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Amaurosis fugax as initial symptom of metastatic clear cell renal carcinoma

Amaurose fugaz como sintoma inicial de carcinoma de células renais metastático

José Renan Miranda Cavalcante Filho¹, Rafael Paiva Arruda², Elson Arruda Linhares³,
Paulo Roberto Lacerda Leal⁴, Irismar de Azevedo Andrade², Karlos Jennysson Sousa Soares³

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RESUMO: *Introdução:* Neste caso, descrevemos a apresentação rara da metástase orbitária de um carcinoma renal de células claras. *Apresentação do caso:* Apresentamos um caso de um homem de 57 anos de idade, que subitamente perdeu a visão em seu olho esquerdo associada à paralisia da abdução do mesmo. A ressonância magnética do crânio revelou um tumor intra-orbitário esquerdo com dimensões de 21,8 x 19,6 mm, provocando um deslocamento centro-lateral com consequente compressão do nervo óptico e do músculo reto lateral homolateral. O paciente foi submetido à remoção cirúrgica da lesão. O diagnóstico histológico indicou metástase de carcinoma renal de células claras confirmada pela tomografia computadorizada abdominal (TC), que apresentou neoplasia no terço superior do rim esquerdo, medindo 3,5 x 3,4 cm com reforço heterogêneo após administração intravenosa de contraste. *Conclusão:* Este caso descreveu a apresentação rara de uma metástase orbital de carcinoma renal de células claras. A apresentação inicial do paciente foi amaurose fugax e paralisia homolateral do nervo abducente. A metástase orbital de carcinoma renal de células claras é rara, no entanto, o exame histológico concluiu o diagnóstico.

Descritores: Carcinoma de células renais; Neoplasias renais/diagnóstico por imagem; Amaurose fugax; Neoplasias orbitárias; Doenças do nervo abducente.

ABSTRACT: *Background:* In this case we describe the rare presentation of orbital metastasis of a clear cell renal carcinoma. *Case Report:* We present a case of a 57-year-old male with suddenly lost the vision in his left eye and homolateral abducens palsy. The magnetic resonance imaging of the skull revealed a left intraorbital tumor with center-lateral displacement measuring 21.8 x 19.6 mm, compressing the optic nerve and the homolateral lateral rectus muscle. The patient underwent surgical removal of the lesion. The histologic diagnosis indicated clear cell renal carcinoma metastasis confirmed by the abdominal computed tomography (CT), which showed a neoplasia in the upper third of the left kidney measuring 3.5 x 3.4 cm with heterogeneous reinforcement after intravenous contrast administration. *Conclusion:* This case described the rare presentation of a clear cell renal carcinoma metastasis in orbit. The initial presentation of patient was amaurosis fugax and homolateral abducens palsy. Orbital metastasis of clear cell renal carcinoma are rare, however, the histologic exam conclude the diagnosis.

Keywords: Carcinoma, renal cell; Kidney neoplasms/diagnostic imaging; Orbital neoplasms; Amaurosis fugax; Abducens nerve diseases.

1. Residente de Neurocirurgia do Hospital Regional de Presidente Prudente, SP. ORCID: <https://orcid.org/0000-0002-8656-5488>. Email: renanmiranda@hotmail.com.
2. Urologista da Santa Casa de Misericórdia de Sobral, Sobral, CE. ORCID: Arruda RP - <https://orcid.org/0000-0001-6440-7350>; Andrade I - <https://orcid.org/0000-0001-7226-1822>. Email: rafa.rpa@gmail.com, irismar_andrade@yahoo.com.br
3. Cirurgião Geral da Santa Casa de Misericórdia de Sobral, Sobral, CE. ORCID: Linhares EA - <https://orcid.org/0000-0002-7416-6391>; Soares KJS - <https://orcid.org/0000-0003-1704-9137>. Email: coelho.linhares@ig.com.br; kajotass@yahoo.com.br.
4. Neurocirurgião da Santa Casa de Misericórdia de Sobral, Sobral, CE. Orcid: <https://orcid.org/0000-0002-2489-3777>. Email: prlleal@hotmail.com

Correspondence: José Renan M. Cavalcante Filho. Rua Jorge Gushiken, 405 – Condomínio Vila Paris – Casa 20. Presidente Prudente, SP.

