Simultaneous choroidal melanoma and renal cell carcinoma in middle-aged patient

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ABSTRACT

BACKGROUND: Although rare, choroid melanoma is the most common intraocular malignancy amongst adults. Similarly, renal cell carcinoma (RCC) is another rare malignancy that counts for 2–3 percent of all adult malignancies. Second primary cancers are not uncommon, even though they occur less frequently than primary cancers.

CASE PRESENTATION: We present a rare choroidal melanoma case in a 55-year-old adult who presented with a one-month history of the painless gradual decrease of vision. His investigations revealed a clinical diagnosis of large choroid melanoma, and upon metastatic workup, he was incidentally found to have synchronous RCC. Both malignancies were treated surgically (and the pathology confirmed the clinical diagnosis of choroidal melanoma and RCC). The patient was free of metastatic or recurrent disease at 12 months follow-up.

KEY WORDS: uveal melanoma; renal cell carcinoma; synchronous

INTRODUCTION

Uveal melanoma (UM), the most common primary intraocular malignant tumor in adults, accounts for only five percent of all melanomas. The disease is more common in Caucasians [1]. In Jordan, the incidence of uveal melanoma is 1.39 cases per million population per year, which is less than in the Western Caucasian population, with a mean age of 45 years [2].

The tumor may involve any part of the uveal tract: choroid (85%), ciliary body (4–7%), or the iris (2–4%) [3]. Besides, small and medium-sized tumors are usually treated by Iodine-125 radioactive plaque therapy [4]. In contrast, large tumors are usually treated by enucleation [5], despite the fact that radiotherapy can achieve 90% local control of UM, up to 50 percent of patients develop hematogenous metastases in the decades following their initial diagnosis. These metastases affect the liver in over 90% of patients. Metastatic UM is always fatal, with an average survival time of fewer than six months [6]. Conversely, renal cell carcinoma (RCC) accounts for approximately 2% to 3% of all adult malignancies [7]. It is also worth noting that one-third of patients who undergo nephrectomy experience distant metastasis [8].

Importantly, both the detection of second primary cancer and the detection of early-stage primary...
cancer can improve survival. Second primary cancers are not uncommon, even though they occur less frequently than primary cancers [9–11]. A retrospective case review of 139 patients with uveal melanoma who were subsequently evaluated by whole-body [18-fluorine-labeled] 2-deoxy-2-fluoro-D-glucose (18FDG) positron emission tomography/computed tomography (PET/CT) imaging showed six patients (4.3%) had second primary cancers as (50%) were synchronous (found at initial staging), and the remaining 3 patients (50%) were metachronous (found at follow-up staging) [12].

We present a case of a middle-aged man who was seen in the King Hussein Cancer Center (KHCC) outpatient clinic in Amman/Jordan with these two malignancies and no other medical history. This case demonstrates the possibility of two different primary tumors coexisting in the same patient, which has significant implications for the individual management of these conditions.

**CASE PRESENTATION**

A 55-year-old gentleman who has no medical co-morbidities with an unremarkable family history of malignancies presented to the clinic with a one-month history of a painless drop of vision in the right eye. On eye examination, visual acuity for the right eye was 6/24, and the left eye was 6/9.

A slit-lamp exam showed a mild right eye cataract; a dilated fundus exam showed a right eye ciliary body melanotic mass on the temporal side with overlying exudative retinal detachment.

The left eye exam was unremarkable. Ocular B-scan ultrasound (U/S) and ultrasound biomicroscope (UBM) scans (Fig. 1) showed a right mushroom-shaped choroidal lesion associated with overlying exudative retinal detachment.

The right eye was unremarkable. Ocular B-scan ultrasound (U/S) and ultrasound biomicroscope (UBM) scans (Fig. 1) showed a right mushroom-shaped choroidal lesion associated with overlying exudative retinal detachment. The management plan was modified to treat the non-metastatic right ciliary choroidal melanoma separately from the second primary renal cell carcinoma. Accordingly, the multidisciplinary treatment plan for the large-sized ciliary body melanoma was right eye enucleation and orbital implant insertion, given the large tumor size beyond the radiotherapy capacity. The right RCC was planned for the right laparoscopic partial nephrectomy.

Therefore, the patient underwent a right laparoscopic partial nephrectomy with intraoperative ultrasound without complications. The pathology report confirmed the diagnosis of papillary RCC, type 2 ISUP grade 3, pT1aNX (Fig. 2).

