Saudi Journal of Ophthalmology (2017) 31, 165–168

Case Report

Congenital distichiasis: Histopathological report of 3 cases

Hind Manaa Alkatan [a,*]; Alicia Galindo-Ferreiro [b,c]; Azza Maktabi [b]; Alberto Galvez-Ruiz [b]; Silvana Schellini [b,d]

Abstract

Distichiasis is a condition clinically presenting as partial or complete accessory row of lashes that emerges from the meibomian glands orifices. It can be an acquired or congenital with an autosomal dominant inheritance. The histopathological features are not well described in the ophthalmic literature, however they include abnormal pilosebaceous units within the posterior lamella of the eyelid and perifollicular chronic inflammatory cell infiltration. In this report, we describe the histopathological findings of three congenital distichiasis cases treated at King Khaled Eye Specialist Hospital (KKESH), Riyadh, Saudi Arabia with discussion on the pathogenesis of such a condition and the differentiating features from ectopic cilia.

Keywords: Congenital, Eyelid, Distichiasis, Cilia, Histopathology

http://dx.doi.org/10.1016/j.sjopt.2017.05.003

Introduction

Distichiasis is a rare acquired or congenital condition that could be either congenital with autosomal dominant inheritance or acquired and is characterized by an abnormal row of lashes that appears to be emerging from the Meibomian glands orifices and can affect one or more eyelids [1–3]. The acquired form is less common and is thought to be related to chronic inflammatory conditions of the eyelids [4]. Many conditions have been reported in association with distichiasis such as: blepharophymosis, hereditary spinal arachnoid cysts, microphthalmus, corneal hypoaesthesia, uvula bifida, cleft palate and some syndromes including: Oculo-cerebro-renal and Pierre Robin syndrome [1,5].

Clinically, the abnormal lashes may cause tearing, photophobia, foreign body sensation and conjunctival injection due to the rubbing effects of these displaced lashes [1]. The histopathological characteristics of distichiasis are not well described in the ophthalmic literature. Therefore we are reporting 3 cases of congenital distichiasis treated surgically, which allowed us to do thorough tissue examination and description of the histopathological findings.

Case reports

Case 1

A 4 year-old female with history of dense abnormal lashes along the lateral part of her left upper lid since birth causing irritation and redness of that eye thus they were regularly removed by the parents. The lashes were described as being thinner at birth but becoming thicker with repeated epilation. There was no family history of similar condition or any systemic association. The location of the lashes was observed upon examination 2 mm below the lash line where the meibomian gland orifices were located. The lid margin in that area was pigmented and the bulbar conjunctiva was com-

Received 27 October 2016; accepted 9 May 2017; available online 18 May 2017.

[a] Departments of Ophthalmology & Pathology, King Saud University College of Medicine, Riyadh, Saudi Arabia
[b] Department of Pathology and Oculoplastic Division, King Khaled Eye Specialist Hospital, Saudi Arabia
[c] Department of Ophthalmology, Rio Hortega University Hospital, Valladolid, Spain
[d] Department of Ophthalmology, Faculdade de Medicina de Botucatu - UNESP, São Paulo, Brazil

* Corresponding author at: Departments of Ophthalmology & Pathology, King Saud University Medical City, PO Box 18097, Riyadh 11415, Saudi Arabia.
e-mail addresses: hindkatan@yahoo.com, hkatan@ksu.edu.sa (H.M. Alkatan).

☆ This project has been approved by the Research Department at KKESH with the following number: RP 1596-CR.

Peer review under responsibility of Saudi Ophthalmological Society, King Saud University

Production and hosting by Elsevier

Access this article online:
www.saudiophthaljournal.com
www.sciencedirect.com

pletely normal (Fig. 1A). The patient underwent surgical excision of the distichiatic lashes with tarso-conjunctival graft. Histopathology of the excised portion of the eyelid showed numerous aberrant hair follicles adjacent to the sebaceous glands located in the posterior lamella. Additional features included significant fibrosis and chronic inflammatory cell infiltration mainly composed of lymphocytes surrounding the follicles and glands (Fig. 1B–D). The postoperative cosmetic appearance of the left upper lid was satisfactory.

