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SHORT COMMUNICATION

Metastatic renal cell carcinoma presenting as a circumscribed orbital mass

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PURPOSE. *To report a case of renal cell carcinoma presenting as a well-circumscribed orbital tumor.*
METHODS. *Retrospective interventional case report.*
RESULTS. *A 60-year-old woman presented with proptosis of the left eye. Imaging showed a well circumscribed tumor in the region of the medial rectus muscle. Excision biopsy revealed a diagnosis of metastatic renal cell carcinoma that was confirmed on abdominal imaging.*
CONCLUSIONS. *Renal cell carcinoma can rarely present as a well-circumscribed orbital mass and should be included in the differential diagnosis of such lesions. (Eur J Ophthalmol 2008; 18: 483-5)*

KEY WORDS. *Renal call carcinoma, Metastasis, Orbit*

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INTRODUCTION

Metastatic lesions of the orbit are rare and account for only 3–7% of orbital tumors (1). Metastasis of renal cell carcinoma to the orbit is exceedingly uncommon, usually presenting in male patients as a diffuse mass. We report the case of a well-circumscribed orbital mass in a female patient that was the first presentation of renal cell carcinoma (RCC).

Case report

A 60-year-old woman presented with a 4-month history of diplopia and proptosis of the left eye. Her medical history was significant for hypertension and migraine. Horizontal diplopia was present on right gaze and the patient complained of discomfort but did not have significant pain. On examination, her best-corrected visual acuity in each eye was 6/9. She had 2 mm left proptosis with restricted adduction of the left eye and diplopia on right gaze. Chemosis and injection of conjunctival and episcleral vessels were noted nasally (Fig. 1A). The remainder of the ophthalmic examination was unremarkable.

The full blood count, serum antithyroid antibodies, and serum TSH levels were normal. Computerized tomography of the orbit demonstrated a well-circumscribed enhancing mass in the region of the anterior medial rectus muscle on the left side (Fig. 1, B and C). Based on a presumptive diagnosis of myositis, the patient was treated with oral corticosteroids for 3 weeks with no response. She was then referred to one of the authors (G.J.D.) for further assessment. Doppler ultrasound of the left orbit showed fusiform enlargement of the medial rectus muscle without significant internal vascularity and magnetic resonance imaging showed a well-defined enhancing mass in the region of the medial rectus muscle. Intramuscular cavernous hemangioma was considered to be the most likely provisional diagnosis.

Excision of the lesion was performed via a transcaruncular approach. The tumor abutted the medial rectus muscle at the musculo-tendinous junction and was well defined except where it was adjacent to the medial wall of the orbit. Macroscopic excision was completed with the aid of a cryoprobe. Significant bleeding was encountered during dissection of the medial aspect of the tumor.

Histopathologic examination of the excised specimen showed a circumscribed tumor surrounded by a thick layer of fibrous tissue forming a pseudocapsule (Fig. 1E). The tumor was composed of sheets of cells with clear cytoplasm, round to oval nuclei, small nucleoli, and even

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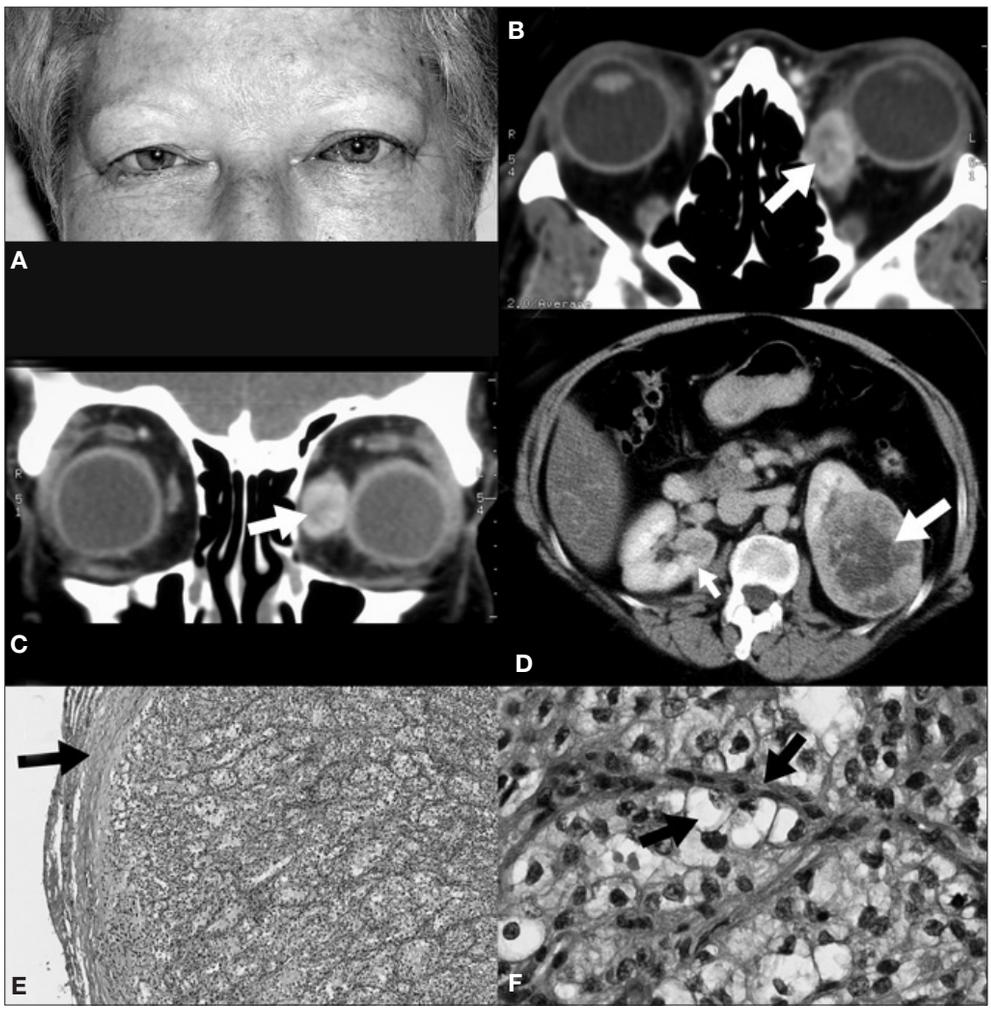