INTRODUCTION

Renal cell carcinoma (RCC) accounts for 3% of all cancers in adult patients and it is twice as frequent in males. Clear cell renal carcinoma (CCRC) is most prevalent variant of this neoplasia, responsible for 70 to 75% of all cases¹. As the most frequent metastasis are breast (48%), prostate (12%), skin (12%) and lungs (8%)². Orbital metastasis are rare and represents less than 2% of all ophthalmic metastasis. We report the case of a patient with renal cell carcinoma who presented the symptoms of orbital metastasis as the initial manifestation of the disease.

CASE REPORT

Male, 57 years old, without comorbidities (systemic arterial hypertension, diabetes mellitus), who did not take any medication for chronic conditions, and ex-smoker, attended the Neurosurgery Department at the *Santa Casa de Misericórdia de Sobral* Hospital. This patient reported that six months ago suddenly lost the vision in his left eye and that it had been deviated medially (Figure 1), with no other associated symptoms.



Figure 1. Patient with esotropia of the left eye due to involvement of the ipsilateral lateral rectus muscle

Upon admission, the patient’s only complaint was of amaurosis of the left eye. He denied other symptoms: hematuria, dysuria, or back pain. Physical examination showed no visceromegaly or abdominal mass. The magnetic resonance imaging of the skull revealed a left intraorbital tumor with center-lateral displacement measuring 21.8 x 19.6 mm, compressing the optic nerve and the homolateral lateral rectus muscle (Figure 2). From the results of the exam, surgical removal of the orbital tumor was indicated (12/02/2016). The histopathological report was compatible with CCRCC and confirmed with

immunohistochemistry (Table 1).

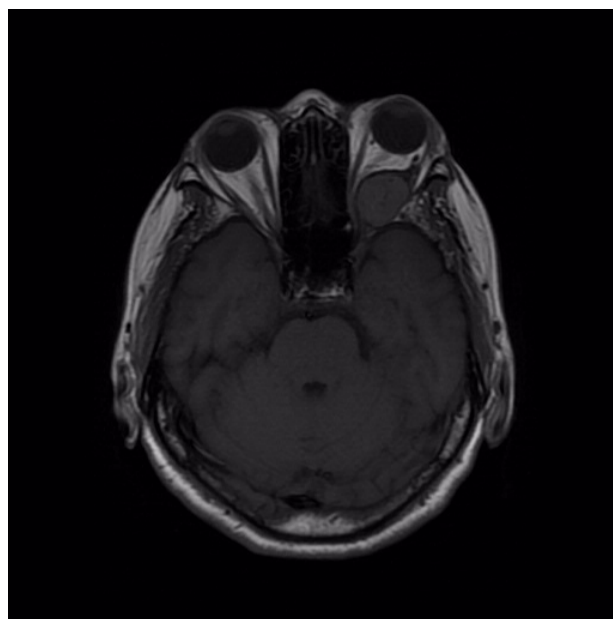


Figure 2. T1-weighted magnetic resonance imaging showing metastatic tumor from renal cell carcinoma in the left orbit. Center-lateral displacement compressing the optic nerve and the lateral rectus muscle. Dimensions: 21.8 x 19.6 mm

Table 1: Left-orbit immunohistochemistry report

Accompanying Files		
Antibody	Clone	Interpretation
RCC	PN15	Positive
EMA	E29	Positive
CK7	OV-TL-12/30	Negative
TTF-1	8G7/1	Negative
VIMENTINA	V9	Negative
CD10	56C6	Negative
PAX8	MRQ-50	Negative
CK20	K20.8	Negative

The clinical investigation of the patient proceeded with the request of an abdominal computed tomography (CT), which showed (neoplasia in the upper third of the left kidney measuring 3.5 x 3.4 cm with

heterogeneous reinforcement after intravenous contrast administration (white arrow) (Figure 3) and a chest CT (nodular image with homogeneous reinforcement after the use of intravenous contrast in the anterior segment of the upper lobe of the left lung. Suggestive of metastatic nodule, associated with left perihilar lymph node enlargement). According to findings from the complementary exams, partial nephrectomy surgery for tumor resection in the left kidney was indicated (30/08/2016).

