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**Case presentation**

# Linear trichoepithelioma on the neck of a 15-year-old girl

**Amanda J Laska[1], Roberto A Belli[2], Todd T Kobayashi[1]**

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**[1] Department of Dermatology, San Antonio Uniformed Services Health Education Consortium, San Antonio, TX**

**[2] Department of Dermatology, US Army Garrison Yongsan, Korea**

**Correspondence:**

Amanda J Laska
59 MDSP/SGMD
ATTN: Dermatology
2200 Bergquist Dr STE 1
JBSA Lackland TX 78236-9908
Phone: 831-238-6699
Email: AJLaska@gmail.com

## Abstract

Trichoepitheliomas are trichogenic tumors that can have various clinical morphologies. These tumors are benign and differentiate toward the outer root sheath of the hair follicle. Solitary trichoepitheliomas arise sporadically, in contrast to multiple trichoepitheliomas, which are usually inherited as an autosomal dominant trait or as part of various genetic syndromes. We report a case of an adolescent female with a linear array of trichoepitheliomas on her left neck.

## Introduction

Trichoepitheliomas, although benign tumors, can have a striking clinical presentation. Cases have been described of trichoepitheliomas with unusual morphology, to include linear, plaque-like, and giant. Several well-documented genodermatoses are associated with the development of multiple trichoepitheliomas, but these tumors can also be seen in otherwise normal patients. Herein we describe a healthy adolescent who had a linear array of trichoepitheliomas on the left side of the neck.

## Case synopsis

A 15-year-old girl was referred to our clinic with the complaint of several lesions on the left neck that were asymptomatic. Per her mother, these lesions had been present since early childhood and had grown proportionally with the child. The patient was otherwise healthy, denying neurologic, cardiac, or musculoskeletal complaints.

Five skin-colored to yellow-pink papules arranged in a linear pattern were noted on the left neck and inferior postauricular region (Figures 1). Inspection of the remainder of the skin was negative for other findings.

