology.

INTRODUCTION

Primary urachal adenocarcinoma (PUA) is an exceedingly rare malignancy originating in the urachus, a remnant of the embryonic allantois and cloaca. The urachus usually disappears after birth, leaving only the median umbilical ligament [1]. But in some cases, it stays and can become a site for several diseases, including cancer. Accounting for less than 1% of all bladder cancers, PUA represents a diagnostic and therapeutic challenge due to its low incidence and nonspecific clinical presentation [2]. Here, we present a case of primary urachal adenocarcinoma, emphasizing its clinical presentation, diagnostic workup, surgical management, and histopathological findings. This report aims to contribute to the growing body of literature on PUA.

CASE PRESENTATION

A 41-year-old female patient, with no significant pathological history, presented to the emergency department in June 2020 with recurrent non-clotting hematuria associated with lower urinary tract symptoms, without any other associated signs, all occurring in the context of stable general condition. An ultrasound showed thickening of the bladder wall with a basal tissue lesion and involvement at the bladder horn. A uro-CT scan revealed a large extra-vesical mass located intra-abdominally, cystic with solid elements inside, invading the bladder at its upper pole without any other associated signs.

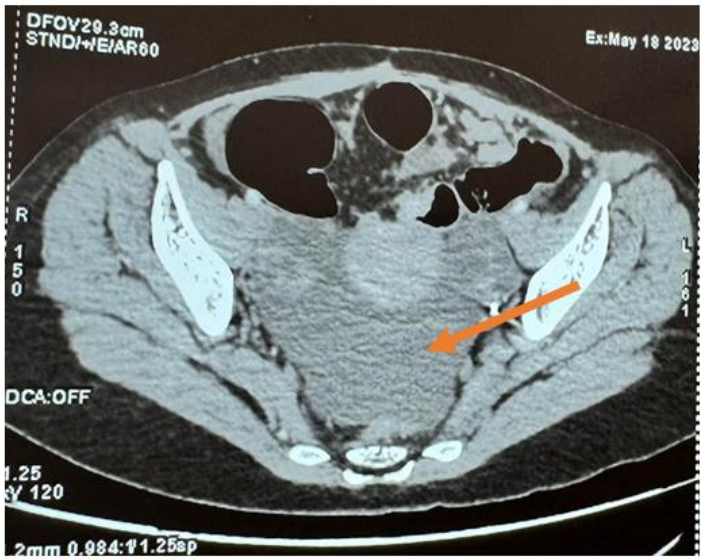

Fig 1: Abdominal CT showing mass at the dome of the bladder

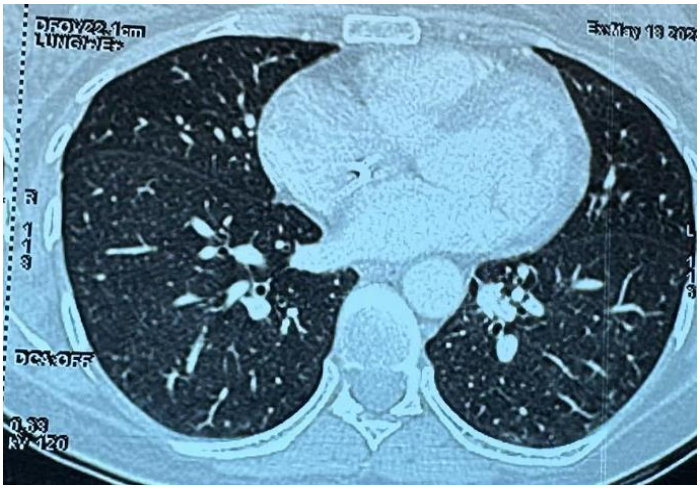

Fig 2: Chest-CTscan showing no secondary pulmonary lesions

The patient underwent exploratory laparotomy with a biopsy of the mass at the level of the urachus. Histopathological analysis was consistent with poorly differentiated tubulovillous adenocarcinoma of the urachus, CK7 positive.

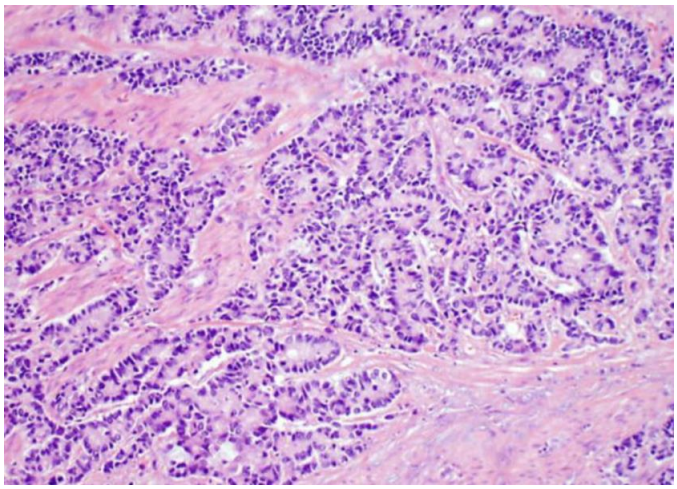

Fig 3: Pathology -Primary urachal adenocarcinoma

The CA125 was 35.71 U/ml, while AFP and BhCG were negative. The patient was operated in May 2020 with a partial cystectomy and left oophorectomy. The histopathological result from surgery favored a mucinous colloid adenocarcinoma measuring 12 × 10 × 6 cm, with no vascular emboli or perineural invasion. The left ovary showed no malignancy, and lymph node dissection was negative (21 nodes negative on the right, 5 nodes negative on the left), with no RAS or BRAF mutations. The patient received adjuvant chemotherapy with FOLFOX regimen (days 1 and 15) from August 2020 to March 2021 and was then placed under surveillance.

