

Divided Nevus of the Eyelids: Three Case Studies

CARMINE ALFANO¹, STEFANO CHIUMMARELLO², FEDERICO DE GADO²,
PAOLO FIORAMONTI², GIANNI BISTONI¹ and NICOLÒ SCUDERI²

¹*Department of Plastic and Reconstructive Surgery, University of Perugia;*

²*Department of Plastic and Reconstructive Surgery, University of Rome "La Sapienza", Rome, Italy*

Abstract. *Divided nevus or kissing nevus is a rare form of congenital nevus that usually occurs on adjacent parts of the upper and lower eyelids of one eye. Most often, the formation is present from birth, but it may also appear later. Three new cases during the period 1996 to 2004 are reported. Two of them were treated in our unit for oncological, functional and aesthetic purposes, and one preferred not to be operated on. All our patients had divided nevus of the left eyelid. In the first case, the reconstruction was achieved with two tarsoconjunctival flaps covered with skin grafts. In the second case, we used a technique based on autologous cultivated conjunctival epithelium. The third patient preferred not to be operated on and is still under clinical follow-up. Because of the severe disfigurement, risk of later malignant change in the lesion, and the possibility of deprivation amblyopia, early surgical treatment is very important.*

Divided nevus or kissing nevus is a rare form of congenital nevus that usually occurs on adjacent parts of the upper and lower eyelids of one eye. When the eyelids are closed the eye appears to be covered by one large nevus. It usually involves contiguous portions of the upper and lower eyelids margins and it may cause functional and aesthetic problems (1). Divided nevus of the lids was first described in 1919 by Fuchs, since that first case less than 40 have followed in literature. It is rare in other locations of the body: in 1993 the first divided nevus of the fingers was reported (2) and in 1978, Sonoda, described the first case on the penis. Since then, others have been reported (3-5). Usually, divided nevi are localized in the median region of the eyelid, even though they might also occur in the canthal regions (6). Size varies from small (few millimetres) to larger forms, the latter of which may even cause mechanical ptosis of the eyelid. The conjunctiva is rarely affected, whereas the cilia

are larger than normal. Bilateral divided nevi of the eyelids have never been reported.

Most often, the formation is present from birth, but it may also appear later; it shows a very slow growth, which may stop at any age. The divided nevus is thought to arise during fetal development at a time when the eyelids are fused. The growth arises from melanoblasts or Schwann cells of a neuroectodermic origin. The lids appear as ectodermal protrusions from the 6th week of fetal development. They grow towards each other and gradually fuse until about the 24th week when they gradually separate. Melanoblasts originate from the neural crest and migrate during the 12 to 14 week stage of gestation to colonize the epidermis. It is no longer a hypothesis that the melanoblasts are present when lids are fused. In fact the divided melanocytic nevus arises just before the 15th week of fetal development (7). Afterwards, when the complete separation between the two eyelids takes place, the single nevus will eventually be divided into two distinct portions: one on the upper and one on the lower eyelid.

We report an update of our experience of divided nevus of the eyelid with three cases observed between 1996 and 2004. Two were treated in our unit for oncological, functional and aesthetic purposes, one preferred not to be operated on.

Patients and Methods

Case 1. The patient was a 22-year-old woman with a congenital divided nevus of the left eyelid. Family history for melanoma was negative. On this established lesion she developed a red nodule on the lower eyelid during her second pregnancy. Inspection showed a pigmented lesion, 1-1.5 cm diameter, affecting the middle and outer of the left upper eyelid. The lower eyelid was also affected, but the pigmented lesion was larger, thicker, and darker, with a central purple red nodule.

With the eyelid closed, these nevi acquire perfect contiguity with their contact surfaces and resemble a single entity. Conjunctiva, sclera and cornea were exempt from melanocytic pigmentation. The patient also had a palpable lymph-node, 1 cm in diameter, localized in the right lateral cervical region.

The rapid growth of the red nodule, together with the presence of lymphadenopathy, led us to take a surgical approach. Surgical

Correspondence to: Paolo Fioramonti, MD, Via Cardinal Cassetta 11, 00165, Roma, Italy. Tel: +393470028772, Fax: +396491525, e-mail: fioramontipaolo@hotmail.com

Key Words: Divided nevus, eyelids, kissing nevus.



Figure 1. A) Case 2: Kissing Nevus affecting the left upper and lower eyelid. The outer surface of the sclera is also affected. B) The post-operative picture clearly shows a satisfactory aesthetic result with a good eye opening.

therapy, after a full thickness excision, was based on a tarsoconjunctival flap for the upper and lower lid, similar to the Tenzel and Hughes procedures. After a tarsorraphy, a full thickness skin graft from the left retroauricular region was used to cover the flaps.

Pathology reports indicated a hyperplastic lymphadenitis for the nodule on the neck and a dermal nevocellular nevus with junctional residuals for the red nodule on the eyelid.

Case 2. The patient was a 25-year-old man with a congenital divided nevus of the left eye involving skin and conjunctiva of the upper and lower eyelids. Family history reported a cleft lip and palate operated on when he was 4 months old. Inspection showed a rilevated lesion, 2.5x1.5 cm, on middle and outer of the left upper eyelid and 2.5x2 cm pedicle on the same surface of the lower eyelid (Figure 1A). The outer surface of the sclera was also affected. Hypertrophic growth of the lesion linked ciliar margin was observed too. The eyelid closed showed a pathognomonic kissing nevus.

