

# Dermoscopy of a rare case of linear syringocystadenoma papilliferum with review of the literature

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**Key words:** linear syringocystadenoma papilliferum, adnexal tumor, dermoscopy

**Citation:** Chauhan P, Kumar Chauhan R, Upadhyaya A, Kishore S. Dermoscopy of a rare case of linear syringocystadenoma papilliferum with review of the literature. *Dermatol Pract Concept*. 2018;8(1):33-38. DOI: <https://doi.org/10.5826/dpc.0801a07>

**Received:** July 22, 2017; **Accepted:** November 7, 2017; **Published:** January 31, 2018

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**Funding:** None.

**Competing interests:** The authors have no conflicts of interest to disclose.

All authors have contributed significantly to this publication.

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**ABSTRACT** Syringocystadenoma papilliferum (SCAP) is a benign hamartomatous tumor arising from pluripotent cells with either apocrine or eccrine differentiation. We report a rare case of de novo linear SCAP in a 12-year-old female child with lesions over the chest along with the dermoscopic findings.

## Introduction

Syringocystadenoma papilliferum (SCAP) is a rare benign hamartomatous adnexal tumor that originates from the apocrine or the eccrine sweat glands [1]. It is a relatively rare neoplasm presenting at birth in 50% of the cases. In around 15-30% of the cases, it develops around puberty [2]. The lesions evolve either de novo or from a preexisting nevus sebaceous. There are three recognized clinical forms of it i.e., plaque, solitary nodular, and linear. Most of the lesions localize over the head and neck region [3]. We hereby present a rare case of de novo congenital linear SCAP over the chest. We also describe the dermoscopic findings observed in our case.

## Case Presentation

A 12-year-old girl presented with slowly growing multiple, itchy, red and raised lesions over her chest just below the

left clavicle which were present since birth. The lesions had significantly increased in size and number in the previous two years. Serous non-foul-smelling discharge from the lesions was occasionally noticed. Associated symptoms suggestive of neurological, ocular or skeletal abnormality were not reported. On clinical examination, multiple grouped dome-shaped papules and nodules with central umbilication were noted to be present in a linear array over the upper part of chest. The surface of the lesions showed central ulceration and crusting (Figure 1).

Dermoscopic examination with a DermLite II Hybrid M (3Gen, San Juan Capistrano, CA, USA; 10× magnification) was performed which revealed milky red papillomatous projections with a central ulceration. At places, white circles were seen over the rim of the milky red areas. Polymorphic vessels were seen within the darker ulcerated areas. Yellowish areas within the ulceration may represent sites of secretion (Figure 2).



**Figure 1.** multiple grouped dome shaped papules and nodules with central umbilication present over the chest just below the left clavicle. [Copyright: ©2018 Chauhan et al.]

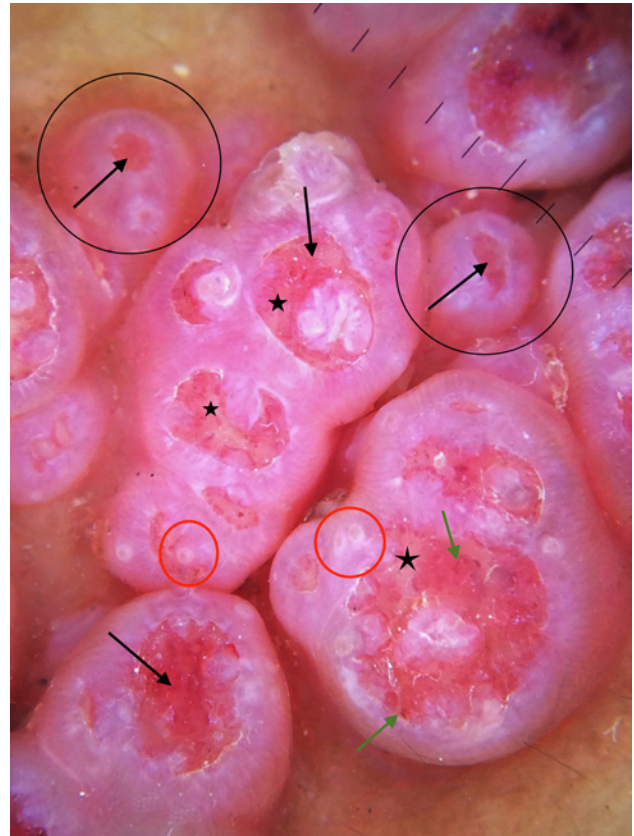
Histopathological examination of the umbilicated papule was done and showed an endophytic tumor with several papillary infoldings lined by glandular epithelium, which consisted of two rows of cells. The inner row had high columnar cells with oval nuclei and faint eosinophilic cytoplasm. The outer row was comprised of small cuboidal cells with round nuclei and scant cytoplasm. Focal areas of decapitation secretion were noted. The core of papillae had many plasma cells, lymphocytes and few polymorphic cells (Figure 3).

