Coincidentally Found Choroidal Metastasis of Renal Cell Carcinoma with Mixed Clear Cell and Sarcomatoid Variant

Koroid Metastazı Tesadüfen Saptanan Böbrek Hücreli Kanserin Karışık Berrak Hücre ve Sarkomatoid Varyantı

Esat NAMAL[1]
Bennur ESEN[2]
Münevver Gül KAYA[2]
Özgül PAMUKÇU[3]
Emre YÜCE[2]

1 Bağcılar Education and Research Hospital, Department of Internal Medicine, Division of Medical Oncology, İstanbul, Turkey
2 Bağcılar Education and Research Hospital, Department of Internal Medicine, Division of Nephrology İstanbul, Turkey
3 Şişli Etfal Education and Research Hospital, Department of Internal Medicine, İstanbul, Turkey

ABSTRACT

Renal cell carcinoma (RCC) is the most common renal cancer that is usually metastatic at admission. Lung, bone and liver are most frequent involvement sites however the brain, skin and eye may be involved less frequently. Among different histologic subtypes, mixed clear cell and sarcomatoid type is presented as few case reports. In the present report, we present a case of RCC in mixed subtype that was coincidentally found with choroidal metastasis.

KEY WORDS: Renal cell carcinoma, Choroid metastasis, Clear cell-sarcomatoid variant

ÖZ

Böbrek hücreli kanser, böbreğin en sık görülen ve genellikle tanı anında metastatik kanseridir. Akciğer, kemik ve karaciğer en sık tutulan alanlar olmakla birlikte beyin, cilt ve gözde tutulabilir. Değişik histolojik alt tipler arasında karışık berrak hücreli ve sarkomatoid varyant sadece nadir olgu sunumları şeklinde görülmektedir. Bu olguda koroid metastazı tesadüfen bulunan karışık histolojik tip olan böbrek hücreli kanser sunmaktayız.

ANAHTAR SÖZCÜKLER: Böbrek hücreli kanser, Koroid metastazı, Berrak hücreli sarkomatoid varyant

INTRODUCTION

Renal cell carcinoma (RCC) is an aggressive malignant tumour that accounts for 3% of all adult cancers with a five-year survival of 10% (1). Approximately 25 % of all cases are metastatic at the time of diagnosis (2). The most common sites of metastasis are lung, bones, liver, and brain but any organ system or site may be involved. Eye involvement of RCC is not extraordinary but choroidal metastasis of the mixed clear cell and sarcomatoid variant is extremely rare. We present a case of RCC with choroidal metastasis without a complaint that may be attributed to eye involvement of RCC during the follow-up period.

CASE REPORT

A 49-year-old male presented with flank pain for the last 2 months. No abnormality was observed on laboratory analysis except mild creatinine elevation (creatinine: 1.5 mg/dl) and mild hematuria. His medical history was unremarkable except hypertension that was well controlled with amlodipine 10 mg/day. On ultrasonographic examination, two masses, one 80x86 mm in size in the upper pole and the other 120x75 mm size in the lower pole of the left kidney, were observed. Computed tomography scan revealed a nonuniform mass originating from the lateral cortex of the left kidney (Figure 1). Thorax computed tomography scan indicated 3.5x2.5 cm, bilateral, multiple nodular mass lesions suggesting pulmonary metastasis of RCC (Figure 2). The patient underwent left radical nephrectomy, and pathologic examination confirmed the diagnosis of mixed conventional (clear cell) and sarcomatoid variant. Fuhrman nuclear grade was 3 for clear cell variant and 4 for sarcomatoid variant. TNM staging of the