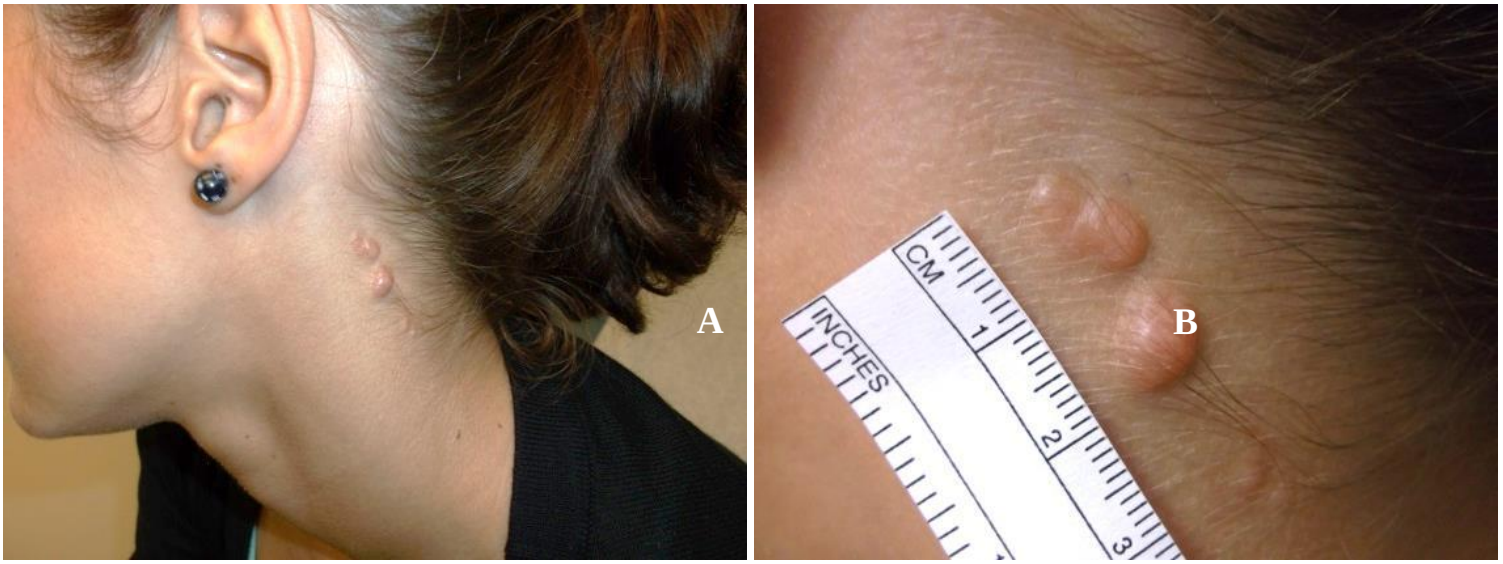


**Figure 1**. (A), Linear array of pink papules on the neck. (B), Pink 3-7 mm papules.

Shave biopsies of two papules showed a normal overlying epidermis with anastomosing strands of uniform basaloid cells in the dermis forming abortive follicular structures (Figures 2, 3). No tumor-stromal clefting was noted. The surrounding stroma was hypercellular and fibrotic with notable papillary mesenchymal bodies.