The diagnosis of linear syringocystadenoma papilliferum was made on the basis of clinical, dermoscopic and histopathologic examinations. The patient was referred to the department of pediatric surgery for surgical excision.

## Discussion

Syringocystadenoma papilliferum (SCAP) is a rare benign cutaneous hamartoma seen to arise from the pluripotent cells with the potential to exhibit either apocrine or eccrine lineage, although apocrine differentiation is more common. It usually involves the head and neck area, taking its origin either de novo or from a preexisting nevus sebaceous in 30% of the cases [1]. The unusual anatomical sites of involvement include arms, breast, eyelids, axilla, scrotum, and inguinal and perineal regions [3].

Three clinical types of SCAP have been described: a) **Plaque type:** presenting as an alopecic patch on the scalp which may enlarge during puberty to become nodular, verrucous or crusted. b) **Linear type:** consisting of multiple reddish pink firm papules or umbilicated nodules 1-10 mm in size commonly occurring over face and neck. c) **Solitary nodular type:** which are domed pedunculated nodules 5-10 mm in size with a predilection for the trunk, shoulder, and axillae [4]. The linear presentation of SCAP is extremely rare with less than 20 cases reported in the literature [5-21] (Table 1).



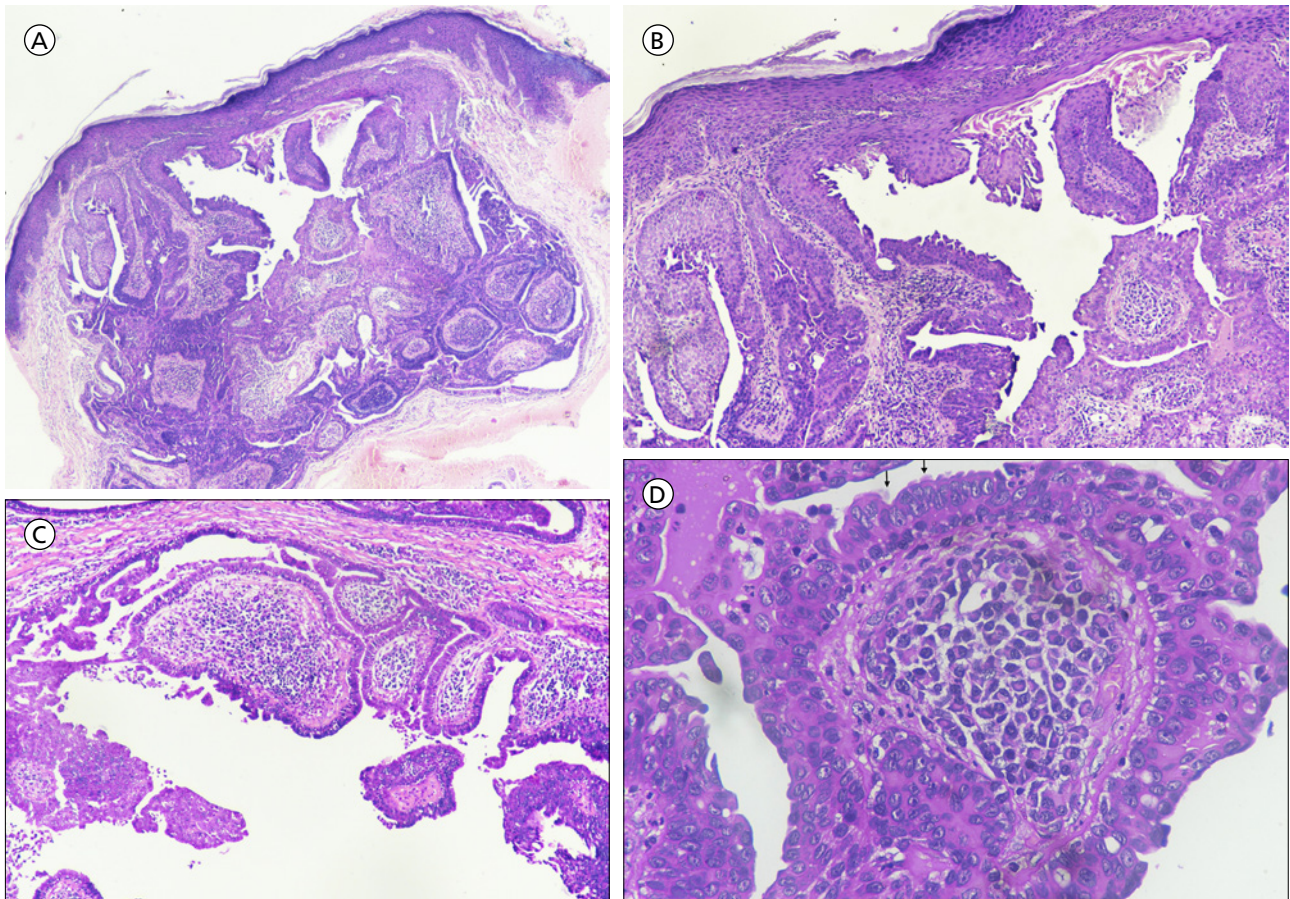
**Figure 2.** Milky red papillomatous projections (black circle) with central ulceration (black arrow). White circles (red circle) over the milky red rim. Polymorphic vessels (green arrows) were also seen. [Copyright: ©2018 Chauhan et al.]

