

Case presentation

Unilateral nevoid trichoepitheliomas on the neck: an unfamiliar presentation

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Abstract

Trichoepitheliomas are epidermal appendageal hamartomas, which usually present as solitary lesions; rarely multiple lesions may be present, mainly involving the centropacial skin symmetrically. We report herein an adolescent male patient with multiple trichoepitheliomas, linearly arranged and dermatomal, present since birth, along the left side of the neck.

Keywords: Trichoepitheliomas, multiple, linear, nevoid

Introduction

Trichoepitheliomas are benign epidermal appendageal tumors, believed to originate from the undifferentiated germinative cells of the pilosebaceous-apocrine unit [1]. Usually solitary, the tumors may rarely be multiple, the latter variety may be inherited in an autosomal dominant pattern. The centropacial region is most commonly affected (80% of cases); other less commonly affected areas include the scalp, neck, and proximal extremities [2]. Rarely, trichoepitheliomas may present as linear papules and plaques in a dermatomal distribution. We present here a case of unilateral, nevoid trichoepitheliomas affecting the neck in an adolescent boy.

Case synopsis

A 14-year-old otherwise healthy boy presented to us with multiple, asymptomatic skin colored papules and nodules, which coalesced to form a linear plaque on the left side of his neck, first noted at birth (Figure 1). The plaque progressively increased in size over the years. Past medical history and family history were non-contributory. Physical examination revealed a non-tender, thick plaque, 8 X 4 cm, on the left side of the neck with a linear configuration, along the C4 dermatome. Detailed examination of the plaque revealed it to be composed of a confluence of multiple, skin colored papules and nodules. Each individual papule was smooth but firm, with no central hairs. The upper part of the plaque had a rough surface. Mucosae, hair, and nails were spared. Histopathological examination revealed a dermal tumor composed of multiple lobules of basaloid cells surrounding a central eosinophilic area, interspersed within a fibro-cellular stroma. Immature follicular differentiation was seen surrounding the lobules (Figure 2). A few horn cysts were scattered around. No connection was seen with the overlying epidermis. The histological appearance was consistent with the diagnosis of trichoepithelioma. We referred the patient to the department of plastic surgery for excision.



Figure 1. Linear, dermatomal plaque.

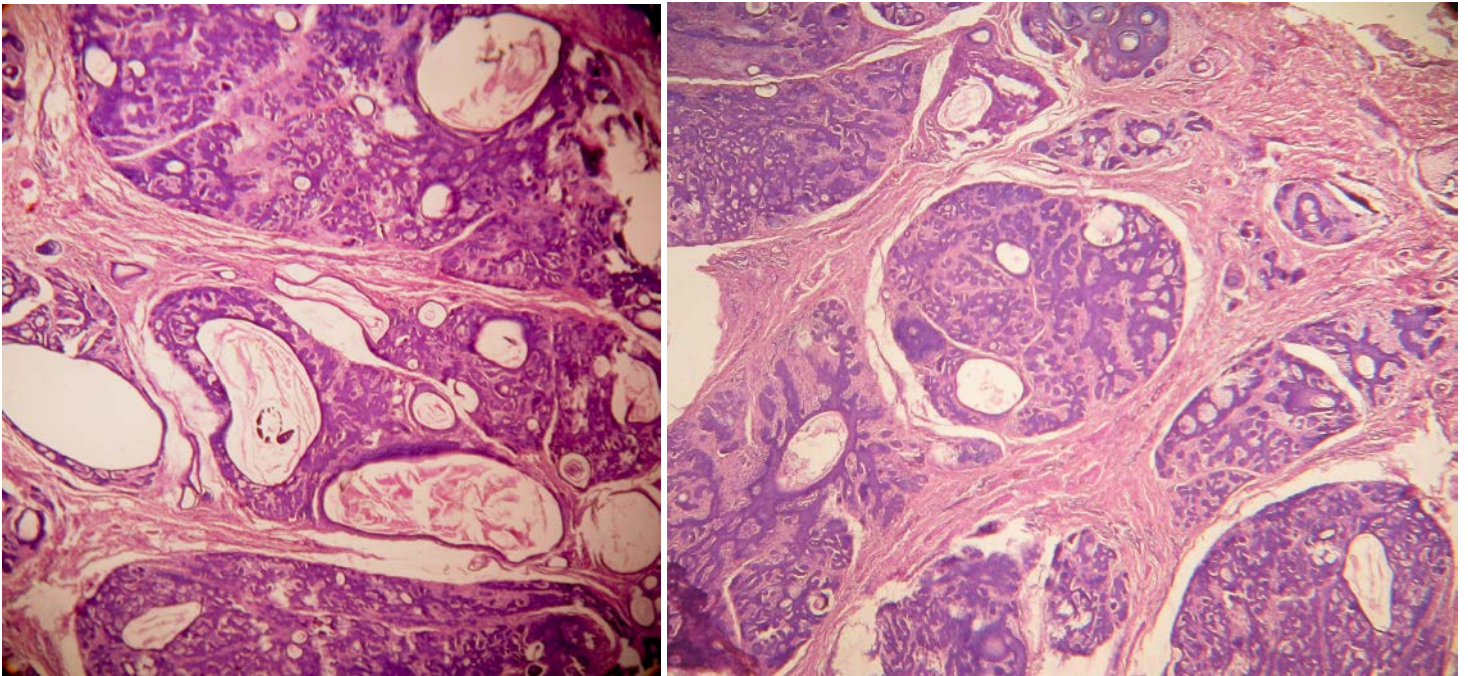


Figure 2 A and B: Basaloid cells surrounding a central eosinophilic area, interspersed within a fibro-cellular stroma .