Cases 2 and 3

They were brothers, the first was a 7 year-old male with long standing bilateral congenital distichiasis presenting recently with photophobia and conjunctival discharge. He was previously treated by electrolysis under general anesthesia. The clinical examination showed similar abnormal rubbing lashes emerging from the meibomian gland orifices of all his eyelids with keratinization of the lid margins and superficial corneal abrasions in both eyes. Interestingly, this patient had positive family history of an affected father and 2 brothers. Similar surgical procedure was performed in both upper lids. Histopathological evaluation of the excised full thickness portions of the lid margin showed several hair follicles within the posterior lamellae surrounded by extensive fibrosis likely due to the cautery, which had also affected both the epithelium and the subepithelial tissue in the form of the absence of most of the meibomian glands and focal chronic inflammatory cells infiltration mainly around the follicles (Fig. 2A–C).

The third case was a 12 year-old male with similar history of congenital distichiasis presenting with conjunctival discharge, photophobia and foreign body sensation in the left eye. Similarly, his left cornea showed recent superficial abrasions in addition to superior corneal scarring. The affected eyelid margin was keratinized and distorted due to previously repeated electrolysis and entropion repair. The same surgical procedure was performed on that eyelid with satisfactory cosmetic result 9 months postoperatively (Fig. 2D). The Histopathological findings were almost identical to the second case with more profound fibrosis, inflammatory process around the follicles, and meibomian glands atrophy. Both patients were genetically tested for lymphedema-distichiasis syndrome and FOXC2 was negative in both.

Discussion

Congenital distichiasis is a rare condition where abnormal thin lashes are observed to emerge from the ostia of the meibomian glands of the eyelids [1–3]. It should be clinically differentiated from ectopic cilia, which are usually thicker and well pigmented, similar to normal lashes. Congenital ectopic cilia also present with grouped hair follicles within the anterior lamella with widened epidermal orifices in contrast to distichiasis where the lashes originate from the posterior lamellae [6]. The acquired aberrant cilia on the other hand arise in

