Metastatic disease from untreated choroidal and ciliary body melanomas

S.J.A. Rankin & P.B. Johnston
Department of Ophthalmology, The Queen's University of Belfast and Royal Victoria Hospital, Grosvenor Road, Belfast, BT12 6BA, Northern Ireland

Accepted 9 February 1990

Key words: metastases, melanoma, choroid

Abstract

We report the case histories of three patients with untreated malignant melanoma of the choroid or ciliary body. Metastatic disease which resulted in death was diagnosed four years in two patients and 13 years in one patient after the diagnosis of choroidal melanoma was suspected. The diagnosis of malignant melanoma was confirmed by histopathology in two of these patients.

Introduction

The natural history of uveal melanoma is poorly documented [1, 2] as most eyes containing a primary malignant melanoma undergo enucleation as the treatment of choice [2]. Zimmerman pointed out that enucleation is not part of the natural history of melanoma which is poorly understood [2]. Recently the value of enucleation was questioned because of an increase in tumour related mortality two to three years post-enucleation [3]. Zimmerman and McLean [4] believe that the peaking of tumour deaths is related to events occurring at the time of enucleation, which include release of neoplastic cells into the circulation and decreased host resistance to disseminated cells from removal of the primary tumour. This view is responsible for the current uncertainty towards enucleation of eyes containing choroidal melanoma and makes it important to report the behaviour of untreated choroidal melanoma. There are few reports of metastatic disease from untreated uveal melanoma [1, 5]. We present the case histories of three patients who died of metastatic disease from untreated uveal melanoma.

Case histories

Patient one was a 62-year-old man who presented with a blind proptotic right eye in 1979. He was first seen 13 years previously with a total retinal detachment in the right eye and an intraocular tumour was suspected. The patient was lost to follow-up until he presented with 8 mms proptosis of his right eye and a palpable mass in the superior orbit. The mediae were opaque so no direct view of the tumour was possible. A chest x-ray revealed a hilar mass. Liver function tests were normal. Over the course of two years he developed a large orbital mass with metastases to liver, lung and axilla (Fig. 3). The patient died and post mortem examination revealed an infiltrating malignant melanoma of the choroid composed of spindle cells.

Patient two was a 50-year-old woman who presented with a three year history of blurred vision. The right visual acuity was perception of light and there was total retinal detachment of this eye without a detectable retinal break. Ultrasound B-scan and fluorescein angiogram did not detect evidence of uveal melanoma. The patient was observed for 14 months by which time a choroidal melanoma
was clinically visible. At this time chest x-ray revealed three small localised lung opacities which were consistent with metastatic disease. Despite this, enucleation was performed to confirm the histological diagnosis. Liver function tests were normal. Histopathological examination of the enucleated eye revealed a 10 mm diameter mixed cell tumour of the ciliary body with scleral infiltration and extraocular extension (Fig. 1). The patient died 18 months post-enucleation with widespread metastatic disease principally involving the liver.

Patient three was a mentally handicapped 63-year-old man who presented in 1983 with a proptotic right eye. He was examined four years previously with rubeosis iridis of the right eye and hyphaema. There was a mature cataract but ultrasound examination showed an intraocular tumour which extended into the orbit. Isotope bone scan revealed multiple bone secondaries of the rib cage. Liver function tests showed a minimal rise in alkaline phosphatase. The patient underwent palliative loco-

Fig. 2. Case 3. Right orbital mass following initial presentation with rubeotic glaucoma.

Fig. 1. Case 1. Right orbital mass and large axillary metastasis.

cal radiotherapy but six months later he had an expanding fleshy mass of the right orbit (Fig. 2). A CT scan revealed extensive destruction of the orbital walls with erosion into the anterior cranial fossa. CT scan of liver showed hepatic metastases. The patient died two months later of bronchopneumonia. Histological diagnosis is unavailable.

Discussion

These patients represent one extreme of the natural history of malignant melanoma. It is estimated that the mortality before enucleation is 1% per year [3] and evidence of metastatic disease before or at the time of discovery of the intraocular tumour is rarely reported. Sobanski reported a series of 10 patients with uveal melanomas who refused enucleation, all died of metastatic disease within 10 years [7]. Various other large series of cases have shown either few or no cases of metastasis before enucleation [8].

Kidd et al. [9] reported a 15 year surgery of 100 cases of malignant melanoma of the choroid which were treated by enucleation at the Royal Victoria Hospital, Belfast. This gives an annual incidence of six to seven new cases per year for a population of 1.5 million. In a similar period our three patients are the only cases of choroidal melanoma which progressed to widespread metastatic disease before recognition or treatment of the primary tumour.