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Primary malignant melanoma of the lacrimal sac: A case report

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Malignant melanoma of the lacrimal sac is very rare and primary malignant melanoma is extremely rare. It is usually diagnosed at an advanced stage after excision or biopsy of a tumor. We treated a patient with tearing and bloody discharge from the left eye. We performed a dacryocystectomy with the suspicion of a chronic dacryocystitis. However, the pathological findings and the immunohistochemical studies showed a malignant melanoma of the lacrimal sac. The patient underwent postoperative irradiation therapy. Follow up two months after surgery revealed no evidence of recurrence.

Early diagnosis is very important for prognosis in patients with malignant melanoma of the lacrimal sac. Because this tumor often presents with symptoms similar to dacryocystitis and may masquerade as a chronic dacryocystitis, it can be difficult to make an early diagnosis.

Key Words : Malignant melanoma, Lacrimal apparatus, Dacryocystitis

INTRODUCTION

Tumors of the lacrimal sac are rare. The most common neoplasms of the lacrimal sac are benign and most are of epithelial origin[1, 2]. Malignant melanoma of the lacrimal sac is a rare condition accounting for 5% of lacrimal sac tumours[3]. Malignant melanoma may develop in other parts of the body in addition to skin, but this is a very rare finding. Only 21 cases of primary malignant melanoma of the lacrimal sac have been reported worldwide. However, early identification of these cases is important because many are potentially lethal if therapy is delayed or inadequate[4]. This malignancy usually has an insidious onset and is often diagnosed at advanced stages after an incorrect diagnosis of dacryocystitis. This is due to the similar initial symptoms of dacryocystitis, benign tumor and malignant melanoma at this location.

CASE REPORT

A 53-year-old woman was referred to the department of ophthalmology with a palpable mass at the left medial canthal region. She complained of a six month history of tearing and a two month history of occasional bloody discharge from the left eye as well as left medial canthal region swelling. The swelling was not associated with pain or visual disturbance. The patient had no previous history of regional surgery or trauma or excessive sun exposure. There was no history of a cutaneous primary melanoma.

The physical examination revealed a cystic, localized, non tender palpable mass at the left medial canthus consistent with an enlargement of the lacrimal sac. Computed tomography scanning of the paranasal sinus showed a soft tissue mass in the left lacrimal sac, attached to the medial orbital wall (Figure 1). However, neither orbital cavity involvement nor bony destruction was detected. In addition, infiltration of the

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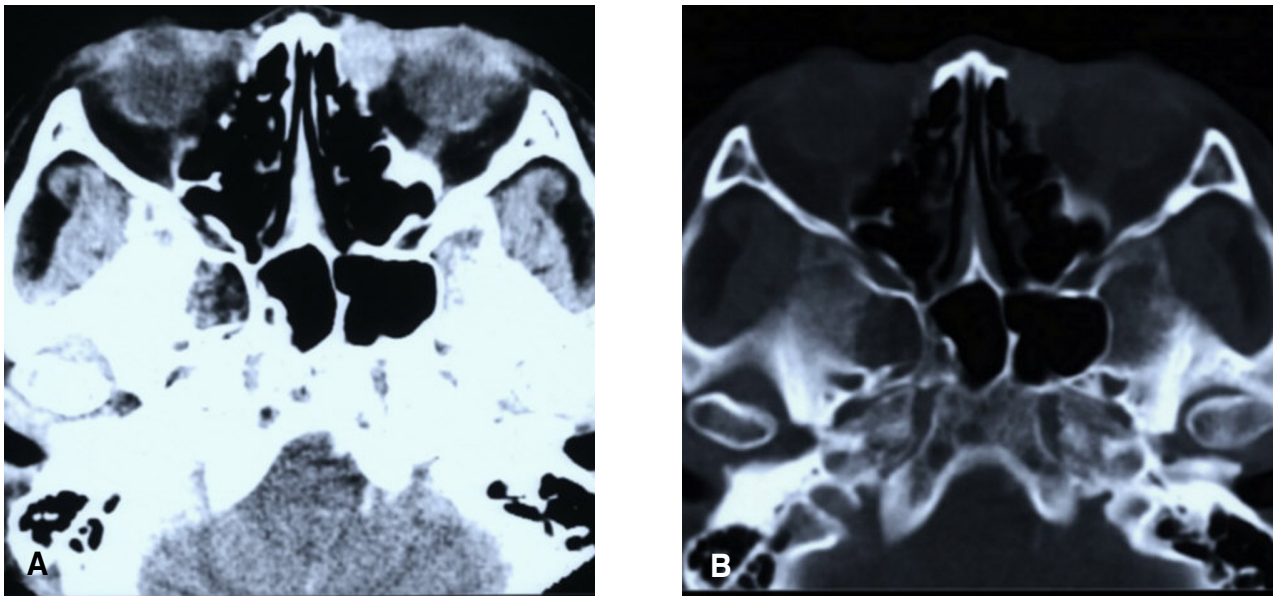

Figure 1. CT findings of melanoma of the left lacrimal sac. A) Contrast enhanced axial CT scans show a well enhancing soft tissue mass in the left nasolacrimal fossa. B) There was no evidence of destruction of adjacent bony structures.

