Case report

Malignant transformation of kissing nevus- a rare entity

Kharel (Sitaula) R, Bhatta S, Shrestha GB, Shrestha JK
Department of Ophthalmology, BP Koirala Lions Centre for Ophthalmic Studies, Institute of Medicine, Tribhuvan University Teaching Hospital, Maharajgunj, Kathmandu, Nepal.

Abstract

Background: Kissing nevus is a congenital nevus in adjacent parts of the eyelids. Malignant transformation of kissing or divided nevi of the eyelids is rarely described. **Objective**: To report a very rare case of malignant transformation of kissing nevus with ocular and extraocular spread.

Case: A 57- year- old man with 6/6 visual acuity in both eyes presented with a kissing nevus present since birth in right upper and lower eyelids which had a slow growth phase. The upper lid in the area of the nevus was thickened with a 20x12x15 mm black pigmented crusted hemorrhagic nodular lesions. The lower lid had a 6 mm black pigmented ulcerated lesion over the pre-existing nevus in the lateral third of the lid with full thickness infiltration. Another 5x4 mm pigmented lesion over the lower medial lid margin with a thickness of about 3 mm extended to the conjunctival side of the lower lid. Right sided pre-auricular and sub-maxillary nodes were palpable. A biopsy of tissue samples from the eyelid and pre-auricular nodes were consistent with malignant melanoma.

Conclusion: Malignant transformation of kissing nevus is rare. It can spread to the conjunctiva, pre-auricular and sub-mandibular lymphnodes.

Key-words: lymph node; kissing nevus ; malignant melanoma; metastasis

Introduction

Divided nevus or kissing nevus is a rare form of congenital nevus that usually occurs on contiguous portions of the upper and lower eyelids of one eye and may cause functional and aesthetic problems (Font RL et al 1996). It was first described in 1919 by Fuchs and is thought to arise during fetal eyelid fusion. The growth arises from melanoblasts or Schwann cells of a neuroectodermic origin during embryologic development (Carmine et al, 2007). When the eyelids are closed the eye appears to be covered by one large nevus. The conjunctiva is rarely affected, whereas the cilia are larger than normal. Though often present from birth, it may also develop later. It shows slow growth, and very rarely, malignant transformation to malignant melanoma (McDonnell et al, 1988).

Malignant melanoma (MM) constitutes approximately 1% of all malignant neoplasms of the eyelid skin (Font et al 1996) and less than 1% of all skin melanomas (Rodriguez - Sains RS et al 1981). Although relatively rare, they are the leading cause of death from primary skin tumors. Eyelid melanomas with conjunctival involvement have been shown to behave in a more aggressive manner than those conûned to the eyelid skin.

Received on: 20.12.2012 Accepted on: 04.03.2012
Address for correspondence: Dr R Kharel (Sitaula), Department of Ophthalmology, BP Koirala Lions Centre for Ophthalmic Studies, Institute of Medicine, Tribhuvan University Teaching Hospital, Maharajgunj, Kathmandu, Nepal.
E-mail:helloranju50@yahoo.com

Recently, inherited mutations in the melanocortin-1receptor gene have been associated with people with red hair, photosensitivity and to an increased risk of cutaneous MM (Tsao et al 2003).

Cutaneous MM can be classiûed into four groups according to the following clinical and histopathologic criteria: (1)nodular melanoma, (2) superûcial spreading melanoma, (3) lentigo maligna melanoma, and (4) acral lentiginous melanoma.

The American Joint Committee on Cancer (AJCC) has recently revised the staging system for cutaneous melanoma to aid in clinical management (Balch et al, 2001). In stages I and II, the thickness of the melanoma and presence or absence of ulceration are used as the basis for staging because of their effect on prognosis. Stage III patients have regional lymph node or in-transit disease, and stage IV patients have distant metastases. Pathologic information about lymph node involvement obtained from lymphatic mapping and sentinel lymphadenectomy is also included.

Kissing nevi of the lid may affect visual development if the increased bulk of the upper lid causes a mechanical ptosis and occlusion of the visual axis, particularly in childhood and adolescence. Because of severe disfigurement, risk of later malignant change in the lesion and the possibility of deprivation amblyopia, early surgical treatment is recommended for all medium and large congenital melanocytic naevi of the eyelid (Papadopoulos et al, 1991; De Pietro et al, 1981). Early reconstructive surgery preferred in order to achieve the best aesthetic result. If detected relative early, however, it may be treated with local resection, possibly combined with brachytherapy, using a custom-designed orbital plaque, or external beam irradiation. Patients with extensive orbital disease should undergo orbital exenteration with the aim of eliminating the potential for metastasis and improving survival.

