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CASE REPORT

The black lacrimal sac: a clinicopathological correlation of a malignant melanoma with anterior lacrimal crest infiltration

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Abstract Primary lacrimal sac melanoma is an extremely rare clinical entity with fewer than 25 cases reported in the literature. We present a well-documented case of lacrimal sac melanoma with anterior lacrimal crest infiltration and discuss the clinical, radiological and histopathological features along with surgical challenges and treatment.

Keywords Lacrimal sac · Malignant melanoma · CT scan orbits

Introduction

Malignancy of the lacrimal sac is rare, and primary malignant melanoma in this region is extremely rare [1–4]. The incidence of melanoma among lacrimal sac tumors varies between 4 and 13 % in different studies [1–4]. The average age at diagnosis is 59 years with men diagnosed at an older age as compared to women [4]. To the best of our knowledge, fewer than 25 cases of primary lacrimal sac melanoma have been reported to date [1–12]. Malignant melanomas are highly invasive with involvement of the nasolacrimal ducts and in some cases the medial orbit [1–6]. Although medial orbital wall involvement has been reported in an advance case of undifferentiated carcinoma, bony involvement in malignant melanomas has fewer reports in the literature [2, 6, 9–13], and these cases are summarized in Table 1. In this clinicopathological correlation, we discuss the presentation of primary malignant melanoma of the lacrimal sac with bony infiltration involving the anterior lacrimal crest, and discuss the surgical challenges with bony involvement, histopathological features and management.

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Case report

A 53-year-old female presented to us with a 1-year history of insidious onset of a slowly progressive, non-tender mass in the left medial canthal area associated with watering. There was a history of recent incision biopsy of the growth 4 months earlier, which was reported to be a malignant melanoma with skeletal muscle infiltration.

On presentation to us, her best-corrected visual acuity was 20/20 in both eyes. Ocular examination revealed a scar of the previous incision biopsy without any cutaneous ulceration (Fig. 1a). The mass was found to be extending just above the medial canthal tendon (Fig. 1a). Syringing showed regurgitation of

Table 1 Reports of malignant melanoma with bony involvement and their treatment

Patient no.	References	Treatment modality	Comments
1	Offret and Haye [10]	Tumor excision with wide bony margins	Surgery included radical sinus and nasal surgery
2	Farkas and Lamberson [11]	Tumor excision with medial orbital resection	Medial orbital wall involvement
3	Glaros et al. [12]	Tumor excision along with bony nasolacrimal duct removal	Patient had widening of the bony nasolacrimal duct
4	Owens et al. [13]	Wide en-block excision along with medial orbital wall resection	Additional medial maxillectomy and ethmoidectomy performed
5	Eide et al. [9]	Wide but incomplete excision along with EBRT	Performed on a recurrence
6	Gleizal et al. [2]	Wide en-block excision of nasolacrimal duct along with surrounding bone	Pre- and postoperative EBRT
7	Lee et al. [6]	Wide en-block excision along with medial orbital wall resection	Additional medial maxillectomy and EBRT performed

blood-stained fluid. There were numerous moles in the head and neck region; however, there was an absence of suspicious lesions (Fig. 1b). The left submandibular lymph node was palpable. Endoscopic nasal examination was unremarkable. At this stage, the plan was to perform imaging, fine-needle aspiration cytology of the left submandibular node and to review the past histopathology slides to confirm the diagnosis.

On imaging, a computed tomography (CT) scan revealed a fairly well-defined isodense mass in the lacrimal sac area (Fig. 1c). Bone windows showed another lesion over the anterior lacrimal crests with early infiltration changes without any nasal or orbital extension (Fig. 1d). Fine-needle aspiration cytology of the enlarged submandibular lymph node did not reveal the presence of any malignant cells. A review of the slides from the previous biopsy confirmed the diagnosis of melanoma. A systemic work-up for metastasis was negative. A decision was taken to perform dacryocystectomy (DCT) followed by focal external beam radiotherapy (EBRT).