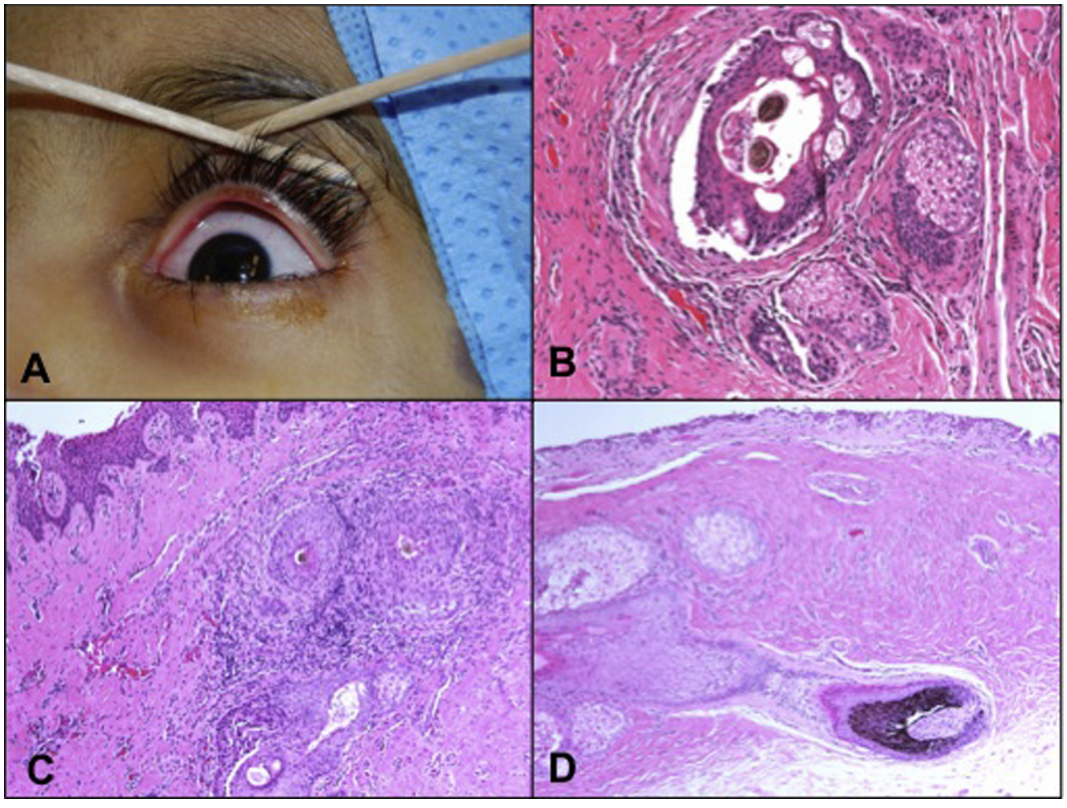

Fig. 1. (A) Preoperative appearance in Case 1 showing hyperpigmentation along the upper lid margin, however lashes are not apparent since they were recently epilated her before the surgery. (B) Histopathological appearance of the aberrant lashes within the tarsus in the post lamellae surrounded by the tarsal meibomian glands. Also note the presence of 2 hair shafts (Original magnification X400 Hematoxylin and Eosin). (C) Perifolliculitis of the aberrant lashes in the eyelid posterior lamellae (Original magnification X100 Hematoxylin and Eosin) in the same case. (D) Histopathological photograph demonstrating the origin of the distichiatic lash from the posterior lamellar part of the eyelid (following surgical splitting). Note the hair follicle and the associated meibomian glands (Original magnification X100 Hematoxylin and Eosin).

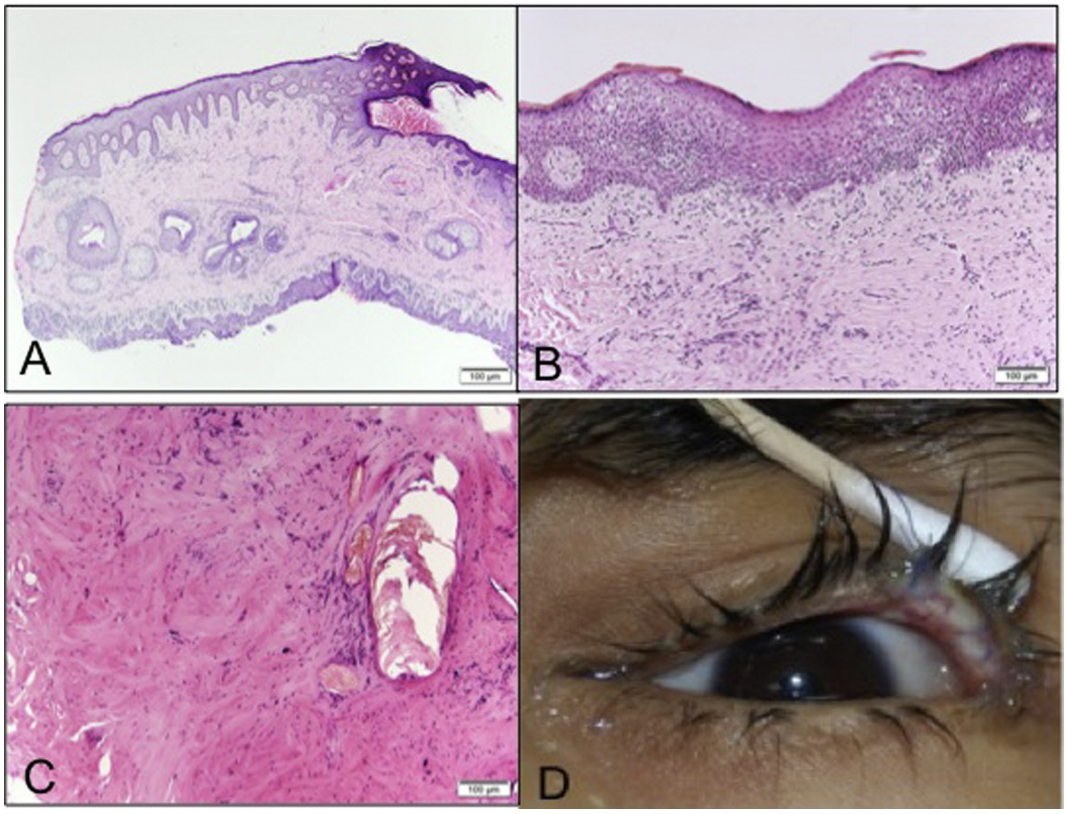

Fig. 2. (A) Lower power histopathological appearance of the excised posterior lamellae of the lid in Case 2 showing numerous hair follicles in the tarsal plate (Original magnification X 40 Hematoxylin and Eosin). (B) Histopathological evidence of cautery effect of the previous electrolysis at the lid margin resulting in acanthosis, hyperkeratosis and sub epithelial dense fibrosis (Original magnification X 100 Hematoxylin and Eosin). (C) Histopathology in Case 2 showing residual hair shaft post-electrolysis with no preserved follicle surrounded by dense fibrosis and focal chronic inflammation (Original magnification X 100 Hematoxylin and Eosin). (D) Case 3 recent postoperative clinical picture following surgical treatment showing the upper lid lateral tarso-conjunctival graft area.