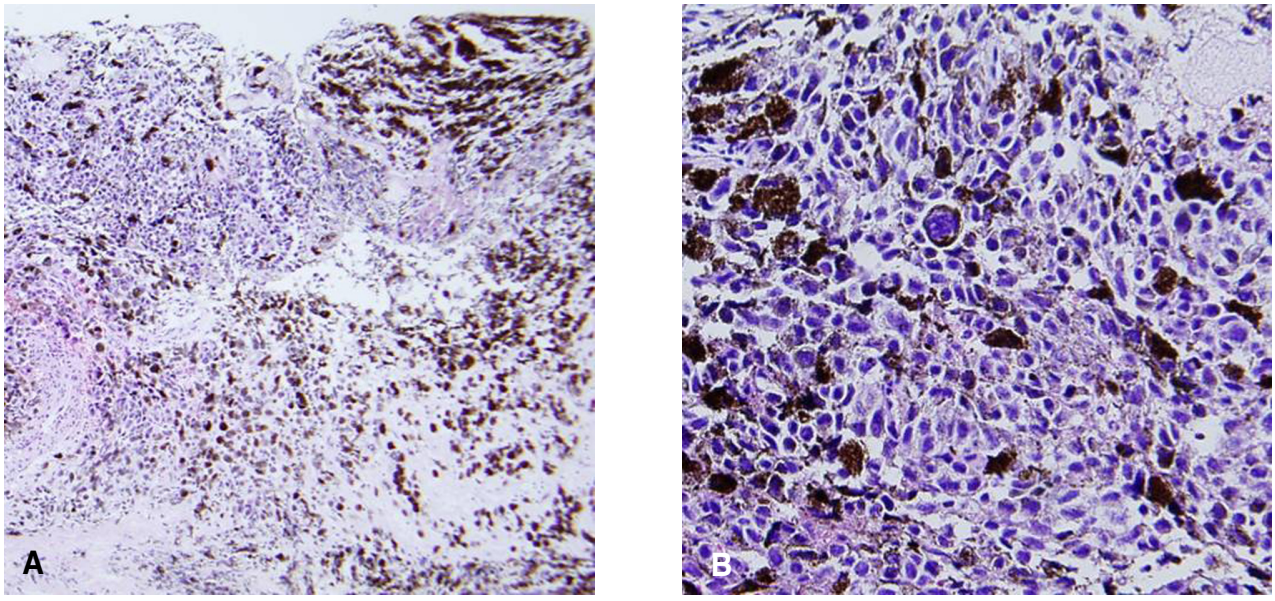

Figure 2. (A) Photomicrography of the lacrimal sac melanoma showing the tumor cells producing melanin pigmentation and infiltrating adjacent stroma. (H&E, ×100) (B) Photomicrography of the lacrimal sac melanoma showing solid growth of malignant cells characterized by production of melanin pigments, hyperchromic nuclei and prominent nucleoli (H&E, ×400).

surrounding soft tissue was not detected, and the mass did not extend into the nasolacrimal duct. There was no associated cervical lymphadenopathy. The patient was taken to the operating room for a scheduled dacryocystectomy. A brown-pigmented soft tissue containing cystic mass filled the lacrimal sac and did not protrude through the nasolacrimal duct. The mass was removed from the nasal and lacrimal bone and dissected from the medial canthal tendon. The cystic mass was thought to be benign and therefore only a dacryocystectomy was performed.

