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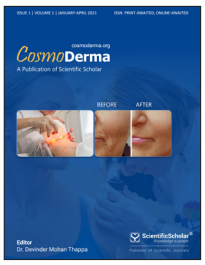

Original Article

Dermal cylindroma – An update and a series of uncommon clinical presentations

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ABSTRACT

Objectives: Dermal cylindroma is an uncommon adnexal tumor. Although frequently occurring in the head-and-neck region as a firm nodule with smooth surface, the tumor may rarely occur on other body sites and present atypically.

Material and Methods: Patients files from the department during the years 2003–2022 have been analyzed. Clinical data, histopathology, and treatment were collected with a focus on atypical presentations. A short literature review has been performed using PUBMED.

Results: We identified six adult Caucasian patients with Fitzpatrick skin type II–III aged between 42 and 74 years, two females and four males. We observed cylindroma of the auricle and the lower leg. One tumor presented as a flat plaque and another one was pedunculated with a mushroom-like shape. Two patients had syndromic cylindroma. One patient presented only with multiple cylindromas of the scalp but no other associated adnexal tumors of Brooke-Spiegler syndrome, while the second one developed multiple malignant tumors of skin and parotid glands.

Conclusion: Dermal cylindroma rarely occurs outside the scalp. The clinical presentation is variable. Syndromic cases may present either oligosymptomatic or with multiple associated malignant tumors.

Keywords: Dermal cylindroma, Brooke-Spiegler syndrome, CYLD gene, atypical presentation

INTRODUCTION

Dermal cylindroma is uncommon benign adnexal tumor. The name was coined by Billroth in 1859 for a salivary gland tumor with similar histopathology.[1] Cylindromas are not restricted to skin but here we will focus on dermal cylindroma only.

On histopathology, cylindromas are composed irregular isles of basaloid epithelial cells arranged like a "jigsaw puzzle" without connection to the overlying epidermis. The cells demonstrate a dual lineage of palisading peripheral smaller cells with hyperchromatic nuclei and more differentiated pale cells and nuclei forming small duct-like structures in the center. The cell islands are embedded into interconnecting hyalinized sheaths.[2]

Cylindroma is slow growing, mostly asymptomatic, and usually small by size. It is not hair-bearing itself and shows no horripilation. The surface is smooth and the shape regular with a nodular appearance. The majority of cylindromas can be found in the head-and-neck region. The tumor shows a clear female predominance. Multiple scalp cylindromas in a single patient are also known as turban tumor. Malignant transformation is rare.[2,3]

Cylindromas may develop either sporadic or in a familial fashion. Sporadic tumors often present as a solitary nodule. They often exhibit *MYB-NFIB* fusion transcripts which are oncoproteins. Activated or overexpressed *MYB* can function in a similar way promoting the development of dermal cylindromas. Familial cylindromatosis (FC) syndromes is mostly associated with bi-allelic truncating mutations of the tumor-suppressor gene *CYLD* that controls the activity of the transcription factor nuclear factor Kappa B. *MYB* target genes become up-regulated in *CYLD*-defective tumors. The *CYLD* gene mutations are involved in the following inherited subtypes: (a) Brooke-Spiegler Syndrome (BSS), (b) FC, and multiple familial trichoepitheliomas. Inheritance occurs in an autosomal dominant pattern. There is no clear relationship between mutations and specific phenotypic expression, suggesting the role of environmental factors. Syndromic forms manifest at earlier age then sporadic types.[4-6] These entities are grouped under the umbrella of the *CYLD* syndrome.[7] Such patients may present a genetic mosaicism of skin.[8]

MATERIALS AND METHODS

Patients files from the department during the years 2003–2022 have been analyzed. Clinical data, histopathology, and treatment were collected with a focus on atypical presentations. A short literature review has been performed using PUBMED.

RESULTS

We identified six adult Caucasian patients with Fitzpatrick skin type II–III aged between 42 and 74 years, two females and four males.

Patients provided verbal and written consent. Presentation follows the recommendations of the CARE list (https://www.care-statement.org/checklist). All the procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the 2008 revision of the Declaration of Helsinki of 1975.

