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Trichilemmal Cyst Nevus with a Sebaceous Nevus Component

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Key Words
Trichilemmal cyst nevus · Sebaceous nevus · Histopathology · Epidermal nevus syndromes · Nevus comedonicus

Abstract
A 23-year-old man with a typical trichilemmal cyst nevus is reported. This recently described disorder is sufficiently characteristic to differentiate it from sebaceous nevus, nevus comedonicus, porokeratotic eccrine nevus, nevus corniculatus, follicular basaloid hamartoma, Munro's nevus and Gardner's syndrome.

Introduction

Epidermal nevi are relatively common and very varied. Sebaceous nevi are the most frequent among them. They mainly occur in the face and on the scalp, but any other body site except for the palms, soles and lips may be affected. In most cases, they are seen in neonates as a hairless plaque. With puberty under the influence of androgens, the nevus may become more yellow and thicker and develop nodules due to the increase in sebaceous glands. Large and systematized sebaceous nevi are arranged along Blaschko's lines and may be part of Schimmelpennig's syndrome. Histologically large mature sebaceous glands – often without associated hair follicles – predominate. Small hair follicles and apocrine glands are often increased in number. Secondary development of other epithelial tumors such as trichoblastoma, syringocystadenoma papilliferum, basal and squamous cell carcinoma is relatively common [1]. We observed a patient who presented with a nevoid lesion clinically reminiscent of sebaceous nevus or nevus comedonicus.

Case Report

A previously healthy 23-year-old man was referred to our outpatient clinic for a right frontoparietal lesion of unclear nature slowly enlarging for about 5 years. He never had any pain. Occasionally, a whitish secretion was noted upon pressure. No similar condition was known in his family. All laboratory examinations were within normal limits. Isotretinoin, up to a maximum daily dose of 60 mg at a body weight of 83 kg, had been given in order to reduce what was thought to be comedones; however, there was no therapeutic response. This prompted us to check and finally revise the diagnosis.

Clinically, a hard, slightly elevated plaque of 12 × 9 cm was seen in the right temporofrontoparietal region (fig. 1). The overlying epidermis appeared normal. Later he developed a smaller lesion on his back in a fountain pattern typical of Blaschko's line. This was in part nodular and had some comedo-like keratin plugs. A biopsy of the dorsal lesion showed small trichilemmal cysts of which one opened to the skin surface (fig. 2a); this was initially misinterpreted as a nevus comedonicus. A biopsy from the forehead lesion showed again multiple trichilemmal cysts with typical epithelial cyst walls without a granular layer and layered keratin, which often retained nuclear remnants at their periphery. Within the cyst keratin, multiple dyskeratotic cells were discernable (fig. 2b). Small foreign-body granulomas were present in the connective tissue between the cysts. In the upper corium, many sebaceous glands both with and without associated hair follicles were present. There were abundant apocrine glands in the mid-dermis. Some cysts opened to the epidermis and displayed small zones of nail-matrix-like epithelium between the cyst opening and the epidermis that gave rise to filiform hyperkeratoses, which retained their nuclei (fig. 2c).

Computed tomography showed a diffusely thickened dermis and subcutis with muscle-equivalent density and moderate enhancement with contrast medium, which reached the tabula externa and could not be clearly delimited from the galea, as well as multiple nodular calcifications. Cortical arrosion was not seen. As the patient did not feel embarrassed by his