Initially we removed the nevus and reconstructed the eyelids with local skin flaps and lateral canthopexy. Starting with cells taken from the biopsy specimen, in 21 days a confluent epithelial sheet of autologous cultivated conjunctival epithelium was developed. We restored the damaged ocular surface with the new epithelium. After three days, conjunctival continuity was re-established with excellent functional and cosmetic outcome (8). A six-year follow-up (Figure 1B) was performed with a conjunctival biopsy and the histological exam showed mucous conjunctival fragments free from significative histopathological alterations. The conjunctiva consisted of a pluristratified squamous, non-keratinised epithelium composed of four to five layers, laying on a basement membrane and with a lamina propria made of well-vascularized connective tissue. Further, the electron microscopy gave evidence of a normal ultrastructure for the conjunctiva.

Case 3. The patient was a 12-year-old girl with a congenital divided nevus of the left eye (Figure 2). Inspection showed a lesion in the left upper eyelid involving all the medial and the outer skin surface, about 6 x 3.5 cm, with 3.5x1.5 cm on the medial area of the lower eyelid. Conjunctiva and cornea were untouched.



Figure 2. Case 3: The eyelid closed showed a pathognomonic Divided Nevus extended to the eyebrow.

The patient came to our attention for aesthetic problems, interested to know if the lesion could be dangerous. We proposed removal of the nevus but the parents refused any surgical treatment. The patient started a periodic 6 month follow-up to keep the lesion and its evolution under control.

Results

Before the malignant potential of these lesions was recognised the therapy consisted of observation and sometimes late surgical removal and reconstruction for cosmetic reasons.

In our first case the reconstruction was achieved with two tarsoconjunctival flaps covered with a skin graft. In the second case, traditional surgery was not able to achieve a complete functional and cosmetic reconstruction so we

decided to combine surgery with the use of autologous cultivated conjunctival epithelium, which in our experience can be a very helpful tool. In the third case the patient preferred not be operated on. We wanted to excise the divided nevus, use an orbicularis oculi musculocutaneous flap and partially incise it to form the contour of the lateral canthus.

Histopathologically, divided nevus is usually a typical intradermal nevus, as in the second case, sometimes showing junctional activity, as in the first case. Interestingly, histopathological differences between upper and lower nevi have never been shown.

Discussion

Divided nevi of the eyelids are very rare, in literature we found only 34 cases have been reported.

Although a divided melanoma of the eyelids has never been described, a malignant transformation of a divided nevus might take place with a very fast growth (9, 10). There is only one reported case of malignant transformation in a divided nevus of the eyelids and this introduces the problem of a differential diagnosis with Dubrouilh's melanosis (*melanoma in situ*) or a neurofibroma, as with pigmented basal cell carcinoma. The reported incidence for 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% for a whole lifetime.

Divided 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 the 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 (11, 12).

The treatment of these lesions is still not subjected to a definite guideline and only a few solutions may be applied. Early reconstructive surgery should be preferred to achieve the best aesthetic result. Dermabrasion has given good results but sometimes cells remain in the deep dermis and, not surprisingly, the lesion recurred fairly quickly.

The usual treatment for small lesions consists of full thickness excision followed by repair with full thickness skin graft from retroauricular region or contralateral eyelid.

Large and deep nevi involving the eyelids may have to be treated in stages. Where the surgical gap repair requires functional support, a flap (tarso-conjunctival, musculocutaneous) is performed.

We do believe that the nevi particular location and uniqueness command a specific evaluation and treatment for each single patient.

References

- 1 Ribuffo D, Cavalieri L, Sonnino M, Tarallo M, Latini C, Muratori L and Scuderi N: Divided nevus of the eyelid: a case report. *Ophthal Plast Reconstr Surg* 12(3): 186-189, 1996.
- 2 Hayashi N and Soma Y: A case of epidermal nevi showing a divided form of the fingers. *J Am Acad Dermatol* 29: 281-282, 1993.
- 3 Kono T, Nozaki M, Kikuchi Y, Ercocen AR, Hayashi N, Chan HH, Ohkubo R and Fukaya E: Divided naevus of the penis: a hypothesis on the embryological mechanism of its development. *Acta Derm Venereol* 83(2): 155-156, 2003.
- 4 Choi GS, Won DH, Lee SJ, Lee JH and Kim YG: Divided naevus on the penis. *Br J Dermatol* 143(5): 1126-1127, 2000.
- 5 Desruelles F, Lacour JP, Mantoux F and Ortonne JP: Divided nevus of the penis: an unusual location. *Arch Dermatol* 134(7): 879-880, 1998.
- 6 Patrinely JR: Surgical management of divided nevus of the eyelid. *Plas Reconstr Surg* 88(2): 331-333, 1991.
- 7 Hamming N: Anatomy and embryology of the eyelids: a review with special reference to the development of divided nevi. *Pediatr Dermatol* 1: 51-58, 1983.
- 8 Scuderi N, Alfano C, Paolini G, Marchese C and Scuderi G: Transplantation of autologous cultivated conjunctival epithelium for the restoration of defects in the ocular surface. *Scand J Plast Reconstr Surg Hand Surg* 36: 340-348, 2002.
- 9 Kasai K and Ogawa Y: The successful treatment of a divided nevus on a young patient using a modified Kunhnt-Szymowski procedure. *Ophthal Plast Reconstr Surg* 6(3): 181-184, 1990.
- 10 McDonnell PJ and Mayou BJ: Congenital divided naevus of the eyelids. *Br J Ophthalmol* 72(3): 198-201, 1988.
- 11 Papadopoulos O: Divided nevus of the eyelid. *Plast Reconstr Surg* 88(2): 331-333, 1991.
- 12 De Pietro WP, Sweeney EW and Silvers DN: Divided nevi: pigmented and achromatic. *Cutis* 27(4): 408-409, 1981.

Received August 29, 2006

Revised October 19, 2006

Accepted October 26, 2006