Discussion

Trichoepitheliomas are benign tumors arising from the undifferentiated pluripotent stem cells of the pilosebaceous-apocrine unit, which can differentiate into follicular or apocrine structures or both [2]. As a result of this remarkable differentiating ability, multiple trichoepitheliomas are often found in association with spiradenomas and /or cylindromas, a condition called Brooke-Spiegler syndrome. Nowadays, trichoepithelioma is regarded as a part of the spectrum of trichoblastoma [1]. Solitary trichoepitheliomas occur much more commonly than multiple trichoepitheliomas. The former occurs as a solitary nodule, usually on the face, generally arising during the second and third decade. The uncommon multiple variant, which has an autosomal dominant mode of inheritance [1], usually appears as small, pearly papules mainly on the centofacial skin. Apart from the face, which is the commonest site for both variants, other sites may be affected, such as the scalp, neck, and proximal extremities [2]. Rarely, multiple trichoepitheliomas may be arranged in the form of linear plaques in a dermatomal distribution; very few such

cases have been reported. There are reports of unilateral multiple trichoepitheliomas arranged in the form of linear plaques in a Blaschkoid distribution, located on the face [3, 4, 5, 6]. In our case also, the multiple trichoepitheliomas were present in a similar linear fashion, but were located on the neck. In all these cases, the lesions were arranged along Blaschko lines, thus representing a nevoid condition. Lambert et al [7] reported a unique case in which a trichoepithelioma was associated with a systematized epidermal nevus. In our case, the lesions were present since birth. Schirren et al [8] also reported a similar case in which linear plaques were present on the neck since birth. However, their case showed histopathological features of both trichoepithelioma and cylindroma (Brooke-Spiegler syndrome).

Although some presentations of trichoepithelioma may be confused with basal cell carcinoma, the presence of stroma surrounding the basaloid lobules containing multiple clefts, with an absence of retraction artifact between tumor cells and stroma helps to differentiate. Tebcherani et al [9] proposed the utility of immunohistochemical markers such as CD 10, cytokeratin 15, cytokeratin 20, and D2-40 to differentiate trichoepithelioma and basal cell carcinoma. Fortunately, the risk of malignant transformation of trichoepithelioma is extremely rare [1]. Sometimes, a wide variety of adnexal tumors may present in a linear fashion, in which case they could be considered in the differential diagnosis. The differentiating points have been tabulated below (Table 1).

Possible therapeutic options for multiple trichoepitheliomas include surgical excision, radiotherapy, dermabrasion, electrodesiccation, and cryosurgery. Recently, successful treatment has been reported with argon laser, erbium-YAG laser [3], and CO2 laser [3, 10]. However, there is chance of scarring with every procedure and the risk-benefit ratio has to be assessed carefully.

In conclusion, unilateral nevoid trichoepitheliomas affecting the neck is an uncommon presentation. The rarity of the case has prompted this report.

Table 1. Adnexal tumors that may present in a linear distribution [2]

Adnexal tumour	Age of onset	Clinical features	Histopathological feature	Treatment	Possible associations
Linear basal cell nevus syndrome	Usually by 3 rd decade	Superficial flesh coloured, red-brown papules or nodules	Focal proliferation of basaloid cells	Surgical excision	Jaw cysts, hypertelorism, skeletal, ocular, CNS defects
Linear comedonal nevus	Childhood to adolescence	Comedones arranged in a linear band	Keratin filled follicular tracts	Keratolytics, Excision for cosmesis	Cataracts
Linear eccrine nevus	Any age	Groups of comedones arranged linearly or in bands, usually unilateral	Keratinous invaginations of epidermis with focal proliferation of cuboidal cells forming glandular structures	Keratolytics, surgical excision	Skeletal, ocular, CNS defects
Multiple linear eccrine spiradenoma	2 nd to 4 th decade	Bluish nodules usually located on the upper part of body. Overlying hypertrichosis may be present	Focal collection of epithelial cells in dermis or subcutis	Surgical excision	May be painful
Linear eccrine poroma	Usually 5 th decade	Pedunculated nodules usually on feet	Collection of small, round basophilic cells in epidermis or upper dermis	Surgical excision	Bowen's disease
Linear syringomas	Usually around puberty	Multiple skin-colored to yellowish firm papules, usually	Cystic structures in dermis; tadpole cells	Excision, Radiosurgery	None

		located on the face (periocular)			
Linear trichoepithelioma (our case)	Childhood to adolescence	Multiple, discrete flesh colored papulo nodules which may coalesce to form a plaque, dermatomal distribution	Focal islands of basaloid cells, horn-cysts, immature follicular differentiation	Surgical excision	None

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