Case report

A 57- year-old male presented with complaints of burning sensation in both eyes. His unaided vision in both eyes was 6/6. He had an 11x4 mm large kissing nevus in adjacent parts of right upper and lower eyelids involving the margins affecting the middle and outer part of the eyelids. The pigmented lesion was larger, thicker, and darker in the upper eyelid. The closed eyelid clearly showed a pathognomonic kissing nevus. On enquiry he said that these lesions had been present since birth. The nevus was very slowly increasing in size but he had not consulted a doctor due to his poor socioeconomic background. He sought treatment at our center because one month prior, he had developed ulceration in the nevi of the right upper lid associated with occasional bleeding from the lesion. Conjunctiva, sclera and cornea were exempt from melanocytic pigmentation. The left eye was completely unremarkable and fundus was normal. No lymph nodes were palpable. Family history for melanoma was negative. The growth of the nevi associated with ulceration and bleeding, led us to advise the patient for excisional biopsy with reconstruction of eyelids and admission was planned. The patient was scheduled to follow up the next day for admission but he did not return for further investigations and procedures.

The patient presented two months later with the complaints of rapid growth of the lesion associated with marked painless diminuation of vision in his right eye. He had had rapid growth of the eyelid lesions after the last visit but summoned faith healers for the treatment. When there was no improvement he returned to us for further treatment.

His visual acuity was perception of light with accurate projection of rays in the right eye and 6/6 in the left. Inspection of right eyelids revealed an ill defined thickening of the upper lid in the area of the nevus with a black pigmented crusted hemorrhagic nodular lesion involving nearly the entire conjunctival surface protruding from the palpebral fissure. On palpation, the nodular lesion was about 20mm x 12mm in the horizontal and vertical dimensions with the a thickness of about 15mm. The right lower lid had a 6 mm black pigmented ulcerated lesion over the pre-existing nevus in the lateral third of the lid along the lid margin. Full thickness of the lid was

involved. Contiguous to it were multiple, small, ill-defined pigmented nodular lesions in the lid margin. There was another pigmented lesion over the lower medial lid margin about 5mm x 4mm in size with a thickness of about 3 mm extending to the conjunctival side of the lower lid. There was also diffuse involvement of the bulbar and palpebral conjunctiva along with fornices on both upper and lower sides. These lesions were associated with areas of ulceration, hemorrhage and crusting. Mucopurulent discharge was present (Figure-1). The nasal part of cornea was visible on levoversion and it was hazy. Upon examination, the left eye was normal.

On evaluation of the lymph nodes, preauricular lymph node 10 x 15 mm in size and two submaxillary nodes 10 x 10 mm in size were palpated on the right side. They were firm in consistency with no tethering of the overlying skin. However the cervical nodes of neck were not palpable.

Because of his poor financial background, he was admitted to the free bed of the eye ward and all necessary investigations and treatment procedures were provided free of charge by the hospital.

The computed tomography scan of the orbit showed homogenously enhanced hyperdense tissue involving the anterior and lateral aspect of pre-septal space of right orbit (Figure-2). A chest X-ray and ultrasound scan of the liver revealed no evidence of metastatic lesions. Liver function test and serum LDH were within normal limits.

A tissue sample from the right eyelid was obtained by incisional biopsy and sent for histopathological examination, which showed round to spindle shaped tumor cells suggestive of malignant melanoma (Figure-3). Fine needle aspiration cytology was carried out in the pre-auricular and cervical lymph nodes. Metastatic tumor cells were present in FNAC of preauricular nodes (Figure-4) but the neck lymph nodes showed no evidence of tumor cells.

Hence, we made a provisional diagnosis of malignant transformation of kissing nevus involving the conjunctiva with periocular lymph node metastasis. Because the malignant lesion was associated with extraocular metastasis and poor visual prognosis, exenteration of the right orbit with lymph node dissection was planned. The patient was briefed about the surgery, the additional need for radiotherapy/chemotherapy, and his prognosis for survival, but he only consented to medical therapy and was unwilling to undergo surgical treatment. Though we counselled him about the gravity of the disease, he left the hospital and did not follow up for the further treatment.

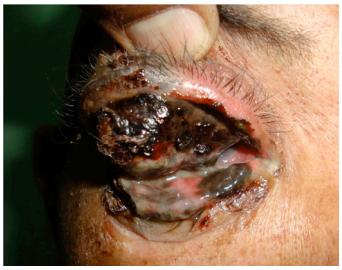

Figure 1: Clinical photography of right eyelids showing infiltrated conjunctiva protruding out associated with pigmentation, ulceration and mucopurulent discharge

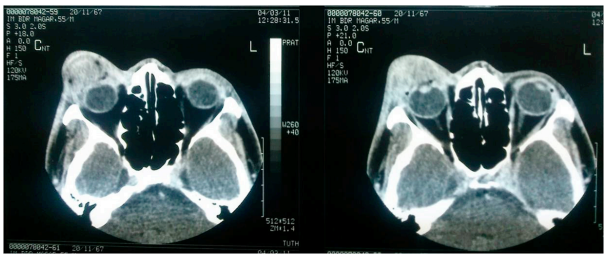

Figure 2: The computed tomography scan of the orbit showing homogenously enhanced hyperdense tissue involving the anterior and lateral aspect of pre-septal space of right orbit.

