Solitary late metastasis of primary renal cell carcinoid tumor to the extraocular muscles imaged with indium-111 octreotide

ABSTRACT
A 75-year-old male presented with right eye pain and proptosis. His history was significant for renal cell carcinoma treated with left nephrectomy 2 years previously. Computed tomography (CT) imaging demonstrated a 1.6-cm enhancing lesion in the right retrobulbar space. Surgical biopsy revealed a low-grade metastatic carcinoid tumor. At retrospective review, the primary renal lesion from 2 years prior was rediagnosed as consistent with a renal carcinoid tumor. Indium-111 octreotide single photon emission CT/CT imaging demonstrated a solitary metastasis within the right extraocular muscles. The patient subsequently developed additional metastases within the remaining right kidney and lung.

Keywords: Indium-111 octreotide, ocular metastasis, renal carcinoid tumor

INTRODUCTION
Carcinoid tumours are a type of neuroendocrine tumour that most commonly arise from the gastrointestinal tract. They are usually slow growing but may present with metastasis.[1] This case details a carcinoid tumour arising from the kidney, which is an uncommon location, and presenting with a rare site of metastasis – the extraocular muscles.

CASE REPORT
A 75-year-old male presented with right eye pain and proptosis. His medical history was significant for left nephrectomy for “papillary renal cell carcinoma” (RCC) 2 years previously. Computed tomography (CT) at that time demonstrated a 9.5-cm mixed attenuation lesion in the left kidney consisting of a solid component and intralesional hemorrhage as an incidental finding during the investigation of an episode of acute diverticulitis. Left nephrectomy was performed, and histology was reported as a 2.5-cm papillary RCC with hemorrhagic component.

On this admission, contrast CT of the orbits demonstrated an enhancing 1.6-cm lesion in the right retrobulbar space [Figure 1a and b]. Surgical biopsy revealed a metastatic low-grade neuroendocrine tumor. Immunohistochemistry was positive for chromogranin and synaptophysin with a proliferation index of 5%. Hydroxyindoleacetic acid was normal at 33 (normal range 2.5–50 µmol/24 h). Subsequent review of the initial left kidney histology with immunohistochemistry revealed a similar morphology to the metastatic deposit in the extraocular muscle, and this was retrospectively reclassified as a low-grade carcinoid tumor. Indium-111 (In-111) octreotide planar and single photon emission CT/CT imaging [Figure 1c-e] at 24 h demonstrated a solitary metastasis in the right retro-orbital soft tissues.

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As the patient was systemically well and histology identified a low-grade tumor, initial treatment was with radiotherapy to the orbital metastasis for symptomatic and local control. The patient remained well on 3-monthly surveillance; however, follow-up octreotide imaging 2 years later demonstrated the development of metastatic disease with avid right renal and pulmonary lesions [Figure 2], at which time systemic treatment with octreotide was commenced.

Carcinoid tumors of the renal origin are extremely rare. 28.6% of cases are diagnosed as an incidental finding in asymptomatic patients.[1-4] In a relatively large series (n = 56), renal carcinoid was misdiagnosed initially as RCC in 15% of cases.[2] A clinicopathological case series reported similar findings - for example, a renal carcinoid was initially diagnosed as papillary RCC due to a “pseudopapillary” appearance but subsequently reclassified after examination of a metastatic liver deposit.[5] Carcinoid tumors occur mostly occur in the gastrointestinal tract or bronchial system of the lungs. Primary genitourinary carcinoids occur in < 1%.[2,6] Liver metastases are most common and can lead to carcinoid syndrome and right-sided valvular heart disease. While 68gallium-labeled somatostatin analogs have become the current standard practice,[7] In-111-labeled octreotide provided the sufficient information to establish a diagnosis in this case. To our knowledge, this is the first case to describe isolated extraocular muscle metastasis from a primary carcinoid tumor with In-111-labeled octreotide, or with 68gallium-labeled somatostatin analogs.[8] Metastases to the orbits have been reported as small case series or case reports but typically occur in the setting of advanced metastatic disease and affect more than one extraocular muscle.[5,9,10]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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