An intracranial lesion discovered in a patient with recurrent conjunctival melanoma: a case report

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Date of submission: 30/06/2013 Date of approval: 27/09/2013

ABSTRACT

Conjunctival melanoma is a rare tumor of the orbit that represents only 1.6% of all non-cutaneous melanomas, with an annual incidence of less than 5 cases per 1 million. Overall metastasis rates range from 14-27%, with 10-40% occurring within the central nervous system (CNS). CNS involvement has shown to be the major cause of morbidity and mortality in these patients, with a median survival of between 2 and 10 months. Therefore, it is crucial to follow these patients closely throughout their lifetime to monitor for disease recurrence. We present a unique case of a 65 year-old woman with a well-documented history of recurrent conjunctival melanoma after multiple excisions. Routine follow up imaging revealed a new intracranial lesion, which presented an interesting diagnostic challenge.

Key words: Conjunctival tumor; melanoma; metastasis

INTRODUCTION

Conjunctival melanoma is a rare tumor of the orbit that represents only 1.6% of all non-cutaneous melanomas, with an annual incidence of less than 5 cases per 1 million. About 14-27% of conjunctival melanomas metastasize, approximately 30% of which proving fatal within 10 years of diagnosis. Generally accepted management of conjunctival melanomas entails surgical excision with subsequent cryotherapy to the resected margins. Because of the relatively high risk of metastasis of these tumors, it is crucial to follow these patients closely after excision with a biannual systemic workup complete with abdominal and chest computed tomography (CT), brain magnetic resonance imaging (MRI), liver function tests, and thorough physical exam. We present an interesting case of a patient with conjunctival melanoma who was found to have a left parietal tumor on routine follow up imaging.

CASE REPORT

A 65 year-old woman with a history of recurrent right conjunctival melanoma presented with new onset of diplopia, pain, photosensitivity, and foreign body sensation in the right eye globe. On physical examination, an ill-defined fleshy non-pigmented mass was noted in the right infero-temporal fornix causing limited abduction of the right eye globe. CT orbital imaging showed an ill-defined soft tissue mass over the left parietal area.
Conjunctival melanomas are uncommon tumors with a metastasis incidence of approximately 14-17%. Increased size and depth of melanomas carry a worse prognosis with tumors thicker than 1.4mm carrying a higher risk of metastasis and death.3 Some studies have concluded that the location of the melanoma carries prognostic implications with bulbar conjunctival tumors having the best prognosis and caruncular melanoma associated with a worse prognosis.3 Melanomas are the third most common source of intracranial metastases following breast and lung carcinomas.2 In general, melanoma metastasis to the central nervous system (CNS) ranges from 10-40% in clinical series, with as high as 66% of patients with CNS metastatic melanoma tumors discovered during autopsy series. CNS involvement has been proven to be the major cause of morbidity

**DISCUSSION**

Conjunctival melanomas are uncommon tumors with a metastasis incidence of approximately 14-17%. Increased size and depth of melanomas carry a worse prognosis with tumors thicker than 1.4mm carrying a higher risk of metastasis and death.3 Some studies have concluded that the location of the melanoma carries prognostic implications with bulbar conjunctival tumors having the best prognosis and caruncular melanoma associated with a worse prognosis.3 Melanomas are the third most common source of intracranial metastases following breast and lung carcinomas.2 In general, melanoma metastasis to the central nervous system (CNS) ranges from 10-40% in clinical series, with as high as 66% of patients with CNS metastatic melanoma tumors discovered during autopsy series. CNS involvement has been proven to be the major cause of morbidity
and mortality in these patients, causing death in up to 95% of patients, with a median survival between 2 and 10 months. Therefore, it is essential to follow these patients closely with a complete systemic workup including abdominal and chest CT imaging, brain MR imaging, liver function tests and a thorough physical exam. Our patient presented an interesting diagnostic challenge, highlighting the importance of acquiring a thorough clinical history to arrive at the accurate diagnosis prior to making management decisions. Routine follow up imaging of our patient illustrated a left parietal lobe lesion highly suspicious for metastasis given the patient’s extended history of repeated orbital melanoma recurrences after multiple excisions. However, meticulous search for prior imaging revealed a brain CT one year earlier that demonstrated the lesion to be unchanged in character and size when compared to the most recent imaging, with features consistent with a meningioma. Thus, diagnosis of a metastatic process was excluded. Also endorsing this diagnosis, intracranial metastasis frequently involves multiple rather than solitary lesions. Dural or extra-axial metastases are found in nearly 10% of patients with advanced cancer. In some cases, only dural metastases are present and they may mimic meningioma, hemangiopericytoma or a solitary fibrous tumor. In men, dural metastases are most likely from prostate cancer; whereas, in women, most dural metastases arise from breast cancer. While dural metastasis may present years after the primary tumor, they are most often seen in the context of known extracranial cancer. Meningiomas encompass a broad group of extra-axial intracranial neoplasms that arise from arachnoidal cells of the meninges, including: Meningiomatous neoplasms or meningiomas, non-meningiomatous mesenchymal neoplasms, both benign or sarcomatosis, melanocytic neoplasms, glial tumors of leptomeninges and tumors of hematopoietic origin. On CT, meningiomas are generally hyper-dense and enhance uniformly. However, histological variances can exist with different imaging appearances due to lipomatosis metaplasia with fatty density and psammomatous calcifications. Some meningiomas may become densely calcified.

On MR, the typical imaging features consist of a rounded and well-circumscribed extra-axial dural-based mass. These lesions may show homogeneous or irregular enhancement, frequently with adjacent dural thickening and enhancement described as “dural tail”. This dural tail has been found to be reactive in most cases and may not represent an extension of the neoplasm. This finding is suggestive, but not specific for meningioma, as can occasionally be observed in other dural-based tumors as well. The most common locations for meningiomas include: the convexities, falx cerebri, as well other locations at the base of the skull and orbits. If any of the less frequent histological variances are present, the MR signal characteristics may vary due to water, fat, or bone content. A psammomatous calcification can be present and will be hypo-intense on MR. Prominent vascularity may indicate a benign angiomatous meningioma. Morphologic variations in meningiomas may range from a well-rounded well-circumscribed mass to a flattened or en plaque configuration. Associated edema may be present in various degrees. In contrast, intracranial melanoma demonstrates a variety of patterns on MR imaging. The most common manifestations illustrate melanotic and amelanotic patterns. The melanotic pattern consists of high signal intensity on T1-weighted images and low signal intensity on T2-weighted images. Amelanotic lesions may appear hypo-intense or iso-intense to the cortex on T1-weighted images and hyper-intense on T2-weighted images. However, they often differ by these trends. Ultimately, when discovering a new intracranial lesion in a patient with a well-known cancer history, it may seem easy to presumptively diagnose the new tumor as metastasis. Our case stress the importance of carefully exploring the patient’s physical exam and history, reviewing all prior imaging, even if from an outside facility. Additionally, we demonstrate the value of consistent collaboration between the ophthalmology and radiology teams to arrive at the correct diagnosis and the best management strategy plan for the patient.