Biopsy and histological examination of the surgical specimen showed solid growth of malignant cells characterized by production of melanin pigments and hyperchromatic nuclei with prominent nucleoli (Figure 2). The tumor cells showed strong

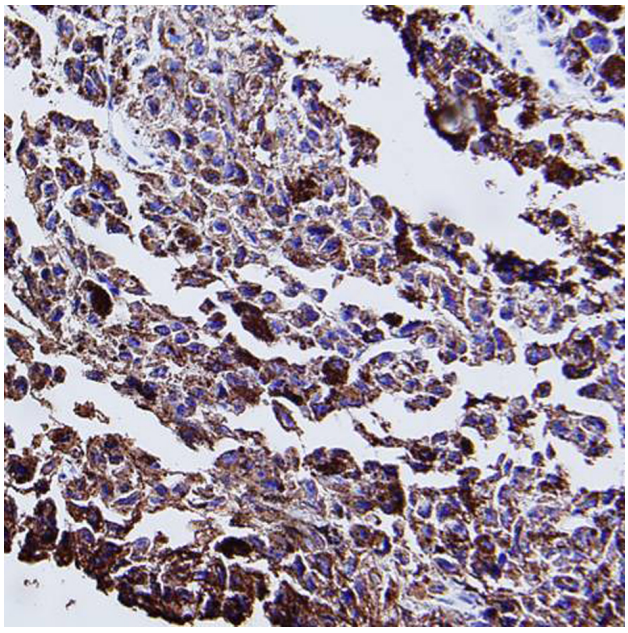

Figure 3. These tumor cells show strong immunoreactivity for anti-melanoma monoclonal antibody, HBM-45 (immunohistochemical staining, ×400).

immunoreactivity to melanoma specific antibody, HBM-45 in the immunohistochemical examination, consistent with a diagnosis of malignant melanoma (Figure 3); whereas the immunoreactivity to the S 100 protein was negative. This histological examination confirmed the diagnosis of malignant melanoma and the resection margins were not free of tumor. A complete metastatic workup including computed tomography scanning of the head and neck, chest, abdomen and pelvis failed to demonstrate lymphadenopathy or other evidence of metastasis. A general physical examination and routine laboratory tests were all within normal limits. The patient underwent 6000 rad of adjuvant irradiation therapy at the left lacrimal region for four weeks after surgery. During the two months follow up after surgery and radiation therapy, the patient had no evidence of local recurrence; however, since then she has been lost to further follow up.

DISCUSSION

Malignant melanoma is a malignancy of the pigment-producing cells known as melanocytes; the organ usually affected is the skin. While melanoma accounts for roughly 4% of all skin cancers, it is responsible for more than 77% of skin cancer deaths. Mucosal melanoma is a rare entity compared to cutaneous malignant melanoma. Mucosal malignant melanoma of the head and neck represents approximately 8% of all melanomas that develop in the head and neck, and generally behaves far more aggressively than cutaneous melanoma[5], patients with this malignancy generally have a poor prognosis despite aggressive treatment[6]. The development of lacrimal sac melanoma has been related to multiple risk factors including: presence of dysplastic moles or nevi, a family history of melanoma, other sites with cutaneous melanoma and an older age. The histogenesis of primary malignant melanoma of the lacrimal sac is not clear; melanocytes are not normally found in the tissue of the lacrimal duct system. Melanocytes originate in the neural crest during embryologic development[7]. Therefore embryologic factors may be important to the explanation of the etiology of this malignancy.

Melanomas arising in the lacrimal sac, similar to other mucosal melanomas, have an insidious onset and limited early visibility that generally result in delayed diagnosis and add to the poor prognosis[8]. In only six of 21 reported cases, of primary malignant melanoma of the lacrimal sac, were there no recurrences. An insidious onset of epiphora is characteristic of chronic inflammation or infection of the lacrimal sac and therefore it is easy to mistake malignant melanoma for chronic dacryocystitis. In most survivors, the diagnosis was made early; this appears to be the most important prognostic factor. Therefore, a computed tomography scan in patients with symptoms of chronic dacryocystitis is an important part of the evaluation[9].

The treatment varies with size and extent of the tumor, and may range from only dacryocystectomy to wide en-bloc resection, post-operative radiotherapy, chemotherapy, and immunotherapy in a variety of combinations[10]. Local recurrence of mucosal malignant melanoma of the head and neck region has been a major factor in treatment failure. Therefore, it has been recently recommended that the resection include a wide field en bloc excision of the whole lacrimal system and the surrounding tissues followed by irradiation. However, the role of adjuvant therapy such as radiation, chemotherapy and immunotherapy remain controversial. In this case, the patient presented with a palpable mass at the left medial canthal region with bloody discharge, and was diagnosed with malignant melanoma after a dacryocystectomy. The resection margins were not free of tumor, and therefore postoperative radiotherapy was added to the treatment regimen. Considering the natural course and the short follow-up, further evaluation with longer follow-up is needed to determine whether recurrence occurs.

In summary, early diagnosis is critical for a favorable prognosis, therefore consideration of the possibility of a malignant melanoma of the lacrimal sac is important when considering evaluation and work up for dacryocystitis. Further study with a larger patient population and longer follow up is required to determine patient outcome after treatment.

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