1. The first patient was a 71-year-old Caucasian man, Fitzpatrick skin Type II, with a history of colon cancer, published previously.[9] He presented with a 2.2 cm large exophytic hairless nodular tumor with pronounced telangiectasias on the left shin [Figure 1]. The working diagnosis was a basal cell carcinoma or a cutaneous metastasis from colon cancer. The lesion was completely removed surgically. Histological examination confirmed the diagnosis of a dermal cylindroma. Defect closure was realized by a tissue advancement flap.
2. The second patient was a 73-year-old woman, Fitzpatrick skin Type II. She presented with a painless pedunculated tumor with an uneven surface on the lateral face, slow growing for >10 years. The patient had no remarkable medical history. She took no medications.

On examination, we observed a 7 cm large tumor with a mushroom-like growth [Figure 2]. No enlarged lymph nodes were detected. We suspected a keratoacanthoma-like squamous cell carcinoma. The tumor was surgically removed by delayed Mohs surgery. Histological examination disclosed basaloid epithelial cells with rare mitotic figures in a jig-saw pattern. The irregular epithelial isles were surrounded by a thin eosinophilic membrane [Figure 3]. Histopathology confirmed a dermal cylindroma.

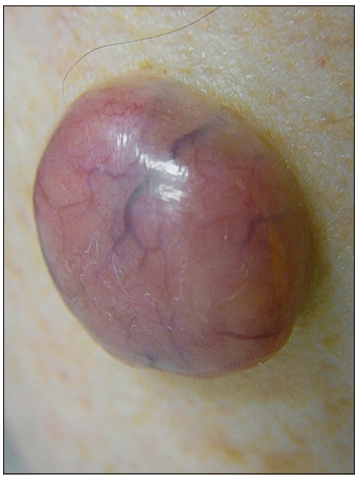

Figure 1: Solitary cylindroma of the lower leg (Case 1), with prominent telangiectasis. (With permission from Wollina U, Haroske G. Kosmet Med. 2012;33 (4):120–1.

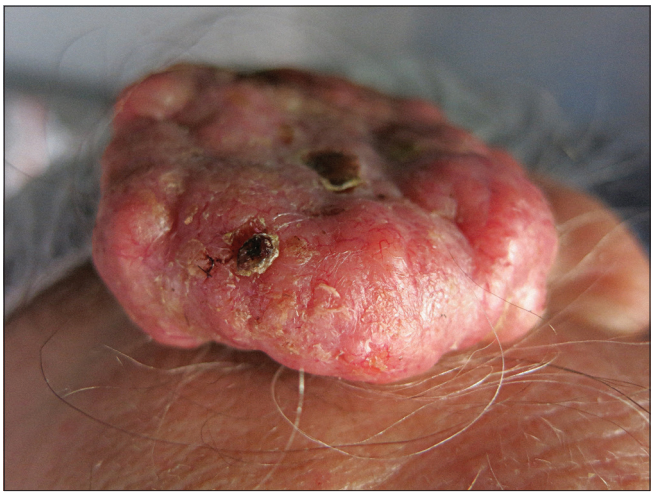

Figure 2: Mushroom-like growth pattern of a pedunculated cylindroma of the lateral face (Case 2). (From Wollina U, Schönlebe J. Open Access Maced J Med Sci. 2018;6 (10):1868-70 with permission).

Defect closure was done by a cheek rotational flap. Details had been published in Wollina and Schönlebe.[10]

3. Case 3 was a 42-year-old Caucasian woman, Fitzpatrick skin Type II, with a painless erythematous flat plaque on her right forehead. Her medical history was unremarkable. The tumor diameter was about 2–4 cm. Dermoscopy revealed telangiectasias [Figure 4]. A basal cell carcinoma was suspected and a diagnostic biopsy was done. Histopathology confirmed a dermal cylindroma and the tumor was removed by delayed Mohs surgery. Defect closure was done by a tissue advancement flap.
4. Case 4 was a 74-year-old Caucasian female, Fitzpatrick skin Type II, with a painless slow-growing tumor of her right ear concha. The tumor diameter was about 3 cm [Figure 5].

