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Case Report

Congenital distichiasis Histopathological report of 3 cases

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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

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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-

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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

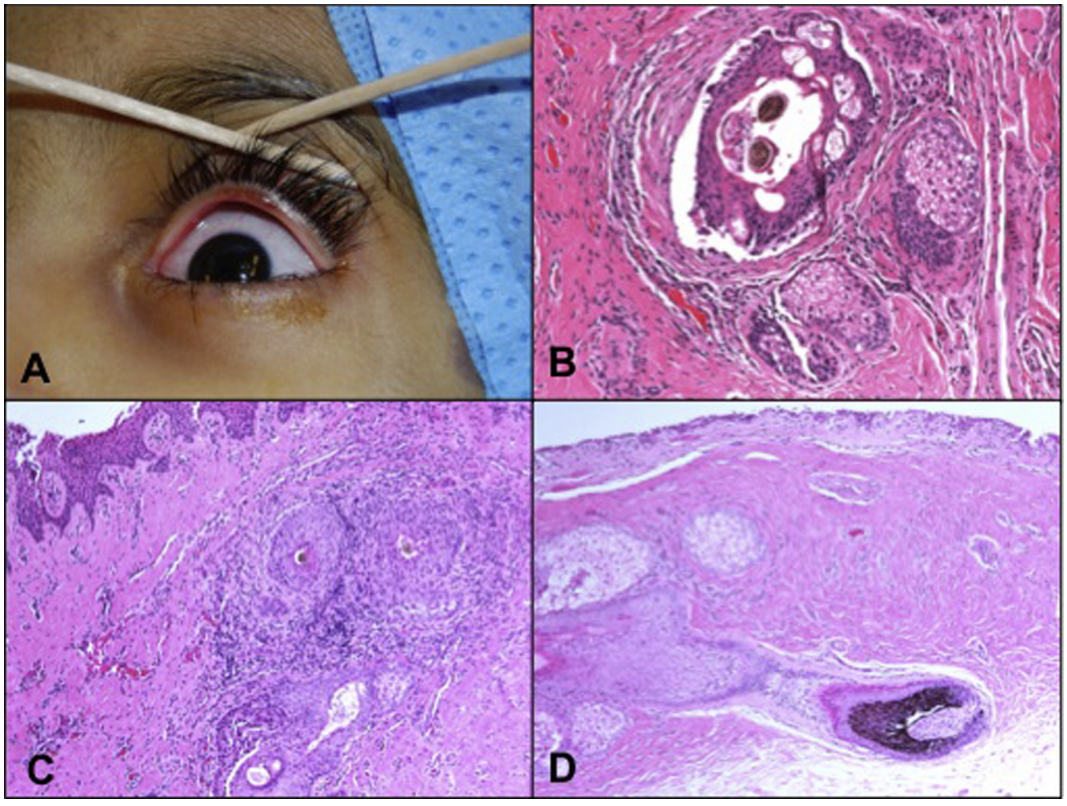

Fig. 1. (A) Preoperative appearance in Case 1 showing hyperpigmentation along the upper lid margin, however lashes are not apparent since they were recently epilated her before the surgery. (B) Histopathological appearance of the aberrant lashes within the tarsus in the post lamellae surrounded by the 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).

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