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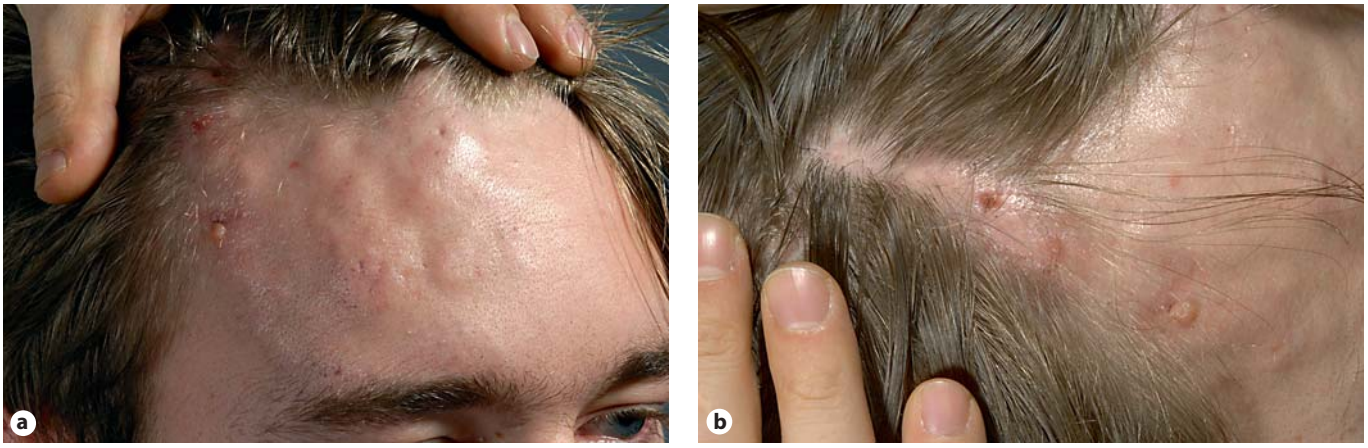

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Fig. 1. a Skin lesions on the right forehead extending into the parietal scalp. **b** Scalp lesion.