A wide-margin DCT with en-bloc excision was performed (Fig. 1e), since the past incision biopsy from the fundus of the sac showed a focal area of skeletal muscle infiltration. The tissue around the previous scar mark from the incision biopsy was excised. A 4-mm margin superiorly, up to the periorbita laterally without breaching it, overlying bone medially (Figs. 1f, 2a, b) and the nasolacrimal duct (Fig. 2c) were included in the wide excision. The overlying anterior lacrimal crest showed a 4 × 3 mm extension of the melanoma (Fig. 1f). A 3-mm surgical margin was marked all around the bony infiltrate and an electric saw was used to gently remove the bony block (Figs. 1f, 2a, b). The nasal endoscopic examination was normal (Fig. 2d).

Gross examination of the lacrimal sac (Fig. 1e) and bone specimen (Fig. 2a) showed an irregular pigmented lesion with a solid blackish cut surface. Microscopic examination of the lacrimal sac revealed denuded stratified columnar lining with few goblet cells. Both specimens showed a cellular and pigmented tumor composed of sheets of epithelioid and spindle cells with a high nucleocytoplasmic ratio (Fig. 2e, f). The nuclei displayed marked atypia with mitoses (average of 6 per 10 high power fields). The majority of the tumor cells contained granular intracytoplasmic melanin (Fig. 2e), which was not seen after permanganate bleach, confirming the melanocytic nature of the tumor (Fig. 2g). Surgical margins were tumor free. Sections from the bony tissue showed tumor infiltrating the marrow spaces, replacing the marrow elements (Fig. 2f). Immunohistochemical staining with HMB-45 was positive in the tumor cells (Fig. 2h). The histopathological features suggested a diagnosis of a mixed malignant melanoma of the lacrimal sac and overlying anterior lacrimal crest.

Following the wide surgical excision, the patient underwent adjuvant external beam radiotherapy. At last follow-up (6 months), a systemic work-up was negative and there was no evidence of local recurrence. The patient is being closely followed up.