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Correspondence Address:
Bennur ESEN
Bağcılar Eğitim ve Araştırma Hastanesi, Nefroloji Bölümü, İstanbul, Turkey
Phone : +90 532 755 85 95
E-mail : bennuresen@yahoo.com

patient that had renal vein involvement was T3b, N0, M1 and he was referred to the oncology outpatient clinic with a diagnosis of stage 4 RCC. PET-CT scanning showed multiple metastases in the lungs, lymphadenomegalies related to metastasis at the right cervical, left supraclavicular and diaphragmatic site, a metastatic lesion at the L2 vertebra, and multiple metastatic involvement of muscular and soft tissues within the abdomen and thorax. Interferon (IFN-alpha) therapy was initiated which was then converted to sunitinib (a tyrosine kinase inhibitor) as a result of his intolerance to IFN treatment. His flank pain gradually improved. On MRI of cranium and orbita, a lesion placed at left bulbus oculi, at the superior margin of optic disc and extending into the vitreus humour was seen within the choroid tissue (Figure 3,4). He underwent radiotherapy for the choroidal metastasis that resulted with substantial improvement but required dexamethasone therapy due to development of optic neuropathy. At the second year of follow-up he was free of symptoms that may attributed to optic involvement of RCC. He died at the third year of diagnosis due to acute renal injury and pneumonia.

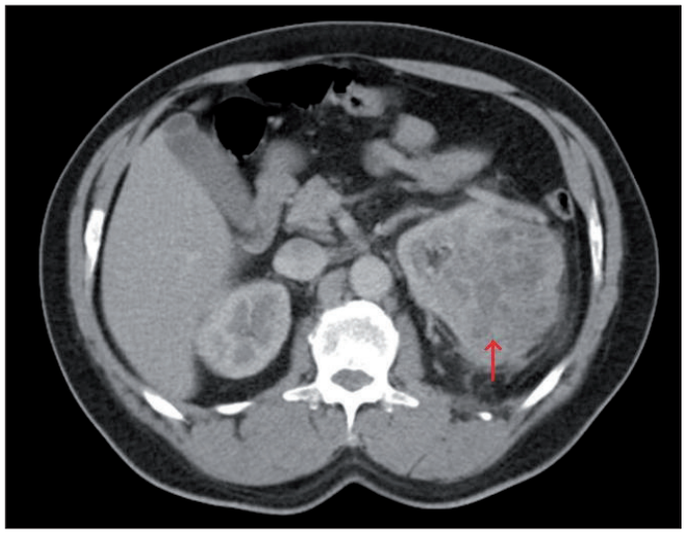


Figure 1: *Computed tomography showing nonuniform masses originating from the lateral cortex of the left kidney.*

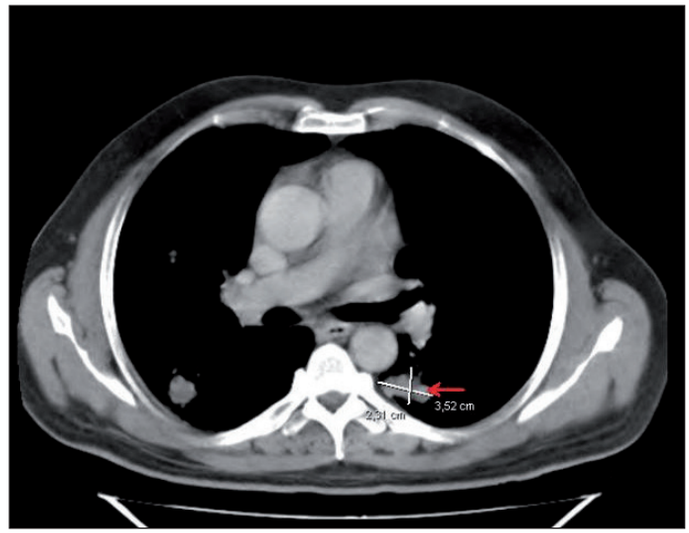


Figure 2: *Thorax computed tomography indicating 3.5x2.5 cm sized, bilateral, multiple nodular mass lesions suggesting pulmonary metastasis of RCC.*