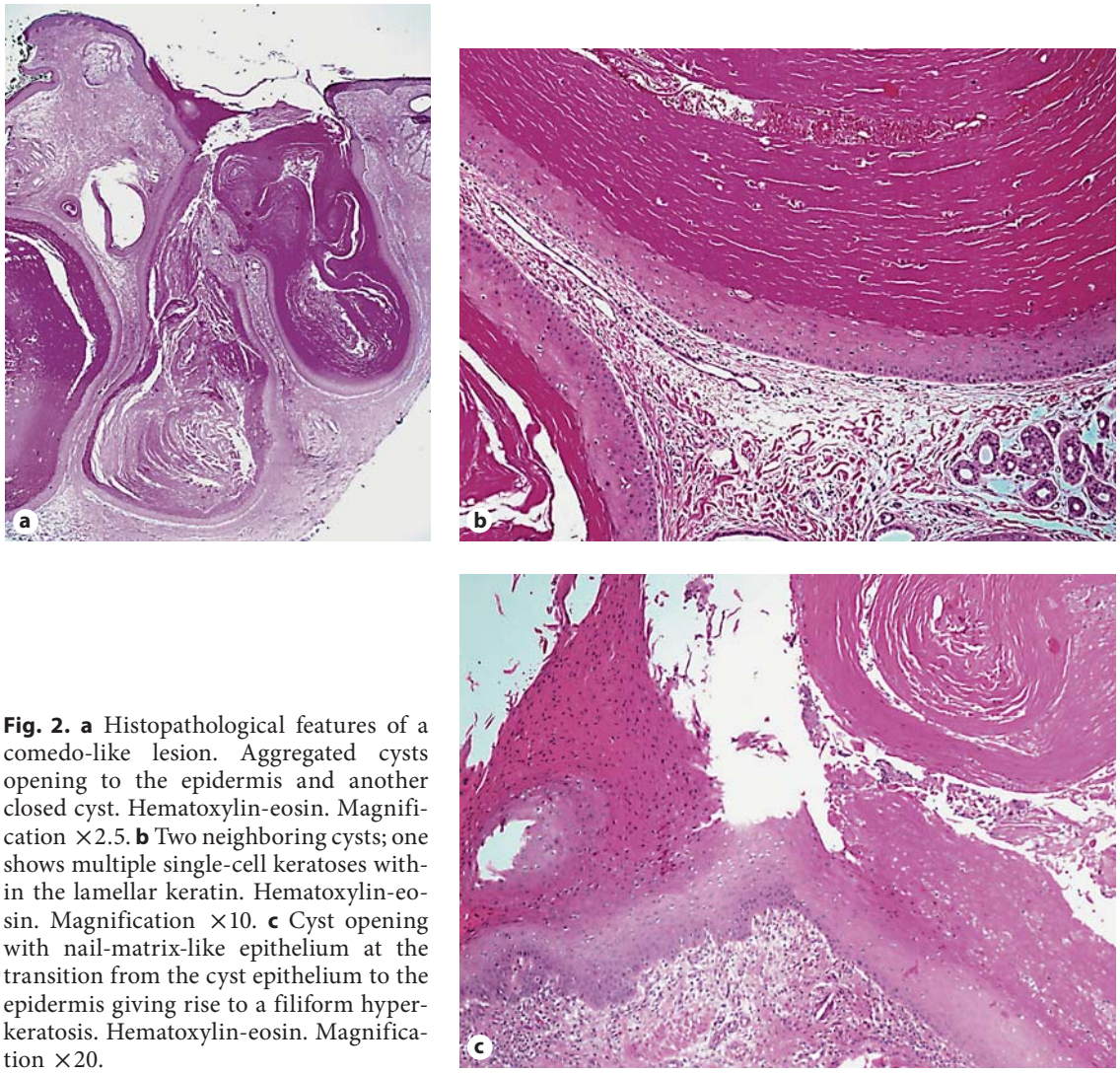

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Fig. 2. a Histopathological features of a comedo-like lesion. Aggregated cysts opening to the epidermis and another closed cyst. Hematoxylin-eosin. Magnification ×2.5. **b** Two neighboring cysts; one shows multiple single-cell keratoses within the lamellar keratin. Hematoxylin-eosin. Magnification ×10. **c** Cyst opening with nail-matrix-like epithelium at the transition from the cyst epithelium to the epidermis giving rise to a filiform hyperkeratosis. Hematoxylin-eosin. Magnification ×20.

lesions, no new treatment was instituted. He was informed about the hypothetical possibility of developing secondary tumors such as basal and squamous cell carcinomas, and a follow-up was agreed upon.

Discussion

Initially, the clinical diagnosis was difficult as there was already the dermatopathological misdiagnosis of a nevus comedonicus. However, after reevaluation of the histological slides, the lack of ordinary comedones was evident as the epidermal invaginations as well as the small cysts all had the structure of trichilemmal cysts. The abundance of sebaceous glands in the upper dermis and of apocrine glands suggested the diagnosis of a sebaceous nevus. A literature research finally revealed another 5 cases very similar to ours [2–5]. Leppard [2] described a 25-year-old patient with unilateral systematized warty lesions since birth that developed from age 10 on lumps that were found to be trichilemmal cysts in histopathology. In additon, there were comedo-like lesions and filiform hyperkeratoses [2, 6, 7]. A case from Spain described an epidermal nevus with multiple small, milia-like trichilemmal cysts [3]. Another case of multiple trichilemmal cysts was even shown to have a mosaicism involving a translocation of chromosomes 1p and 9q [4], the impact of which remained unclear [5]. Multiple trichilemmal cysts in linear arrangement on the scalp were seen in a patient with type 2 segmental glomangiomatosis [6].

Tantcheva-Poor et al. [5] described a case of a 31-year-old woman with an organoid nevus distributed along Blaschko's lines on the face, scalp and right thigh. They stressed the particularity of multiple trichilemmal cysts and coined the term of trichilemmal cyst nevus or nevus trichilemmocysticus [5]. In their case, the condition started with filiform hyperkeratoses and comedo-like lesions in a Blaschko-linear distribution and later developed nodules. The lesions increased in number and size with age. Dermatopathology of our case revealed a numerical increase in sebaceous and apocrine glands typical of a sebaceous nevus and in addition many densely packed small trichilemmal cysts, which dominated the histopathological features, and some filiform hyperkeratoses, but no typical comedones. The lesions were very similar to those of Tantcheva-Poor et al. [5]; however, the cysts in our biopsy were not as large as in their case. The comedo-like lesions in our patient were confined to the back and much less marked. In contrast to that case, a sebaceous nevus component was present in our patient, but the diagnosis of a typical sebaceous nevus had to be discarded. There was no acantholysis. The trichilemmal cysts were smaller. Inflammation and secretion of a whitish substance only occurred after manipulation by the patient. Thus, our case is best described as a trichilemmal cyst nevus with features very similar to, though not identical with, that of Tantcheva-Poor et al. [5].

Tantcheva-Poor et al. [5] also presented a table of differential diagnoses listing trichilemmal cyst nevus, nevus comedonicus, linear follicular basaloid hamartoma, porokeratotic eccrine nevus, nevus corniculatus, Munro's nevus and Gardner's syndrome [5]; however, sebaceous nevus was not considered.

Whether our patient should be followed up like typical sebaceous nevus cases remains to be seen. The prediction of Happle's group that more cases of trichilemmal cyst nevi with less marked involvement will be observed has come true with our observation.

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