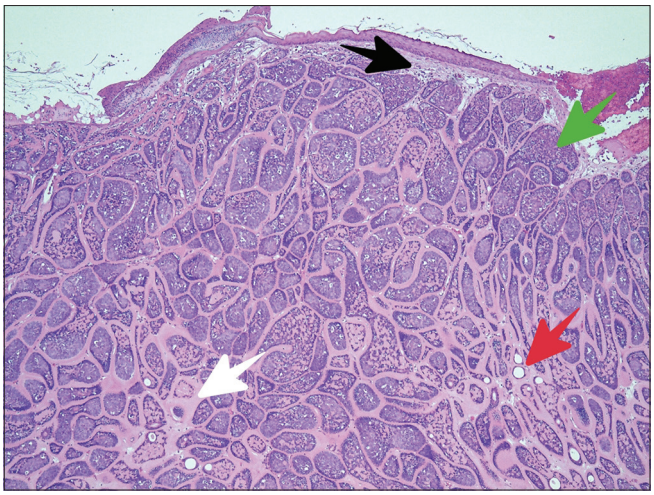

Figure 3: Histopathology of dermal cylindroma (Hematoxylin Eosin, ×40) of case 2. Green arrow: Basaloid tumor isles, White arrow: Hyalinized sheaths, Red arrow: Duct like structures, Black arrow: Free Grenz zone to the overlying epidermis.

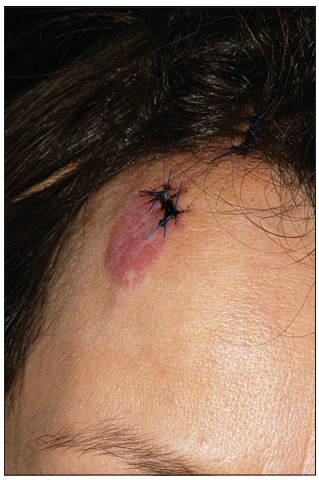

Figure 4: Flat erythematous plaque with telangiectasias on the right forehead (Case 3).

We performed a diagnostic biopsy that confirmed a dermal cylindroma. Complete excision was performed in the ENT department with reconstruction of the concha.

5. Case 5 was 59-yearl-old Caucasian male, Fitzpatrick skin Type II, with multiple painless nodules on the scalp. He had a positive family history for nodular scalp tumors of unknown entity. On examination, we observed >20 erythematous or skin colored firm nodules of variable size, the largest had a diameter of about 4 cm. Some tumors presented an eroded surface [Figure 6].
   We suspected multiple trichilemmomas or BSS. The tumors were surgically removed in general anesthesia. Defect closure was done by a combined approach of rotational flaps and meshed graft transplantation. Histopathology confirmed multiple dermal cylindromas. Genetic analysis detected a *CYLD1* mutation leading to the final diagnosis of FC. There had been no other findings suggesting classical BSS.
6. Case 6 was a 72-year-old Caucasian man, Fitzpatrick skin Type I-III. He suffered for more than 50 years from

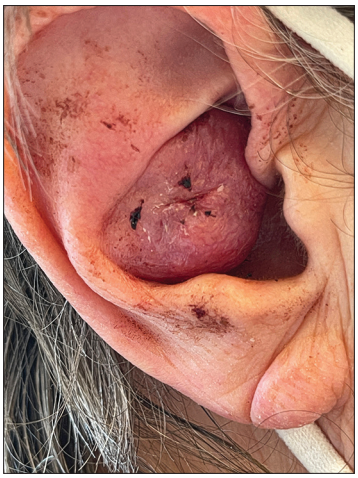

Figure 5: Dermal cylindroma of the ear concha (Case 4).

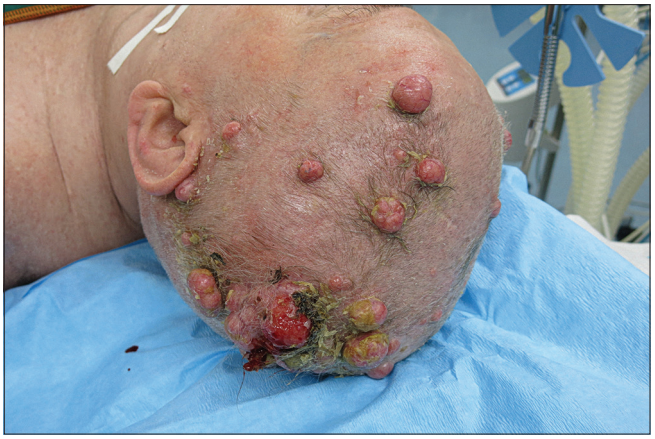

Figure 6: Familial cylindromatosis (Case 5).