Thirteen cases of linear SCAP were females and the majority of patients had onset at birth or in early childhood. Only two cases of late-onset linear SCAP have been reported; one at 19 years and another 21 years old. Surprisingly, the majority of the cases arose de novo. Only three cases of linear SCAP have been reported to be associated with nevus sebaceous [21], tubular apocrine adenoma [8], and apocrine cystadenoma and hidrocystoma [10]. Hitherto, only three cases of linear SCAP over the chest have been described [6,8,20]. Of these, one case of linear SCAP in a 12-year-old male was associated with tubular apocrine adenoma[8]. The remaining two were cases of de novo linear SCAP over the left side of the chest in one [6] and over the left breast in the second case [20]. To the best of our knowledge, our patient represents the third case of de novo linear SCAP over chest.

All the published cases of linear SCAP were histopathologically proven. However, there is a lack of documentation of dermoscopic findings in linear SCAP, as none has been previously reported.

We also highlight the dermoscopic features of SCAP as the actual potential of dermoscopy has not been tapped in the diagnosis of adnexal tumors. In 2011, Bruno et al. [22], described the dermoscopic features in SCAP associated with nevus sebaceous for the first time. The authors described a





**Figure 3.** (a) Scanner view showing an endophytic tumor with several papillary infoldings in the upper dermis (H&E, x40). (b) Histopathological examination showing invagination of surface epithelium and irregular papillary projections protruding into the lumen (H&E, x100). (c) Several papillary infoldings lined by glandular epithelium consisting of two rows of cells with inner row of high columnar cells having oval nuclei and faint eosinophilic cytoplasm and outer row having small cuboidal cells with round nuclei and scanty cytoplasm. (d) Higher magnification showing core of papillae having many plasma cells, lymphocytes and few polymorphic cells. Decapitation secretion is appreciated in the luminal layer (black arrow) (H&E, x400). [Copyright: ©2018 Chauhan et al.]

polymorphous vascular pattern comprised of irregular linear and glomerular vessels, some of which were surrounded by a whitish halo and others grouped in a horseshoe arrangement on a pinkish-white background. The dermoscopic findings in our case are similar and corroborate the earlier findings of Bruno et al. [22] To the best of our knowledge, ours is the first case describing dermoscopic findings of de novo linear SCAP.

Dermoscopy findings in seven cases of SCAP associated with nevus sebaceous located in the head and neck regions documented by Zaballos et al. [23] are noteworthy. The authors noted that the most common dermoscopic pattern associated with their SCAP cases was a symmetric erythematous lesion with “exophytic papillary structures,” followed by a central depression, ulceration and vessels (hairpin vessels, polymorphous vessels and comma vessels). Similarly, Dumen et al., [24] have also documented dermoscopic features in a case of SCAP with nevus sebaceous. The dermoscopic examination in the aforementioned case revealed central yellowish-white discoloration, polymorphic vessels including irregular dotted, hairpin-like, glomerular and linear vessels with a

surrounding pinkish-white rim and peripheral hairpin like vessels. These findings are also similar to that seen in our case.

The dermoscopic morphology of the other adnexal tumors of apocrine origin needs to be elucidated. Recently, dermoscopic features of 22 cases of apocrine hidrocystoma were reported [25]. A homogenous area that occupies the whole lesion with arborizing vessels was found to be the most common dermoscopic pattern in apocrine hidrocystoma. Similarly, tubular apocrine adenoma (TAA) is another adnexal tumor of apocrine origin whose dermoscopy is hitherto unclear. Ito et al, [26] noted coexistence of short fine telangiectasias and large blue-gray ovoid nests arranged in a floriform pattern to be the specific dermoscopic finding in TAA.