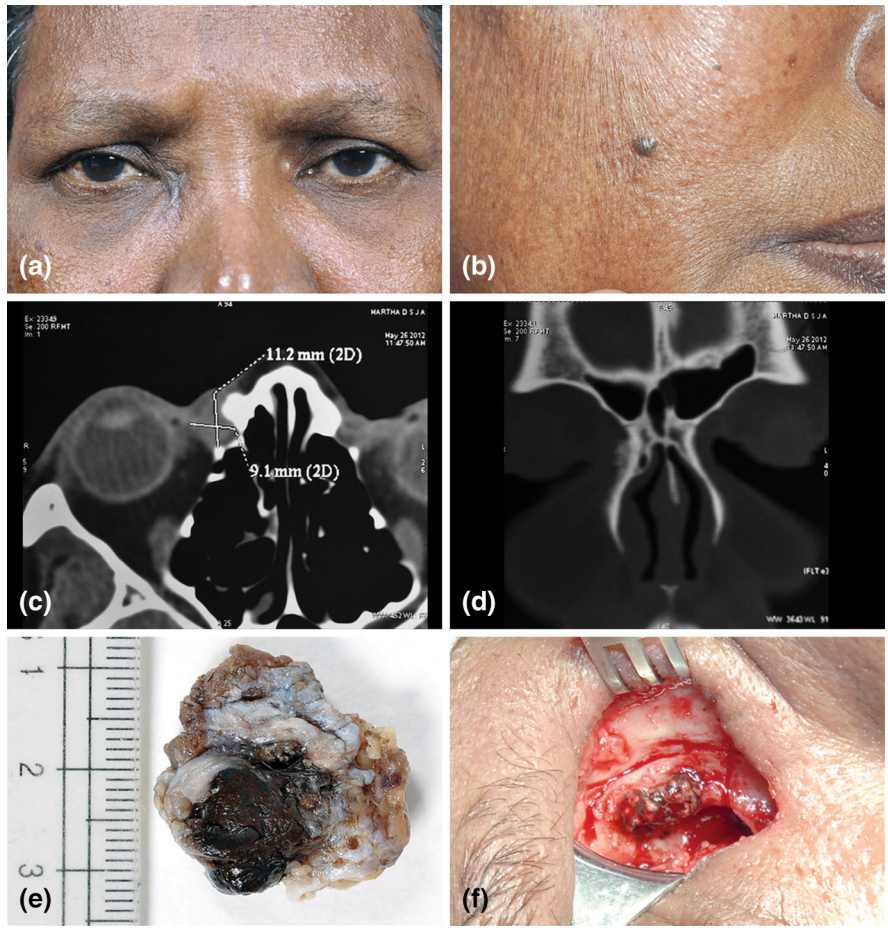

Fig. 1 External clinical photograph showing the past scar over the lacrimal sac area and fullness over and above the medial canthal tendon (**a**). External clinical photograph of the right cheek showing numerous moles (**b**). CT scan orbits (axial cut) showing enlargement of the lacrimal sac with an isodense lesion (**c**). CT scan orbits (coronal cut with a bone window) showing an irregular lesion over the anterior lacrimal crest with superficial infiltration (**d**). Lacrimal sac with the *black* tumor and wide surgical margins following en-bloc excision (**e**). Intraoperative photograph showing a *blackish mass* over the anterior lacrimal crest. Note the wide surgical margins taken around the lesion (**f**)

Discussion

The majority of lacrimal drainage system tumors arise in the lacrimal sac [1]. Although its origin is disputed, the tumor possibly arises from melanocytes located in the epithelium of the lacrimal drainage system or the underlying stroma [1–6]. The development of lacrimal sac melanoma has been related to multiple risk factors including older age, presence of dysplastic moles or nevi, delayed presentation, past history of surgery or interventions like incision biopsy, family history of melanoma and other sites with cutaneous melanoma [7]. In our patient, the first four of these six risk factors were present.

It is difficult to distinguish clinically from the more common chronic dacryocystitis. Diagnosis delay is mostly because of the overlap in the presenting symptoms of chronic watering, insidious onset and limited early visibility of the lacrimal sac [1–8]. Typically, patients have the clinical triad of epiphora, blood-stained reflux from the lacrimal punctum, and a mass in the region of lacrimal sac, partly above the medial canthal tendon [1–9]. The patient in this study had all these features. The usual sequence of events is epiphora, followed by recurrent bouts of dacryocystitis, a mass in the medial canthal area, and finally, epistaxis and involvement of the regional lymph nodes. Imaging modalities like the dacryocystogram and CT scan are of useful adjunctive value in aiding the diagnosis [1–4].

Malignant melanoma of mucosal tissue is far more aggressive and lethal than cutaneous melanoma [1–4, 14]. However, the rarity of this tumor and the lack of sufficient follow-up data prevents reliable prognosis. A low index of suspicion, insidious onset, delay in diagnosis along with factors that help the tumor to spread like regional vascularity and nasolacrimal route further increases mortality [1–4, 14]. Hoyt et al. reviewed 15 patients with mucosal melanomas of the

