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CASE REPORT

Metastatic melanoma after 23 years of primary ocular melanoma

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SUMMARY

We describe a case of 52-year-old man who presented with an episode of tonic-clonic seizures. He had right ocular melanoma 23 years ago with subsequent enucleation which was the standard treatment at that time. CT scans of the brain and of the thorax-abdomen-pelvis revealed widespread metastatic lesions in the brain, lung and liver. Further investigations including bronchoscopy with cytopathology uncovered that the metastatic disease was a recurrence of ocular melanoma. He received palliative radiotherapy and died 6 months later. Ocular melanoma is often associated with fulminant metastatic disease after a period of dormancy. Thus, despite successful treatment of the localised disease at initial presentation, an effort is needed for optimal long-term follow-up plan in order to improve survival in case of recurrence.

BACKGROUND

Ocular melanoma is the most common primary cancer of the eye. The incidence of ocular melanoma is six cases per million populations per year with about 70% of cases identified in the fifth through seventh decades.¹⁻⁴ Patients with ocular primaries have high incidence of distant metastases, and short subsequent survival.

CASE PRESENTATION

A 52-year-old Caucasian male was brought to the emergency department after an episode of seizures. It started with involuntary movement in the right hand lasting for 2-3 min, dizziness followed by episode of generalised tonic-clonic seizures witnessed by his wife. He reported of headache while recovering from the postictal phase.

He had a history of (1) enucleation of right eye 23 years ago for ocular melanoma and (2) depression. He was a non-smoker and worked in a rubber factory.

His vitals were stable on presentation.

INVESTIGATIONS

Routine laboratory investigations including full blood count, renal profile, liver function tests, coagulation profile and ECG were unremarkable. Chest radiograph revealed left hilar lymphadenopathy and nodules in the left lower lobe.

Initial brain CT scan was organised to rule out any possible space occupying lesion; it revealed two nodular ring-shaped lesions (each about 2 cm in diameter) situated in the left frontal and parietal lobe with ring-like uptake of contrast with adjacent

significant vasogenic oedema with no midline shift (figure 1). Appearances were suggestive of metastatic disease with primary lesion elsewhere in the body. Brain MRI with contrast confirmed the above findings.

He subsequently had a thorax-abdomen-pelvis CT scan with contrast which showed large left hilar lymphadenopathy with 3-4 soft tissue density nodules in the left lower lobe, the largest measuring ~4 cm (figure 2). Liver showed a 2 cm hypodense lesion in segment 8 (figure 3). Initial diagnosis was left lung primary malignancy with satellite lesion in ipsilateral lung and metastatic disease in brain and liver.

A bronchoscopy with transbronchial biopsy of the mediastinal lymph nodes was organised in a tertiary care centre. Melanoma markers including HMB45, MelA and S100 were positive which confirmed melanocytic lineage. However, BRAF V600 mutation was not detected.

DIFFERENTIAL DIAGNOSIS

1. Lung malignancy
2. Recurrence of ocular melanoma.

TREATMENT

The patient was informed of the above diagnosis and referred to medical oncologist and radiation oncologist for further care. He received palliative radiotherapy which was well tolerated by him.

OUTCOME AND FOLLOW-UP

However, about 6 months later patient died of metastatic disease.

DISCUSSION

Ocular melanoma is the second most common type of melanoma after cutaneous and most common primary malignancy of eye with 95% of melanomas occurring in uvea and remainder arising in conjunctiva. The incidence of ocular melanoma is six cases per million population per year with about 70% of cases identified in the fifth through seventh decades.²⁻⁴ In Europe uveal melanoma shows a decreasing incidence from north-to-south from over 8 per million in northern countries to <2 per million in southern countries.⁵ Risk factors like host pigmentation factor including light eye colour, fair skin, ultraviolet rays' exposure and atypical cutaneous or iris naevi have been implicated.⁶⁻⁸ The incidence of uveal melanoma has remained stable for the last three decades; however conjunctival melanoma has shown an increasing trend.⁹



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Figure 1 Brain CT scan pre and post contrast study showing ring enhancing lesions left frontal region with extensive perilesional oedema, consistent with metastatic lesions.

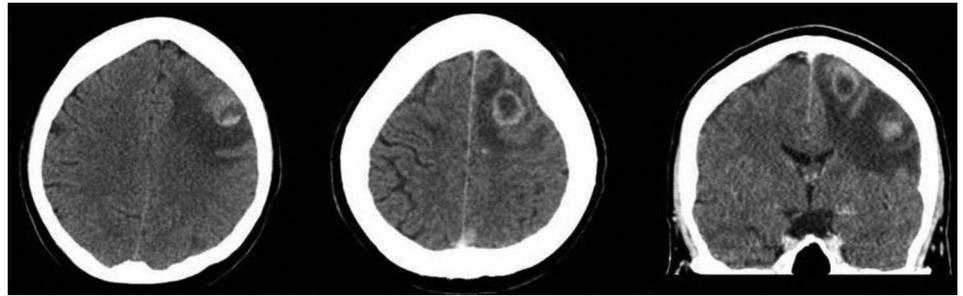


Figure 2 Thorax CT scan showing multiple nodules in the left lower lobe.



Figure 3 Liver CT scan showing 2 cm hypodense metastatic lesion in segment 8.

The most common symptoms of ocular melanoma are blurring of vision, irritation in eyes and photopsia. However, a fair number of patients may be asymptomatic and ocular melanoma may be an incidental finding during eye examination.

Enucleation was the standard treatment for ocular melanoma and used in most of the patients at that time. For most small and medium size tumours, radiation is the current recommended treatment, for instance plaque radiotherapy (aka

brachytherapy), proton beam radiotherapy and stereotactic radiotherapy. There are no studies showing that one form of radiation is better than the other form. Surgery is often recommended for tumours of larger size and for iris melanomas in particular. Also, surgery may be recommended for recurrent disease, after initial radiation treatment.¹⁰

It was suggested that manipulation of the globe and the consequent fluctuations in intraocular pressure may actually cause dissemination of tumour cells.¹¹ Late recurrence of melanoma is common; the 5-year and 10-year recurrence rates are 25% and 34%, respectively. In our patient the recurrence occurred more than two decades after the initial presentation. Hepatic metastases developed in 92% and in 55% of these, the liver was the only organ involved initially. Pulmonary parenchymal metastases developed in 31%. Bone involvement accounted for 23%, mostly affecting the spine. Again 17% had skin or subcutaneous metastases.¹²

The prognosis of metastatic melanoma is poor and currently there are no approved treatments for metastatic disease, though there are several palliative treatments, as well as new clinical trials offered in the USA and Europe. Liver directed treatment may be offered and systemic treatments using agents such as ipilimumab (Yervoy), may be given.¹⁰

Learning points

- ▶ Increasing cases of recurrence after prolonged period of quiescence has been reported with dim prognosis in most circumstances.
- ▶ Despite paucity of a proven effective treatment regime and therefore questionable benefit of surveillance, an optimal follow-up plan may still seem favourable.
- ▶ Therefore, more efforts towards follow-up care in terms of method of screening and time interval needs to be encouraged, which may seemingly improve patient outcome.

Contributors SRK was involved in patient care and writing and editing of the case report. PRB was involved in patient care, editing of case report, data collection and consent form. NR, consultant radiologist, was involved in all imaging studies for the patient and providing images for the purpose of case report. SA was involved in patient care and data collection.

Competing interests None declared.

Patient consent Obtained.

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