In November 2021, the patient presented with a CT scan showing a radiological progression with two peritoneal nodules measuring 6.2 mm and 7 mm. The case was discussed at the oncology-urology staff meeting, and the decision was made to perform a cystoscopy, which came back normal, and to refer the patient for surgical totalization with intraperitoneal chemotherapy. However, the patient refused the surgery. A PET-CT scan was then performed, showing peritoneal nodules and pelvic effusion, suggesting peritoneal carcinomatosis. CA125 was elevated at 200 U/L and ACE at 102 U/L.

The patient was started on first-line chemotherapy with FOLFIRI and panitumumab, completing the chemotherapy in January 2024, with clinical and radiological stability, as well as normalization of tumor markers. She then began maintenance treatment with Fufol and panitumumab. Unfortunately, the patient was lost to follow-up.

She returned in February 2024 with a notable deterioration in overall status (PS 3), cachexia, abdominal distension, and respiratory distress. The decision was taken to admit her to the hospital and start palliative treatment until her health recovers. Regrettably, the patient succumbed five days post-admission.

DISCUSSION

Primary urachal adenocarcinoma (PUA) is a rare malignancy, accounting for 0.2% of all malignant tumors of the urinary tract and approximately 0.01% of all cancers. Among bladder cancers, PUA constitutes less than 1% of cases [3]. The incidence is estimated to be around 1 case per 5 million people annually, though this varies slightly by geographic region and study population [4]. PUA typically presents in adults, with the highest incidence in the fifth and sixth decades of life, and is slightly more common in males than females, with a male-to-female ratio of approximately 1.5:1 [5].

The pathophysiology of primary urachal adenocarcinoma (PUA) is the malignant transformation of urachal remnants, embryonic structures that remain postnatally. The urachus links the bladder dome to the umbilicus during fetal development but usually regresses before birth, resulting in a fibrous cord [6]. In some instances, fragments of the urachus survive, establishing a possible locus for cancer. The interplay of enduring urachal remnants, chronic inflammation, genetic alterations, and perhaps metaplasia fosters a milieu favorable to malignant transformation. The pathways implicated are analogous to those seen in colorectal and mucinous adenocarcinomas, indicating the glandular differentiation often noted in PUA [7]. Primary urachal adenocarcinoma (PUA) often presents with nonspecific symptoms, which can delay diagnosis. Symptoms typically result from the tumor's proximity to the bladder and urachus and its potential for local invasion [8]. While hematuria is the hallmark symptom of PUA, a combination of urinary symptoms, abdominal pain, or palpable mass should prompt investigation. Early detection is challenging due to the overlap with benign conditions like UTIs or bladder inflammation. The diagnosis of primary urachal adenocarcinoma (PUA) is complex and requires a multidisciplinary approach combining clinical evaluation, imaging studies, and histopathological analysis.

The diagnosis of primary urachal adenocarcinoma (PUA) relies on Sheldon's criteria, which include [9]:

- Tumor location in the bladder dome or anterior wall.
- Presence of urachal remnants in or near the tumor.
- No primary involvement of the bladder urothelium.
- Tumor extension beyond the bladder.
- Exclusion of metastases from other primary adenocarcinomas.

Imaging (ultrasound, CT, MRI), cystoscopy, and histopathological examination are crucial tools for confirming the diagnosis and differentiating PUA from other cancers.

Histologically, PUA is most commonly mucinous and exhibits glandular differentiation with variable IHC expression. A combination of histological patterns and IHC markers is used to confirm its diagnosis and origin.

The Mayo Clinic staging system is favored for its simplicity, while the Sheldon system provides a more detailed description of disease spread. Accurate staging is critical for prognosis and guiding treatment decisions [10].

The treatment of primary urothelial carcinoma (PUC) relies on a multimodal strategy. In the adjuvant context, surgery (partial or radical cystectomy) is followed by chemotherapy, often using regimens such as FOLFOX or CAPOX, especially in cases with a significant risk of recurrence [11]. Radiotherapy is used

sparingly, especially for cases with good surgical margins. For metastatic illness, chemotherapy is the primary treatment, using FOLFOX or CAPOX, with targeted medicines such EGFR inhibitors (e.g., cetuximab) or PD-1/PD-L1 inhibitors (e.g., pembrolizumab) for particular mutations [6]. Palliative care is crucial for symptom management. Clinical trials and genetic profiling provide novel treatment alternatives.

The prognosis for urachal carcinoma is generally poor, with a five-year survival rate ranging from 40% to 60% [12]. Key prognostic factors include tumor size, stage at diagnosis, and the presence of metastases. Early-stage tumors tend to have a better prognosis, underscoring the importance of early detection. A significant challenge in managing primary urachal adenocarcinoma (PUA) is the lack of specific treatment guidelines, owing to its rarity. Further research is needed to better understand the biology of PUA and to develop targeted therapies. Additionally, raising awareness among clinicians about this rare cancer can lead to earlier diagnoses and, potentially, improved outcomes.

CONCLUSION

In conclusion, primary urachal adenocarcinoma (PUA) is a rare and aggressive cancer with a poor prognosis, marked by a five-year survival rate of 40% to 60%. Early detection significantly improves outcomes, but challenges persist due to the lack of specific treatment guidelines. Current management relies on surgery and chemotherapy regimens similar to those for other cancers, but there is a pressing need for targeted therapies. Future progress depends on further research into PUA's biological mechanisms, better early detection methods, and the development of personalized treatments. Increasing clinician awareness and international collaboration are key to improving patient outcomes.

Consent: Written informed consent is obtained from the patient for publication and any accompanying images.

Ethical Approval: This study is exempt from ethical approval in our institution since it doesn't involve experimental treatment.

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Conflict of Interest Statement: None

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