![Figure 1A. B-scan ultrasound shows a mushroom-shaped choroidal lesion associated with overlying retinal detachment and low-to-moderate internal reflectivity suggestive of choroidal melanoma; B. 4x hematoxylin and eosin (H&E) slide (ciliary body malignant melanoma); Pigmented predominantly spindle cell tumor arranged in fascicles arising from the ciliary body; C. 40x (ciliary body malignant melanoma); the tumor is of mixed cell type with foci showing Large and small Epithelioid cells with abundant eosinophilic cytoplasm, prominent nuclei, and inclusions. This tumor exhibits prominent tumor-infiltrating lymphocytes and macrophages. Abundant melanin pigmentation is seen](image-url)
Four weeks later, after full recovery from the previous operation, the patient underwent a right eye enucleation and orbital implant insertion. The pathology showed ciliary body melanoma, mixed cell type, pT4b with no extra-scleral extension with features including domed shaped, placed inferotemporal anteriorly (ciliary body) with a greatest basal diameter of 28 mm and 10 mm distance from the optic disc (Fig. 1). At 12 months follow-up, the patient was doing well with the excellent prosthetic eye, with no disease recurrence for either the eye or the kidney, and the whole-body CT scan showed no distant metastasis.

**DISCUSSION**

Despite being infrequent, the prevalence of multiple primary malignancies is increasingly widely identified because of advanced diagnostic equipment and the extended survival of cancer patients. The reasons for different multiple neoplasms are unclear, but several risk factors, including family history, genetic mutation, environmental factors, smoking, and radiation exposure, have been suggested to be involved. Multiple primary tumors are considered the presence of more than one synchronous or metachronous cancer in the same patient. The diagnostic criteria for multiple primary cancer remain the same criteria advocated by Warren and Gates in 1932 [13]. Namely:

- each cancer must be definitively malignant by histopathology;
- they must be histologically different;
- the possibility of metastasis among the cancers must be excluded.

In this case report, we described a synchronous occurrence of different pathological conditions (uveal melanoma and RCC), which were managed accordingly.

In the literature, two studies showed cases of clinical second primary neoplasms long with uveal melanoma had been reported between 2000 and 2020. Ten out of 333 patients (3.3%) had synchronous second cancers, and six out of 139 patients (4.3%) had second primary cancer (50%) were synchronous, and (50%) were metachronous [12]. In the present case, the 55-year-old middle-eastern patient with an olive-brown skin colour has a negative medical and family history. He has no history of chemotherapy or radiation exposure. The UM and RCC were asymptomatic and were diagnosed incidentally. And all lesions were resected successfully.

Uveal melanomas have an incidence of 6 per 1 000 000 per year in white adults and are more common in white than in non-white races, with a mean age of 60.14. The main treatment for UM depends on the size and location of the tumor. Large tumors or tumors close to the optic nerve usually require enucleation. Smaller tumors can be treated by local excision, local radiation, or proton beam irradiation [15]. Differential diagnoses of the patient involved hemorrhage in the subretinal or suprachoroidal space, choroidal hemangioma, solitary choroidal granuloma, and choroidal metastasis. The clinical and echographic features were typical for primary uveal melanoma, which was confirmed with histopathology after enucleation.

Renal cell carcinoma is the most common malignant tumor of the adult kidney and the most lethal of all malignancies. It is more common in males in the 6th and 7th decade and usually involves the upper pole. It has a satisfactory prognosis after surgical resection. Radical nephrectomy is the primary treatment for localized RCC [16]. There was no sign of renal problems in the patient’s medical history. Normal blood test results, urine analysis, and kidney function tests ruled out renal impairment. As
the case presented here is not in the advanced stage of UM and RCC, there were no complications or signs of organ impairment except right-eye vision. Important to remember that UM and RCC in their early phases can be missed, left untreated, and escalate to irreversible symptoms, particularly in developing countries. It is essential to establish whether these tumors are hereditary by interviewing patients with multiple primary neoplasms. This information might be helpful for doctors to assess cancer risk and optimize treatment. This case report has a limitation: we did not test for the specific mutation of the patient and his relatives as this test is unavailable. However, the systemic workup for this patient who presented with uveal melanoma could reveal another primary tumor, and this approach saved his life.

REFERENCES