Volume 22 Number 11
November 2016

Photo vignette

Linear syringocystadenoma papilliferum on the retroauricular area associated with nevus sebaceus

Algun Polat Ekinci[1], Nesimi Buyukbabani[2], Lale Mehdi[3], K. Didem Yazganoglu[4], Can Baykal[5]

Dermatology Online Journal 22 (11): 14

[1]Specialist of Dermatology and Venereology, Istanbul University, Istanbul Medical Faculty, Istanbul, Turkey

[2]Professor of Pathology, Istanbul University, Istanbul Medical Faculty, Istanbul, Turkey

[3]Resident of Dermatology and Venereology, Istanbul University, Istanbul Medical Faculty, Istanbul, Turkey

[4]Associate Professor of Dermatology and Venereology, Istanbul University, Istanbul Medical Faculty, Istanbul, Turkey

[5]Professor of Dermatology and Venereology, Istanbul University, Istanbul Medical Faculty, Istanbul, Turkey

Correspondence:

Algun Polat Ekinci, MD
Istanbul Universitesi
Istanbul Tıp Fakültesi Deri ve Zührevi Hastalıklar Anabilim Dalı
34093 Capa-Istanbul / Turkey
Tel. +90 2126352939 Fax +90 2126353107
Email: algunekinci@yahoo.com

Abstract

Syringocystadenoma papilliferum is a rare cutaneous adnexal tumor that usually arises in the head and neck region. It may develop *de novo* or within a nevus sebaceus. Linear syringocystadenoma papilliferum is an uncommon variant of this benign tumor. We report a child with linear retroauricular distribution of syringocystadenoma papilliferum. A background nevus sebaceus was shown histologically. Total excision was curative with no recurrence. An association between the linear variant of syringocystadenoma papilliferum and nevus sebaceus has not been reported previously.

Keywords: syringocystadenoma papilliferum, linear syringocystadenoma papilliferum, nevus sebaceus, retroauricular

Introduction

Syringocystadenoma papilliferum (SCAP) is a rare cutaneous tumor prominently located on the head and neck region [1-3]. Linear SCAP is a more rare form of this benign tumor [1, 2, 4, 5]. Although SCAP may frequently emerge from nevus sebaceus (NS) [4], intriguingly, linear SCAP associated with NS has not been previously reported.

Case synopsis

A 12-year-old boy presented with an elevated asymptomatic plaque on the back of the left ear. The plaque had been present since early childhood and increased gradually in size over the years. Dermatological examination revealed a linear plaque of approximately 4 cm on the retroauricular region and extending to the scalp. The growth consisted of multiple, skin-colored verrucous papules and nodules coalescing into a linear plaque and showing erosion on some sites (Figure 1).

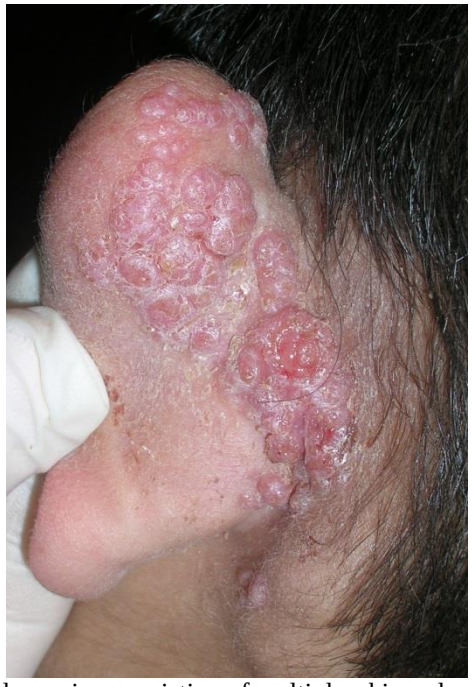

Figure 1. A linear plaque on the retroauricular region consisting of multiple, skin-colored to pinkish verrucous papules and nodules coalescing into a linear plaque and showing erosion on some sites.

Two subcutaneous, solid, immobile lesions on the postauricular region were evaluated by ultrasonography and the diagnosis of reactive lymphadenopathy was suggested. There were no other remarkable cutaneous or systemic findings. A punch biopsy revealed epidermal hyperkeratosis and parakeratosis associated with proliferation of structures composed of papillary projections lined by two rows of cells: an outer layer of cuboidal cells and an inner (luminal) layer of columnar cells with some evidence of decapitation secretion (Figure 2a). This was consistent with a diagnosis of syringocystadenoma papilliferum (SCAP). The lesion was excised completely. Histopathological analysis of excision material showed features of SCAP presenting as multiple foci (Figure 2) and a background lesion, which had features compatible with NS. Findings included epidermal verrucous projections, accompanied by abnormal sebaceous and sweat glands in the dermis (Figure 2b). There was no recurrence at the 1-year follow-up examination.