Figure 3A,B: *MRI examination of cranium and orbita, a lesion placed at the left bulbus oculi, at the superior margin of optic disc and lies in the vitreous humour within the choroid tissue.*

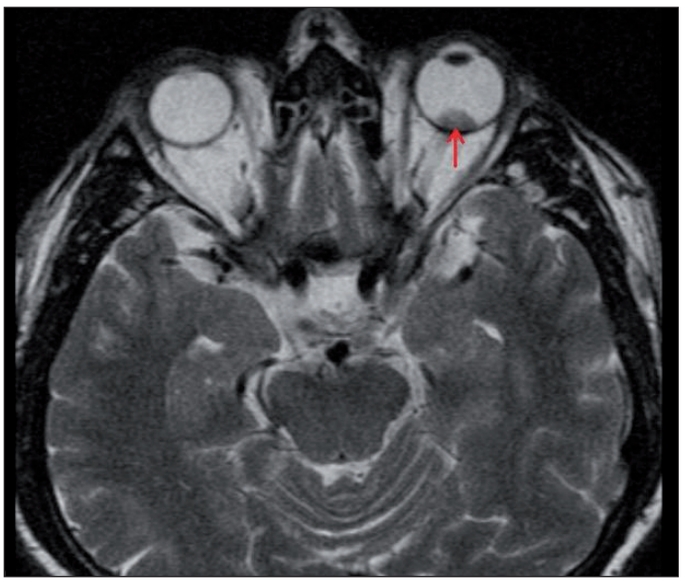


Figure 4: *Orbital MRI showing choroidal metastasis in the left bulbus oculi which penetrated into the vitreous humour within the choroid tissue.*

DISCUSSION

Although optic involvement of RCC is rare, choroid, iris and ciliary body are common intraocular metastasis sites that can be diagnosed at the initial evaluation as well as may be seen decades after nephrectomy. The most common presenting symptoms are pain, blurred vision, red eye or headache but a minority of cases may be asymptomatic. The choroid is the most common intraocular metastasis site because of the higher blood flow. Choroidal metastasis of RCC has no characteristic appearance, and patients may present with variable symptoms that may lead to confusion in the differential diagnosis.

Approximately half of RCC cases metastasize to the lung, bone and liver via the lymphatic or venous routes (2). Brain, cranium or orbita metastases account for 5% of all patients with RCC (3). The reported incidence of choroidal metastasis among all malignancies is 0.07% (2). Breast and lung malignancies are the most common primary tumors that are associated with choroidal metastases. The frequency of eye involvement in RCC is 3.6%. When all choroidal metastases are considered, RCC make up only 3% (4). Clear cell carcinoma is the most frequent histologic subtype, and the most common RCC type associated with choroidal metastasis (5). A minority of RCC cases may have mixed histologic type that may have different behaviour from conventional (clear cell carcinoma) type of RCC. Mixed clear cell +sarcomatoid subtype is extremely rare and there is limited data regarding the rate of choroidal metastasis.

Patients with choroidal metastasis of RCC have a poor prognosis with a median survival rate of 8 months (5). Extrenal radiotherapy is most appropriate treatment choice that is associated with symptomatic improvement. Improvement rate of symptoms such as visual loss and pain may vary between 26 and 81% (6). However a group of patients may require corticosteroid therapy to sustain improvement rates or to prevent neuropathy. Our patient experienced optic neuropathy that responded well to steroid administration. A small group of patient may require surgical procedures like enucleation (7) Removal of primary tumor may enable spontaneous regression of choroidal metastasis (8).

As any site may be involved in RCC, a multidisciplinary approach is necessary to avoid misdiagnosis of metastasis when a diagnosis of RCC is confirmed. Owing to the fact that patients with RCC may present with variety of symptoms depending on extension of metastasis, RCC is known as an 'internist tumor'. Regardless of symptoms at baseline evaluation or during the follow-up period, patients with RCC should be evaluated systematically at each outpatient visit.

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