psoriasis. On the trunk, multiple skin-colored to reddish papules and nodules were found. Small yellowish papules were seen in the nasolabial fold and the retroauricular fold. Multiple tumors had been surgically removed during the last years including multiple cylindromas and spiradenomas of the scalp and trunk, trichoepitheliomas of the face, shoulder, and trunk. Ten years before, a malignant cylindroma of the scalp and multifocally growing basal cell adenocarcinomas of both parotid glands were diagnosed and treated. In addition, we observed a malignant spiradenocarcinoma on his flank. With multiple cylindromas, salivary gland tumors, spiradenomas, and trichoepitheliomas, he fulfilled the diagnostic criteria for BSS. Details had been published in Köstler *et al*.[11]

Findings of the cases are summarized in [Table 1].

DISCUSSION

Cylindroma occurs only on hair bearing skin. It was originally classified as apocrine tumors due to apocrine secretion patterns on light and electron microscopy.[12,13] Later, immunohistochemical findings suggested an eccrine origin.[14-19] However, recent investigations argued for a possible follicular origin since cylindromas are expressing CD200 that is not found among eccrine tumors.[20]

We report six cases of cylindroma, four sporadic, and two syndromic. Case 5 we classified as FC since the patient was negative for (multiple) spiradenomas or trichoepitheliomas. There was no sign of salivary gland involvement either by basal cell adenoma, membranous-type, or basal cell adenocarcinoma. In the current classification of head-and-neck neoplasia, FC, BSS, and *CYLD* cutaneous syndrome are considered an entity with different names.[21] In contrast, Case 6 was a classical BSS.

Table 1: Summary of presented atypical cases of dermal cylindroma.

Case No.	Age (y)/sex	Remarks
1	71/Male	Nodular tumor with pronounced telangiectasias on the shin
2	73/Female	Pedunculated tumor on the lateral face, mushroom-like growth
3	42/Female	Erythematous flat plaque on the forehead, telangiectasias
4	74/Female	Slow-growing tumor of the ear concha
5	59/Male	Multiple nodules on the scalp, positive family history, *CYLD1* mutation, no other tumor entities suggesting classical BSS
6	72/Male	Multiple tumors of head-and-neck, trunk and arms, classical BSS with multiple cancers of skin parotid glands

BSS: Brooke-spiegler syndrome

Remarkable was the occurrence of multiple malignancies such as malignant cylindroma, basal cell adenocarcinoma of both parotid glands, and spiradenocarcinoma.

The sporadic cases showed an unusual localization or an unusual appearance. In Case 1, we observed a dermal cylindroma of the lower leg. The pronounced telangiectasias were remarkable. The leg is a very rare localization – we found only two other reports.[22,23]

Case 4 presented with a cylindroma of the ear concha. There are sparse reports on the involvement of the ear by cylindroma. Most tumors were from the external auditory canal,[23-30] very rare on tragus[31] or auricle.[32] A few tumors of genital area,[33] the chest,[34] or abdominal skin[35] have been reported as well.

In Cases 2 and 3, the appearance of cylindroma was peculiar. The second case was remarkable because of the size of the tumor and the pedunculated mushroom-like growth pattern.[10] The third case missed the nodular shape of common cylindroma but appeared as a flat plaque with telangiectasias raising the differential diagnosis of basal cell carcinoma.

All tumors could be surgically removed without any complications.

CONCLUSION

Cylindroma is a benign adnexal tumor mostly found in the head-and-neck region that can occur on unusual body parts and with atypical clinical presentation. This unfolds a broad range of possible differential diagnoses, both benign and malignant. Early clinical suspicion and confirmation of diagnosis by histopathology offers the best prognosis.

Statement of human and animal rights

All the procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the 2008 revision of the Declaration of Helsinki of 1975.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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