Congenital trichofolliculoma: a very rare presentation

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Abstract

Trichofolliculoma is an uncommon hair follicle hamartoma. It usually appears during adulthood on the face or scalp as a single, asymptomatic, skin-colored papule/nodule with small protruding hairs. Histopathological features are diagnostic. Very rare congenital cases have been reported. Herein, we report a congenital trichofolliculoma in a 15-year-old girl.

Keywords: trichofolliculoma, congenital, hair follicle tumors, hamartoma

Introduction

Trichofolliculoma is an uncommon hair follicle hamartoma. It usually appears during adulthood on the face or scalp as an isolated, skin-colored papule or nodule and exhibits a central umbilication from which small hairs may protrude [1]. The lesion may be confused clinically with an epidermal inclusion cyst, dilated pore of Winer, or basal cell carcinoma [2]. Histopathologically it reveals a central dilated follicle, containing keratin and hair shafts with multiple hair follicles in different stages of development [3].

Case Synopsis

A 15-year-old girl presented with an asymptomatic hairy papule on her nose since birth. Examination revealed a skin-colored papule with a central umbilication and small protruding hairs (**Figure 1**). The lesion was excised and histopathological examination demonstrated a central dilated follicular cavity with radially arranged hair follicles in different stages of development consistent with trichofolliculoma (**Figure 2**).

Case Discussion

Trichofolliculoma is a rare hair follicle hamartoma/tumor. It is considered by most authors to be a hamartoma rather than a neoplasm as it has all components of the hair follicle in an aberrant distribution [4]. Its differentiation is midway between a hair follicle nevus and a trichoepithelioma [5]. It usually presents in adulthood as a single lesion on the face especially around the nose. However, it is reported to occur in other sites such as the external auditory meatus, intranasal area, genitalia, lip, and vulva [6]. Rare cases of congenital trichofolliculoma have been reported in the literature [7-11]. There are

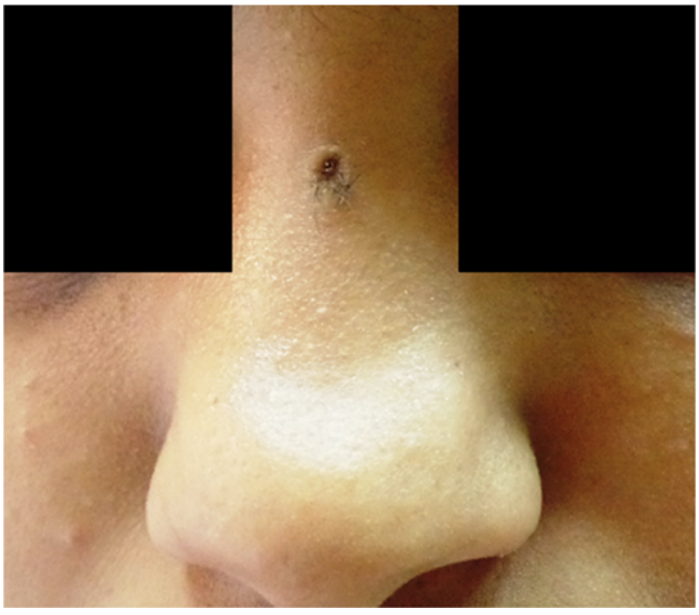

Figure 1: *Trichofolliculoma. A skin-colored papule on the nose with a central umbilication and small protruding hairs.*

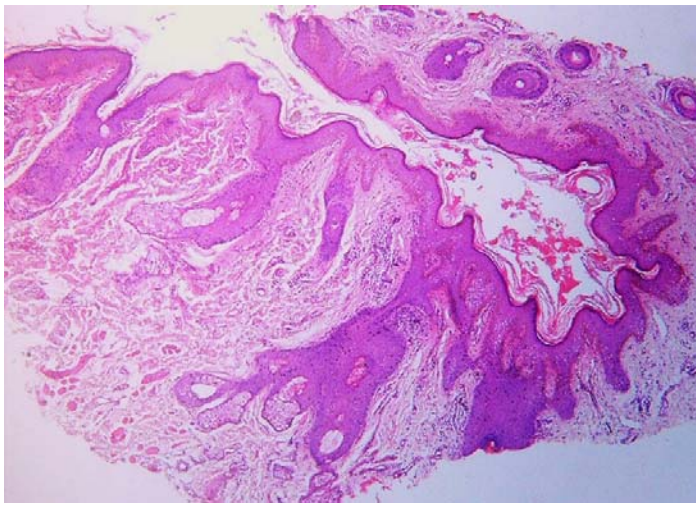

Figure 2: *Histopathology of trichofolliculoma. A central dilated follicular cavity with radially arranged hair follicles in different stages of development (H&E, original magnification X40).*

no reported systemic or skin abnormalities associated with trichofolliculoma [2].

Histopathologically, fully developed trichofolliculoma appears as a well-circumscribed lesion consisting of a central dilated cavity lined by stratified squamous epithelium with an infundibular keratinization pattern that is continuous with the surface epidermis. The cavity usually contains keratin and sometimes vellus hair shafts. Numerous vellus hair follicles, in different stages of development, radiate from the wall of this central cavity [3]. These follicles may in turn give rise to secondary or even tertiary follicles [5]. Rupture of the cystic space can lead to a granulomatous reaction in response to the extruded keratin in the surrounding dermis. Incidentally, trichofolliculomas may show epidermolytic hyperkeratosis or acantholytic dyskeratosis [12]. These findings may vary according to the age of the lesion. Schulz and Hartschuh suggested that trichofolliculoma undergoes changes corresponding to the stages of the normal follicle cycle [13]. In older lesions, catagen and telogen follicles are seen [5]. Merkel cells can be visualized in the outer sheath of the small follicles [14]. Sebaceous differentiation may be seen within the follicles or the rudimentary structures. Sebocytes are seen more often in late stage lesions. A variant of trichofolliculoma in which large sebaceous follicles connect to a central cavity has been reported as a sebaceous trichofolliculoma [15]. It has some similarities to the folliculosebaceous cystic hamartoma (FSCH) which has been argued to be a later stage in the evolution of trichofolliculoma [16].

Misago and his colleagues studied the chronological changes of 40 trichofolliculoma lesions and found that the regressing secondary follicles were not replaced by sebaceous elements. They considered that FSCH is a distinct entity, not a very late stage of trichofolliculoma [17]. Trichofolliculoma with a well-developed stroma may be mistaken for a fibrofolliculoma, the most common skin lesion seen in Birt Hogg Dube syndrome. Fibrofolliculoma is characterized by epithelial cords radiating from the main infundibular wall, not secondary hair follicles as seen in trichofolliculoma [18]. Clinically, the differential diagnosis of trichofolliculoma includes dilated pore of Winer, epidermoid cyst, and basal cell carcinoma [2].

Conclusion

Trichofolliculoma is a benign hamartoma with a nearly stationary course. Excision may be performed for aesthetic purposes and is curative. It has no malignant potential although a single case of trichofolliculoma with perineural invasion has been reported in the literature [19].

Potential conflicts of interest

The authors declare no conflicts of interests/[the following potential conflicts].

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