中文題目:老年原發性輸尿管神經內分泌腫瘤:個案報告 英文題目: Primary Neuroendocrine Carcinoma of the Ureter in elderly-A Case Report 作 者:林楷傑¹, 吳珮瑜², 蔡宜純^{1,3,4} 服務單位:¹高雄醫學大學附設醫院內科部,²高雄市立小港醫院腎臟內科,³高雄 醫學大學附設醫院腎臟內科,⁴高雄醫學大學附設醫院一般醫學內科

Introduction:

Neuroendocrine carcinoma (NEC) of the urinary tract is extremely rare as it accounts for less than 0.05% of primary urinary tract malignancies. Among NEC, reports addressing the cases that originate from the ureter are limited. We reported a case of a 97-year-old female with ureteral NEC who had an initial presentation of recurrent urinary tract infection (UTI). To our knowledge, this is the oldest age to be reported. Furthermore, we discovered the rapid progressive disease developed over a span of 4 months with evidence of imaging documentation.

Case Report:

A 97-year-old female had significant past medical history for controlled hypertension and rheumatoid arthritis. She had recurrent urinary tract infection two times in the past 4 months, both with the same urinary bacterial culture of Escherichia coli (E. coli). This time, she presented to our emergency room with fever, chills and consciousness disturbance for 2 days. Upon arrival, her body temperature was 38.7°C, and had tachycardia with blood pressure of 87/51mmHg. Laboratory findings showed elevated lactate (3.81mmol/L) and C-reactive protein level (163.16mg/L). Urine analysis showed pyuria and was also positive for leukocyte esterase and nitrite. She was admitted to the intensive care unit with a diagnosis of severe urosepsis.

Urinary tract ultrasound showed left side hydronephrosis and hydroureter. Abdominal computed tomography (CT) demonstrated some soft tissue lesions in the left upper third ureter with upstream obstructive nephropathy (Figure 1), and was highly suspicious for malignancy.

Therefore, ureterorenoscopic tissue biopsy was performed and left double-J tube was placed for left ureter stricture. The ureter biopsy specimen revealed primary small cell carcinoma (Figure 2). Microscopic examination revealed infiltrating solid nests in growth pattern with small- to large-sized hyperchromatic nuclei, indistinct nucleoli and little cytoplasm. Focal tumor necrosis was also seen but there was no lymphovascular invasion nor perineural invasion. The immunohistochemical (IHC) studies showed dot-like positive of Cytokeratin (AE1+AE3), positive of Synaptophysin and CD56, and focally positive of Chromogranin A. Based on histological features and IHC profile, the diagnosis of NEC of the ureter was established. However, due to the patient's advanced age and comorbidities, she refused to receive further treatment for the cancer.

Conclusion:

Ureteral NEC is an aggressive form of cancer with poor prognosis, yet diagnosis is difficult in the early stages of the disease. Our patient, a 97-year-old female with a rapid disease progression of 4 months, is the oldest age to be recorded to date. The present case emphasizes that albeit rare, nephrologists and urologists must always keep in mind the possibility of ureter NEC when dealing with urinary tract malignancies in adults.



Figure 1. Representative computed tomography (CT) images of patient prior and upon diagnosis. The coronal section (A) and axial section (B) of non-contrast abdominal CT of the patient on November 2018, 4 months prior to cancer diagnosis.

The coronal section (C) and axial section (D) of non-contrast abdominal CT revealed some soft tissue lesions (white arrows) in the left upper third ureter with upstream obstructive nephropathy upon diagnosis of ureteral neuroendocrine carcinoma.



Figure 2. Pathology from patient's ureter biopsy. (A) Hematoxylin–eosin stain (H&E) of the ureter tumor in gross look in 40x. (B) growth pattern with small- to large-sized hyperchromatic nuclei, indistinct nucleoli and little cytoplasm (H&E 400x). (C,D,E) The immunohistochemical (IHC) study showed positive of CD56, Synaptophysin, and focally positive of Chromogranin A. (F) The dot-like positive of Cytokeratin (AE1+AE3).