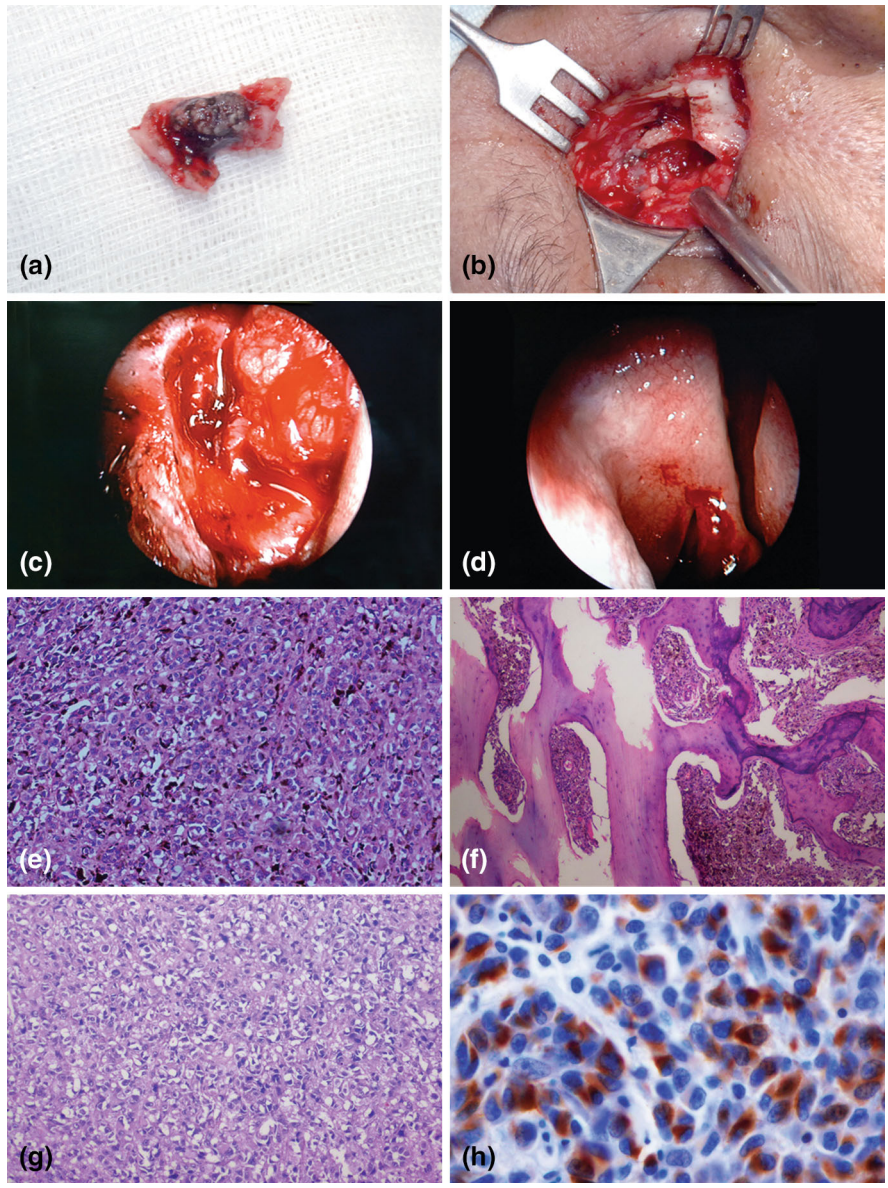

Fig. 2 The bony lesion following its removal (**a**). Intraoperative view of the field following wide surgical excision (**b**). Endoscopic view of the nasolacrimal duct showing no residual tumor in the vicinity (**c**). Endoscopic view of the middle meatus showing a normal nasal examination (**d**). Microphotograph showing sheets of tumor cells with intracytoplasmic melanin pigment (H&E, ×100) (**e**). Microphotograph of the bone lesion showing infiltration of marrow spaces by the pigmented tumor cells, replacing the marrow elements (H&E, ×40) (**f**). Intracytoplasmic pigment is not seen after permanganate bleach confirming the melanocytic nature of the pigment (H&E, ×100) (**g**). Immunohistochemical staining with HMB-45 showing positive staining of the tumor cells (**h**)

head and neck and found a median survival of 1.8 years, with a 5-year survival of only 10 %. They noted a local recurrence in 80 % of patients after a median time of 10 months [14]. Gleizal et al. reviewed 22 cases of lacrimal sac tumors and found that 73 % of the cases recurred. However, the follow-up of those who did not recur was short and variable, thus hinting that the recurrences could still be higher [2]. Therefore, a thorough evaluation for recurrence and metastasis is warranted.

Unfortunately due to the lack of experience with lacrimal sac melanomas, there are no standard treatment guidelines. Wide surgical excision with tumor-free margins is the preferred treatment [1–4, 7–14]. Radiotherapy, chemotherapy or immunomodulatory agents like alpha-interferons have all been described as an adjuvant modality of management, but with questionable efficacy [1–14]. Although the patient in the current study underwent a wide surgical resection including the surrounding bones, the patient was subjected to adjuvant radiotherapy along with close follow-up to monitor recurrence and metastasis.

In conclusion, although rare, lacrimal sac melanoma may masquerade as chronic dacryocystitis. Early

recognition and treatment appears to be an important prognostic factor and is likely to reduce mortality and improve survival of these aggressive tumors.

Disclosure We have full control of all primary data and agree to allow International ophthalmology to review the data on request.

Conflict of interest The authors indicate no financial conflict of interest.

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