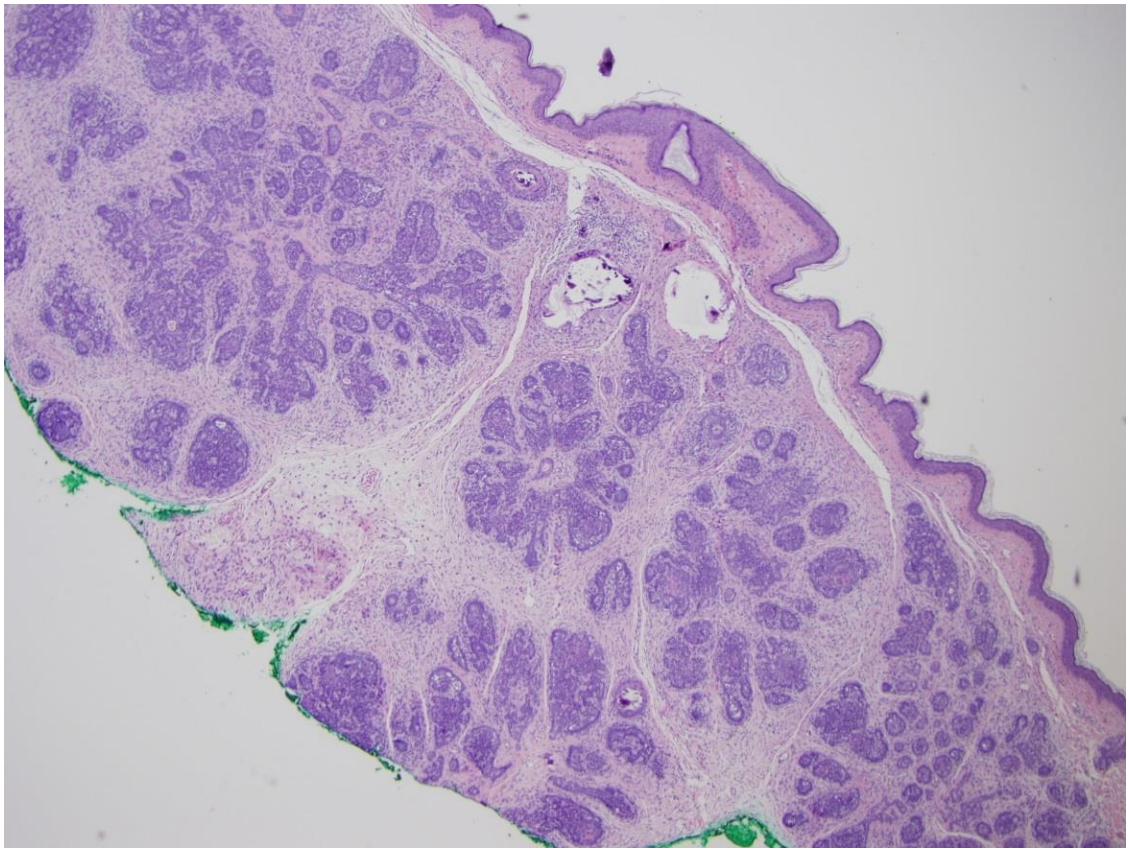

**Figure 2**. Basaloid cells in dermis forming abortive follicular structures. H&E, 10x.

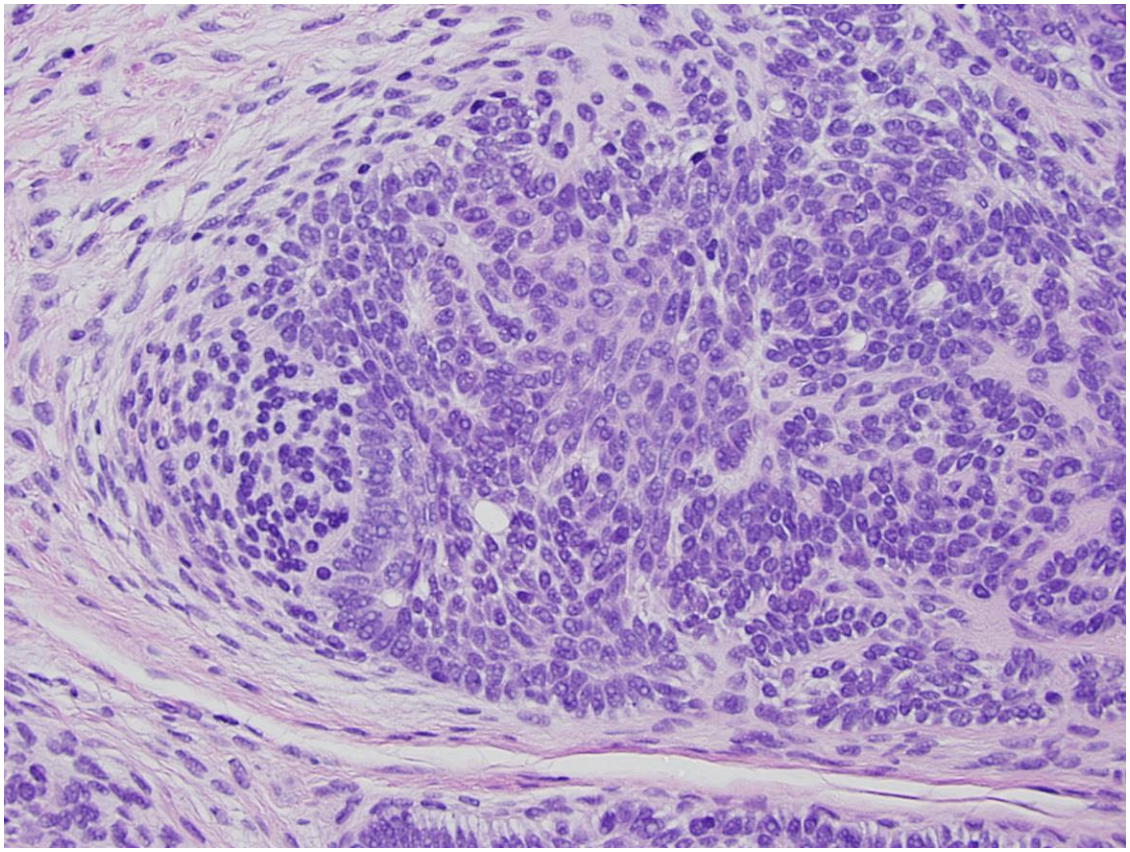

**Figure 3**. Uniform basaloid cells within hypercellular stroma. H&E, 40x.

## Discussion

Trichoepitheliomas exist in both solitary and multiple forms. Solitary trichoepitheliomas are more common than multiple trichoepitheliomas and typically arise in the fifth decade. They are considered small if less than 2 centimeters in diameter or giant if greater than 2 centimeters. Reports of giant solitary trichoepitheliomas comment that they are usually pedunculated, up to several centimeters in diameter, and found on the head, perianal region, and thighs of adults [1]. They arise in adults in the 7th decade and must be differentiated from basal cell carcinoma. Surgical excision is the treatment of choice, however local recurrence and transformation into basal cell carcinoma may occur [2].

Patients with multiple trichoepitheliomas usually develop them in childhood or adolescence [3]. They appear as small skin-colored papules distributed symmetrically along the nasolabial folds and central face most commonly, but involvement of the scalp, neck, and proximal extremities may also be noted. Gene defects in the *CYLD* gene, which encodes an enzyme with deubiquitinase activity, is found in three inherited syndromes, two of which feature multiple trichoepitheliomas [4]. Brooke-Spiegler syndrome is characterized by multiple trichoepitheliomas, spiradenomas, and cylindromas and multiple familial trichoepithelioma 1 is considered a subset of Brooke-Spiegler syndrome and features multiple trichoepitheliomas. The third syndrome, familial cylindromatosis, is also related to a *CYLD* mutation. Linear trichoepitheliomas have only been reported a few times in the literature and are usually found in otherwise healthy patients. Most of the reported cases in the literature discussed patients of Fitzpatrick skin type IV-VI with lesions involving the face. du Toit et al. reported a South African patient with a linear trichoepithelioma on the nasal dorsum and Chang reported an African-American female with unilateral configuration of trichoepitheliomas on the left nasolabial fold and left cheek following the lines of Blaschko [1, 5]. Geffner et