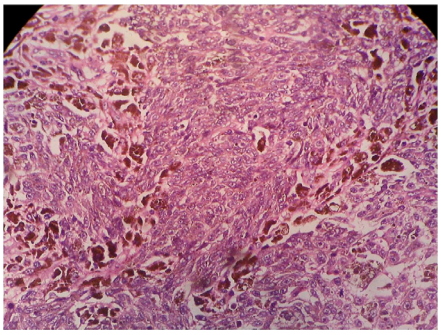

Figure 3: Histopathological examination of tissue sample from eyelid showing round to spindle shaped pigmented tumor cells.

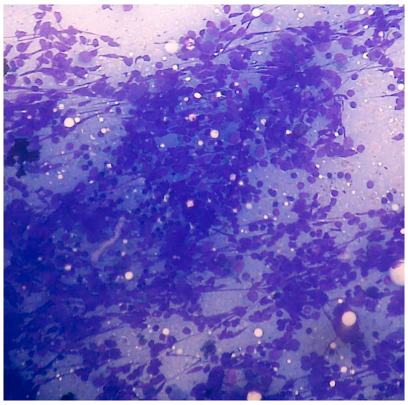

Figure 4: Fine needle aspiration cytology of preauricular lymph node with presence of the metastatic tumor cells.

Discussion

Malignant transformation of kissing or divided nevi of the eyelids is very rarely described. We herein report a very rare case of malignant transformation of kissing nevus with ocular and extraocular spread.

The reported incidence of congenital melanocytic nevi of malignant change is very variable, ranging from 2% to 30%, depending on the length of follow-up, with an average of 14% over a lifetime (Carmine et al, 2007).

The survival rates of patients with malignant melanomas generally depend on the depth of invasion. Breslow (Breslow et al, 1970) thickness is a well-known prognostic indicator for cutaneous melanoma and it has been found that lesions histopathologically measuring 0.76 mm or less are associated with a 5-year survival rate of 100%, whereas patients with tumors that had invaded more than 1.5 mm had a 5-year survival rate of only 50% to 60%. The presence of regional lymph node metastases is the single most important prognostic factor for most solid neoplasms. The presence of ulceration may be another high-risk histologic feature and may predict nodal metastasis, and the presence of extracapsular extension increases the risk of nodal recurrence. At the time of the first visit, the patient was in a potentially surgically curable stage. However, because he did not pursue further treatment, the malignancy became more extensive and the above mentioned complications suggest a poor prognosis. Hence, it is not only important for ophthalmologists to perform early biopsy of suspicious lesions when excision and reconstruction are easy, but to also promote awareness among patients so that they understand the severity of the disease. This will allow them to choose the most appropriate treatment that will lead to the best prognosis.

Conclusion

At the earliest sign of malignant transformation of a kissing nevus, prompt management is required. The tumors can grow very rapidly and progress from a surgically curable stage to an extensive disease with metastasis requiring exenteration with a poor survival prognosis.

Reference

Balch CM, Buzaid AC, Soong SJ, et al (2001). Final version of the American Joint Committee on Cancer staging system for cutaneous melanoma. J Clin Oncol; 19:3635–48.

Breslow A (1970). Thickness, cross-sectional areas and depth of invasion in the prognosis of cutaneous melanoma. Ann Surg; 172:902–8.

Carmine A, Stefano C, Federico DG, Paolo F, Gianni B and Nicolò S (2007). Divided nevus of the eyelids: three case studies; 21: 137-140.

De Pietro WP, Sweeney EW and Silvers DN(1981). Divided nevi:pigmented and achromatic. Cutis; 27(4): 408-409.

Goggins WB, Tsao H (2003). A population-based analysis of risk factors for a second primary cutaneous melanoma among melanoma Survivors. Cancer; 97:639–643.

Font RL (1996). Eyelids and lacrimal drainage system. In: Spencer WH, ed. Ophthalmic Pathology: An Atlas and Textbook, 4th ed. Philadelphia: W.B. Saunders; v4, chap. 11, 2270–8.

McDonnell PJ and Mayou BJ (1988). Congenital divided naevus of the eyelids. Br J Ophthalmol; 72(3): 198-201.

Papadopoulos O (1991). Divided nevus of the eyelid. Plast Reconstr Surg; 88(2): 331-333.

Rodriguez-Sains RS, Jakobiec FA, Iwamoto T (1981). Lentigo maligna of the lateral canthal skin. Ophthalmology; 88:1186–92.

Source of support: nil. Conflict of interest: none