Fig. 1 - (A) *Clinical photograph showing left proptosis and congestion of nasal conjunctiva on the left side.* **(B, C)** *Axial (B) and coronal (C) computed tomographic scans demonstrate a well-circumscribed enhancing mass in the region of the left medial rectus muscle.* **(D)** *Computed tomographic scan of abdomen shows a large left renal mass consistent with a diagnosis of renal cell carcinoma (large arrow). A smaller lesion is also noted in the right kidney (small arrow).* **(E)** *Photomicrograph of excised orbital lesion illustrates a well-circumscribed tumor mass surrounded by a fibrous capsule (arrow) and composed of sheets of clear cells (hematoxylin-eosin, x100).* **(F)** *Higher magnification photomicrograph of the same specimen showing sheets of polygonal clear cells with small nuclei separated by fine fibrovascular septae (arrows).*

chromatin (Fig. 1F). Numerous capillary-size vessels were noted with areas of hemorrhage. Mitotic figures were scant. Immunohistochemical stains were performed and the tumor was positive for CAM 5.2, AE1/AE3, and CD10. These features were characteristic of a clear-cell carcinoma of renal origin.

The patient underwent computed tomography of the chest and abdomen that showed a left renal mass lesion in keeping with renal cell carcinoma with extension into the left proximal renal vein and a small lesion in the right kidney (Fig. 1d), and multiple hypervascular lesions within the liver and lungs, suspicious of further metastatic lesions. The patient was referred to medical oncology for further management and is on novel antiangiogenesis agents, sunitinib and sorafenib.

DISCUSSION

We report an unusual manifestation of RCC that presented as a well-circumscribed orbital mass adjacent to the medial rectus muscle/tendon. RCC very rarely metastasizes to the orbit and in their comprehensive review on the subject Shome and coworkers could find only 27 reported cases (2). Males are affected in more than 90% of cases and the orbital metastasis may occasionally be the presenting manifestation (2). The orbital mass is usually diffuse, although a circumscribed mass has been documented (3, 4). Radiologically, the mass could not be distinguished from the adjacent muscle, leading to the initial diagnosis of myositis. Circumscribed orbital masses that show enhancement on contrast are usually of vascular

origin, commonly cavernous hemangioma and hemangiopericytoma. The differential diagnosis also includes peripheral nerve sheath tumors, fibrous histiocytomas, lymphomas, and metastases.

Histologic examination revealed a clear-cell tumor and this greatly helped narrow the differential diagnosis since clear cell lesions are rare in the orbit. These include metastasis from RCC, alveolar soft part sarcoma, granular cell tumor, and paraganglionoma. Though morphologic differences exist between these lesions, today the definitive diagnosis is based on the immunohistochemical findings.

Metastatic RCC is an aggressive disease, and even in the recent past, was associated with a very poor prognosis, with a 5-year survival of less than 10%. However, novel antiangiogenic agents such as sunitinib and sorafenib, which inhibit VEGF and PDGF pathways, have improved the progression-free survival in these patients (5).

This case demonstrates that RCC metastasis can present as a circumscribed orbital mass and should be included in the differential diagnosis of intramuscular tumors.

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