al. reported a 10 year old African American female who presented with extensive linear grouped papules on the left shoulder, left anterior and posterior trunk, and left lower extremity that were found to be trichoepitheliomas [6]. Strauss reported a unilateral trichoepithelioma on the face of a white female, which at that time was the first report of a linear trichoepithelioma in a Caucasian patient [7]. Oh et al. discussed a large unilateral facial plaque on the face of an Asian boy involving the nasal dorsum, left nasal sidewall and forehead that represented coalescent trichoepitheliomas [8]. Like our patient, all these patients were otherwise healthy and lacked stigmata of a genetically inherited disorders.

It is believed that trichoepitheliomas arise from the primary epithelial tissue and differentiate toward the outer root sheath of the hair follicle. On histopathology, they are characterized by basaloid proliferations in the dermis surrounded by a hypercellular fibromyxoid stroma that shows interstromal clefting. Papillary mesenchymal bodies represent abortive hair follicles and horned cysts consist of a keratinized core surrounded by basophilic cells. Treatment of trichoepitheliomas is difficult and requires

destructive modalities. Surgical excision, curettage and electrodessication have been used successfully. The ablative carbon dioxide laser is useful for patients with multiple trichoepitheliomas [9]. Imiquimod and retinoic acid were shown in a study by Alessi et al. to have 80% efficacy and this is a viable option for patients desiring medical treatment [10]. Our patient declined further treatment.

In summary, this patient displays a linear array of trichoepitheliomas on the left neck in the absence of other features in a Caucasian female. This presentation is unique as many of the reported patients with linear trichoepitheliomas have been of African-American or Asian ancestry. Also, our patient did not have facial involvement. According to our literature search, this is the second reported case of a linear trichoepithelioma in a Caucasian patient, and the first of a linear trichoepithelioma in a Caucasian patient not involving the face. This report can be added to the literature as a rare presentation of trichoepithelioma.

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