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Case Report

Rare Choristoma of the Tarsal Conjunctiva: Critical Inspection Prevents Unnecessary Mutilation

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Abstract
We report a case of a 26-year-old male patient with an incidental finding of a flesh-colored tumor with few vellus hairs on its surface, located on the fornix and the tarsal conjunctiva of the left lower eyelid. Histology of the biopsy showed a choristoma consisting of abundant vellus hairs, sebaceous glands, and sparse lacrimal gland tissue. Recognition of conjunctival vellus hairs is significant and raises the suspected diagnosis of choristoma, which can be confirmed by a small sample biopsy. A complete excision is unnecessary and possible surgical complications can be prevented. Choristomas of the tarsal conjunctiva are very rare and, to our knowledge, this is the first histological documentation of a choristoma containing vellus hair located on the tarsal conjunctiva. In addition, we review the histopathological findings of choristomas and their differential diagnoses.

Introduction

Choristomas are benign congenital lesions composed of mature tissue elements in an abnormal location. Most conjunctival choristomas are epibulbar on the scleral surface. They are uncommon lesions, mainly presenting in children [1–3]. Tarsal choristomas are very rare.

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In the literature, we have found just two documented tarsal cases, one containing lacrimal gland tissue, the other containing bone [4, 5]. To our knowledge, this is the first histological documentation of a choristoma containing vellus hair located on the tarsal conjunctiva.

Vellus hair is fine, barely pigmented, short hair. It can normally be found on the majority of the body surface of children. It is differentiated from the thicker, longer, and pigmented terminal hair (e.g. eyelashes, facial hair, scalp hair). In contrast to vellus hair, terminal hair histologically shows a central medulla or core.

The aim of this case report was to highlight the importance of clinically recognizing the discrete vellus hairs and thus making the presumptive diagnosis of rare tarsal choristoma. Consequently, to confirm the diagnosis, a sample biopsy is sufficient and a surgical complication after excision can be avoided. In addition, we provide an overview of the choristoma literature and discuss the different terms used.

Case Report

A 26-year-old male patient with a history of facial and eyelid dermatitis presented with dry eyes and blepharitis. Upon ophthalmological examination, a firm conjunctival lesion of the inferior tarsus/fornix was found incidentally (Fig. 1a). Since the lesion was only visible after eyelid eversion, the patient had not noticed it before. Macroscopically, the lesion presented as a flesh-colored tumor of 6 mm × 3 mm with several vellus hairs visible on the tarsal surface, distant from the eyelid margin (Fig. 1b). A diagnostic biopsy was performed, which included deep tarsal tissue (Fig. 1c). The histological examination showed a superficially and deeply located tarsal tumor, containing a cluster of hair follicles with and without vellus hairs. No terminal hairs were present. In addition, few sebaceous glands, sparse lacrimal tissue, and chronic inflammation were found. The covering conjunctiva was inconspicuous, without atypia (Fig. 2). These findings were consistent with the diagnosis of a choristoma.

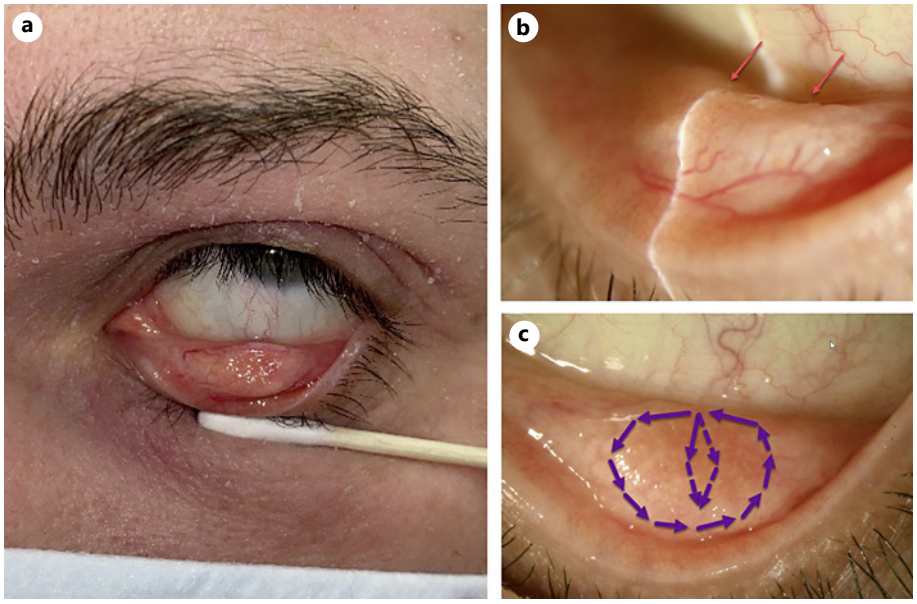

Fig. 1. Clinical picture: **a** overview of the conjunctival tarsal lesion (pre-operative). **b** Fine vellus hairs on the surface of the lesion. **c** Outer arrows delineate the whole lesion, inner arrows the sample biopsy.