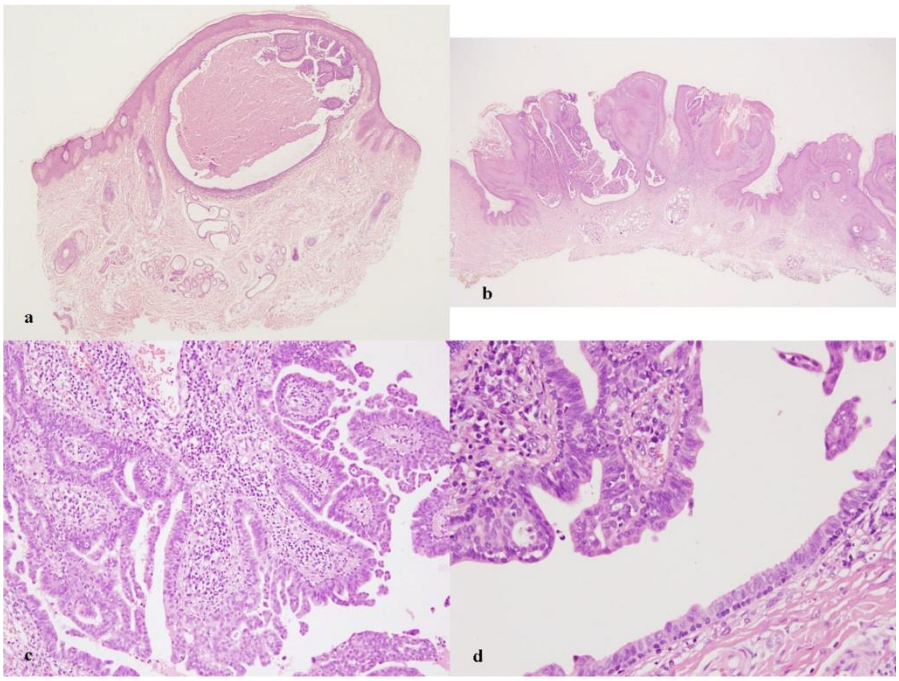

Figure 2. a) Punch biopsy shows papillary projections to the small cyst lumen suggesting a diagnosis of syringocystadenoma papilliferum (H&E, 40x); **b**) Panoramic view of excision shows background nevus sebaceus, characterized by broad papillomatous epidermal projections and abnormal sweat glands in the upper dermis. Papillary projections of syringocystadenoma papilliferum are seen on the center-left. On the left end of the specimen perilesional normal skin can be seen for comparison (H&E, 20x); **c)** Papillary projections and prominent stromal inflammatory infiltrate (H&E, 200x); **d**) Double layered surface and covering apocrine epithelium, plasma cell rich stromal infiltrate, typical of syringocystadenoma papilliferum (H&E, 400x).

Discussion

SCAP is a rare benign adnexal tumor usually presenting at birth or early childhood. It may become more prominent during and after puberty [1-4]. It is more commonly located on the head and neck area [1-3]. The pinna is reported to be one of the sites of

involvement [3]. It may emerge either from normal skin or from an existing NS [1, 3, 4]. The linear form is very rare and may also be seen on anatomical sites other than the head and neck [1, 4, 5]. The clinical picture of linear SCAP consists of multiple linearly arranged, skin-colored to pinkish papules and/or nodules, which may have smooth, hyperkeratotic, verrucous, eroded, or ulcerated surfaces. Individual papules and nodules may be separate or coalescent, sometimes causing a conglomerated appearance [1, 2, 4, 5]. Linear tumors may be very large, reaching lengths up to 20 cm [5]. Interestingly, an association between linear SCAP and NS has not been reported previously. Unlike previous reports, the linear tumor in our patient was shown to coexist with NS. Moreover, our patient showed an unusual location for a linear variant of SCAP.

Clinically, differential diagnoses of linear verrucous lesions in childhood mainly include hamartomatous diseases, such as NS, epidermal nevus, nevus comedonicus, basaloid follicular hamartoma, and viral warts [1, 4]. NS is more common on the scalp, but may also be located on the retroauricular area. Round or oval lesions are more typical, but linear lesions may also be observed. Interestingly, the clinical presentation of the tumor in our case did not clearly favor NS and this was identified only on the background of the SCAP after histopathological examination.

Conclusion

This case report describes the rare linear morphology of SCAP located on the retroauricular region, a previously undescribed location. The coexistence of linear SCAP and NS has also not been previously reported. Awareness of the linear variant of SCAP among other linearly arranged verrucous lesions in childhood may be of critical importance with regard to clinical management.

References

1. Ghosh SK, Mandal RK, Bandyopadhyay D, Mukhopadhyay SM. Adult-onset linear syringocystadenoma papilliferum over the inguinal fold: a case report with emphasis on mast cell staining pattern. Dermatol Online J. 2012 Nov 15;18(11):16. [PMID: 23217957]
2. Martorell-Calatayud A, Sanz-Motilva V, Garcia-Sales MA, Calatayud-Blas A. Linear syringocystadenoma papilliferum: an uncommon event with a favorable prognosis. Dermatol Online J. 2011 Aug 15;17(8):5. [PMID: 21906485]
3. Bakshi J, Parida PK. Syringocystadenoma papilliferum: report of first case on the pinna. J Laryngol Otol. 2006 Dec;120(12):e45. Epub 2006 Nov 14. [PMID: 17105677]
4. Chauhan A, Gupta L, Gautam RK, Bhardwaj M, Gopichandani K. Linear syringocystadenoma papilliferum: a case report with review of literature. Indian J Dermatol. 2013 Sep;58(5):409. [PMID: 24082219]
5. Kar M, Kar JK, Maiti S. Giant linear syringocystadenoma papilliferum of the back. Indian J Dermatol Venereol Leprol. 2012 Jan-Feb;78(1):123. [PMID: 22199089]