the superior palpebral conjunctiva, in association with the anatomical location of the crypts of Henle as well as the presence of chronic inflammation [7]. In all our patients, the distichiatic lashes were observed to be shorter, thinner and less pigmented than the normal lashes in the corresponding patients and their origin was well demonstrated in relation to the meibomian gland orifices at the lid margins. The pathogenesis is also different between the 2 conditions. In congenital distichiasis, a developmental anomaly occurs in which complete pilosebaceous unit(s) (with hair and glandular structures) is present in the posterior lamellae. This likely represents a failure of the primary epithelial cells to differentiate selectively into a sebaceous gland only. Instead, it develops into a complete pilosebaceous unit, in which case it would not represent a true metaplasia as has been previously suggested. However, we cannot totally eliminate an in utero injury leading to metaplasia in congenital distichiasis, but this seems unlikely [6,7].

Histopathologically, features and findings of distichiasis are not well described in the ophthalmic literature. Distichiatic lashes have been described to arise within the dense connective tissue of the tarsal plate but marked fibrosis and intense chronic inflammatory cell infiltration were not always present [8]. The histopathological features of distichiasis in our study were almost identical where abnormal pilosebaceous units were observed in the posterior lamella of the eyelids associated with perifollicular chronic inflammatory cell infiltration. The distichiatic lashes emerged from the ducts of the tarsal Meibomian glands. Several hair shafts were present in the first case within a single lumen of the duct of these tarsal glands. This last finding was against the clinical diagnosis of ectopic cilia as mentioned earlier. We believe that the extensive fibrosis, epithelial changes of the lid margin and the marked inflammatory process in Cases 2 and 3 were due to the history of repeated epilation and/or electrolysis.

Few treatment modalities including epilation, electrolysis, lid margin cryotherapy, split of the posterior lamella and the use of grafts have been described with limited benefit, however this is beyond the scope of our report [9,10].

In conclusion, we are describing the unique histopathological findings in congenital distichiasis with emphasis on the origin and pathogenesis of the abnormal lashes in this condition. Ophthalmologists and ophthalmic pathologists should be able to differentiate this condition from ectopic cilia.

Conflict of interest

The authors declared that there is no conflict of interest.

References

1. O'Donnell BA, Collin JR. Distichiasis: management with cryotherapy to the posterior lamella. *Br J Ophthalmol* 1993;**77**:289–92.
2. Fox SA. Distichiasis. *Am J Ophthalmol* 1962;**53**:14–8.

3. Wolfley D. Excision of individual follicles for the management of congenital distichiasis and localized trichiasis. *J Pediatr Ophthalmol Strabismus* 1987;**24**:22–6.
4. Scheie HG, Albert DM. Distichiasis and trichiasis: origin and management. *Am J Ophthalmol* 1966;**61**:718–20.
5. Hoover RE, Kelley JS. Distichiasis and lymphedema: a hereditary syndrome with possible multiple defects. A report of a family. *Trans Am Ophthalmol Soc* 1971;**69**:293–306.
6. Jakobiec FA, Yoon MK. Histopathologic proof for the origin of ectopic cilia of the eyelid skin. Graefe's archive for clinical and experimental ophthalmology = Albrecht von Graefes Archiv fur klinische und experimentelle. *Ophthalmologie* 2013;**251**:985–8.
7. Hase K, Kase S, Noda M, Ohashi T, Shinkuma S, Ishida S. Ectopic cilia: a histopathological study. *Case Rep Dermatol* 2012;**4**:37–40.
8. Raymond-Letron I, Bourges-Abella N, Rousseau T, Douet JY, de Geyer G, Regnier A. Histopathologic features of canine distichiasis. *Veterin Ophthal* 2012;**15**:92–7.
9. Anderson RL, Harvey JT. Lid splitting and posterior lamella cryosurgery for congenital and acquired distichiasis. *Arch Ophthalmol* 1981;**99**:631–4.
10. White JH. Correction of distichiasis by tarsal resection and mucous membrane grafting. *Am J Ophthalmol* 1975;**80**:507–8.