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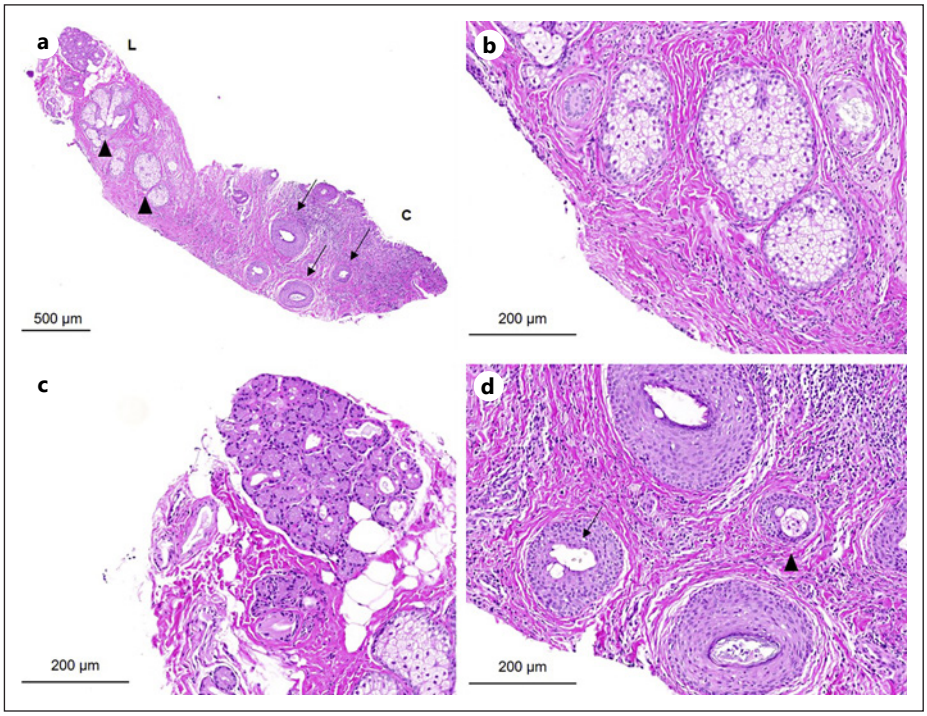

Fig. 2. Histological findings: **a** overview with subconjunctival hair follicles with and without vellus hairs (arrows), sebaceous glands (arrow heads), and sparse lacrimal tissue (L) covering conjunctiva (C). Absence of terminal hairs. **b–d** Higher magnification of sebaceous glands (**b**), lacrimal gland tissue (**c**), hair follicles with vellus hairs (arrow head, **d**), and hair follicles without vellus hairs (arrow, **d**).

Discussion and Conclusions

In reviewing the literature, we searched the PubMed database for the terms conjunctival choristoma, tarsal choristoma, conjunctival hamartoma, tarsal hamartoma, dermolipoma, lipodermoid, and follicle nevus. We noticed how confusing the nomenclature is. Congenital epibulbar dermoid and lipodermoid are the most commonly reported choristomas and are predominantly diagnosed during childhood (60–70% of cases) [3]. However, by definition, as recently performed for epibulbar lacrimal glands reported by Milman et al. [6] most should be classified as hamartomas and not choristomas. Choristomas and hamartomas have in common that they both arise from pluripotent rests of germ cells, which can differentiate into various mature tissues. The difference between the two entities is that the newly grown tumor tissue is alien in the case of choristomas and not in the case of hamartomas [7]. For naming our lesion, this definition is not very helpful.

The evidence of sparse sebaceous and lacrimal glands, which are not alien in this location, may suggest the diagnosis of a hamartoma. However, due to the vellus hairs, which do not occur in the conjunctiva, the tumor should be classified as choristoma. Even though vellus hair can theoretically develop backward from terminal hair follicles such as eyelashes, we consider the vellus hair on the tumor surface as foreign since the tumor was in no close proximity to the eyelid margin. Overall, our tumor raises the question whether the distinction choristoma versus hamartoma is meaningful.

Tarsal choristomas are very rare. Conjunctival choristomas are mostly located epibulbar between the conjunctiva and sclera. In the literature, we have found just two documented cases of choristomas of the tarsal conjunctiva, one containing lacrimal gland tissue, the other

containing bone [4, 5]. In addition, we found one smooth muscle hamartoma of the fornix extending to the inferior tarsal border [8].

In contrast, we found several publications concerning ectopic cilia of the eyelid. However, most of them were located epidermal and contained terminal hair [9–11].

One reported case of cilia was on the tarsal conjunctiva but very close to the eyelid margin. Histology shows the close relationship to the eyelashes and a marked inflammatory reaction suggesting the diagnosis of cilia inversum rather than a newly formed hair [12]. Furthermore, we found three reports of clear-cut hair located on the tarsal conjunctiva [13–15]. The clinical photograph shown by Guler et al. [13] closely resembles the macroscopic findings of our patient. However, these publications all only feature clinical images and no histology. In contrast to previous publications, we are the first to document histologically a choristoma of the tarsal conjunctiva containing vellus hair.

The limitation of this case report is that only a single patient case is described and that there has not been a long-term follow-up. Further, systematic studies are needed to achieve a systematic terminology of conjunctival choristomas/hamartomas.

In conclusion, clinical detection of conjunctival vellus hairs is significant and raises the suspicion of choristoma, which can be confirmed by a small-sample biopsy. A complete excision is not necessary and possible surgical complications can be avoided.

Acknowledgment

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Statement of Ethics

Research was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. Written informed consent was obtained from the patient for publication of this case report and any accompanying images. This retrospective review of patient data did not require ethical approval in accordance with local/national guidelines.

Conflict of Interest Statement

The authors declare that they have no competing interests.

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Authors Contributions

Daniela Mihic-Probst and Viola Katharina Vetter performed the histological examination and were major contributors in writing the manuscript. Maria E. Correa-Perez and Karla Chaloupka provided the clinical photographs and the clinical case presentation. Daniela Mihic-Probst, Viola Katharina Vetter, Maria E. Correa-Perez, and Karla Chaloupka read and approved the final manuscript.

Data Availability Statement

All data that support the findings of this study are included in this article. Further inquiries can be directed to the corresponding author.

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