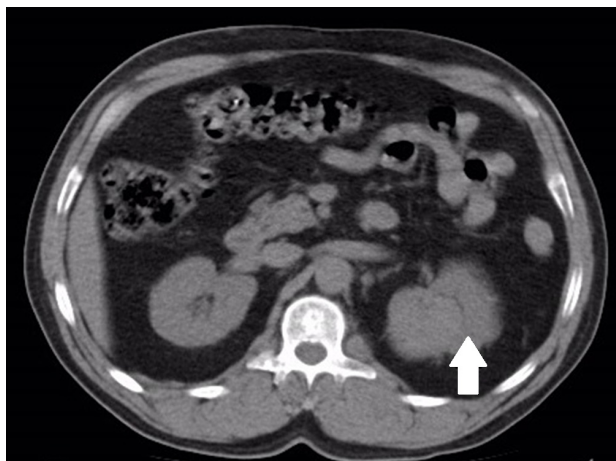


Figure 3. Axial section of computed tomography of abdomen revealed tumor mass in the left kidney measuring 3.5 x 3.4 cm

DISCUSSION

Although the case reported belonged to the epidemiological group most commonly affected by RCC (male and in the 5th and 6th decade of life), it is worth highlighting due to its unusual clinical presentation and atypical evolution of ocular symptoms. Classically, the most characteristic symptoms of metastatic orbital RCC are: proptosis, ptosis, and pain, all of which are absent in this report. Another unusual feature present in this case was the sudden onset of symptoms, amaurosis fugax in the left eye and ipsilateral esotropia due to involvement of the optic nerve and lateral rectus muscle, respectively.

RCC treatment depends on the evolution stage of the disease, and it presents options for clinical and surgical treatments. Molecular-targeted therapies are currently the clinical treatment of choice. They are more effective against RCC than chemotherapy, with response rates of approximately 40 and 10%, respectively. There are several ongoing studies investigating new drugs for the systemic treatment of RCC³.

Until recently, complete RCC resection was gold standard therapy in cases with strictly localized disease.

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Surgery occurred adequately, without complications, and the patient was discharged after 4 days. Histopathology of the tissue removed during nephrectomy revealed renal parenchyma, hosting a neoplasia of epithelial origin composed of atypical cells, irregular nuclei and macronuclei, with abundant clear cytoplasm in tubular and cordonal arrangements suggestive of CCRC (Figure 4).

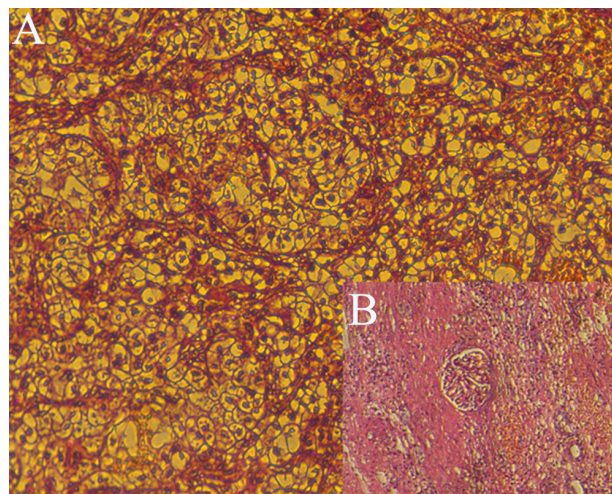


Figure 4. A: Histopathology of the tissue removed during partial nephrectomy of the left kidney Renal parenchyma hosting neoplasia of epithelial origin composed of atypical cells, irregular nuclei and macronuclei, with abundant clear cytoplasm in tubular and cordonal arrangements suggestive of Clear Cell Renal Carcinoma. B: Some healthy glomerular regions interspersed with others affected by neoplasia can be seen. H&E staining x20

However, it is now known that in smaller tumors it is possible to proceed with less aggressive approaches. In tumors of up to 4 centimeters, partial nephrectomy is chosen because there is no significant difference in survival, and long-term recurrent disease in detriment of radical nephrectomy⁴.

Surgical treatment also plays an important role in patients who already have distant metastasis. One study compared isolated immunotherapy versus nephrectomy plus immunotherapy in patients with metastatic RCC. The results showed greater long-term survival in the group of patients submitted to therapy combined with surgical approach⁴.

CONCLUSION

In this case report, even though the patient presented metastasis to lung and orbit after partial nephrectomy, he evolved with good postoperative recovery in a one-month segment; and the patient was referred to a reference center for oncology for post-surgical follow-up.

relationships and affiliations relevant to the subject matter or materials discussed in the article. No competing financial interests exist.

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