The present report aims to emphasize that the dermoscopic examination can act as an extremely valuable, noninvasive and inexpensive tool in the diagnosis of SCAP especially in children when a traumatic procedure like a biopsy can raise the apprehension and discomfort of the child and the parents. However, further studies are needed to corroborate the dermoscopic findings seen in our case. Moreover,

**TABLE 1. Summary of the histopathologically proven cases of linear SCAP**

Age /Sex	Age at Onset	Symptom	Location of the Lesions	Clinical Morphology	Association	Author, Year of Publication	Ref
10, F	Since birth	-	Upper part of the left arm	Multiple yellowish-red papules, with central umbilication in several of the papules	-	Rostan et al, 1976	5
16, F	Since birth	-	Left side of chest	Skin-colored, firm papules and nodules. Few were ulcerated discharging a serosanguinous material	-	Premalatha et al, 1985	6
2, M	Since birth	-	Posterior neck below hairline	7 waxy erythematous oval papules	-	Goldberg et al, 1985	7
12, M	Since birth	Occasionally mildly pruritic	Upper chest	Grouped shiny erythematous papules focally coalescing into plaques	TAA	Epstein et al, 1990	8
11, F	Since birth	-	Right thigh	4 plaques consisting of a dozen pea-sized pink papules and small nodules with crusted surface. Central umbilication in some.	-	de Bliiek et al, 1999	9
14, F	Early childhood	Pruritus	Left inner thigh	6 discrete, erythematous papules	Apocrine cystadenoma, hidrocystoma	Patterson et al, 2001	10
20, F	Since birth	Sudden growth in lesion, recent onset of pain	Nape of neck	Skin-colored and erythematous warty papules arranged vertically	-	Dawn et al, 2002	11
5, F	6 months of age	-	Scalp	Grouped skin-colored, umbilicated papules along with cauliflower-like moist reddish alopecia plaque	-	Laxmisha et al, 2007	12
51, M	early childhood	-	Nape of neck	3 ulcerated nodules (2-3 cm) small, skin-colored and yellow papules in close proximity to the nodules	-	Narang et al, 2008	13
19, F	At birth	-	Extensor site of proximal part of right upper extremity	Multiple discrete, erythematous, 0.5-1 cm sized pseudovesicular papules	-	Gönül et al, 2008	14
20, F	At 19 years	Pruritus, history of bleeding	Left-sided occipital part of the scalp extending to the nape of neck	Highly elevated, moist, fetid, vegetated and pinkish lesion	-	Yaghoobi et al, 2009	15

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**TABLE 1. Summary of the histopathologically proven cases of linear SCAP (continued)**

Age /Sex	Age at Onset	Symptom	Location of the Lesions	Clinical Morphology	Association	Author, Year of Publication	Ref
40, F	Since birth	-	Right lower abdomen	Multiple erythematous papules and nodules with erosion on the surface of some of the lesions	-	Yap et al, 2010	16
18, F	Early childhood	-	Right inguinal & pubic region	3 large fleshy erythematous, exuberant verrucous plaques with adherent whitish slough Skin-colored, discrete papules with slight umbilication	-	Pahwa et al, 2011	3
15, F	Early childhood	Occasionally pruritus	Left side of neck	20 discrete erythematous vesicle-like papules	-	Martorell-Calatayu et al, 2011	17
12, M	Since birth	-	Lower back	Fleshy cauliflower-like erythematous papulonodular lesion with increase in vascularity and oozing of serosanguinous fluid	-	Kar et al, 2012	18
36, M	21 years of age	-	Right inguinal fold	Multiple skin-colored, dome-shaped, firm, non-tender nodules	-	Ghosh et al, 2012	19
10, F	2 months	-	Right lower abdomen	Multiple erythematous papules, coalescent plaques	-	Chauhan et al, 2013	1
35, F	Since 8 months	Mild irritation	Left breast	2 erythematous, multilobular exuberant plaques	-	Bandopadhyay, 2014	20
12, M	Since birth	-	Back of the left ear extending to scalp	Multiple, skin-colored verrucous papules and nodules, showing erosion on some sites	NS	Ekinci et al, 2016	21
12, F	Since birth	-	Over chest just below left clavicle	Multiple grouped domeshaped papules and nodules with central umbilication, ulceration and crusting	-	Present case	

Abbreviations: F, female; M, male; NS, nevus sebaceous; Ref, reference; TAA, tubular apocrine adenoma

we wish to encourage the incorporation of dermoscopy as an integral part of clinical skin examination so that the dermoscopic patterns can be defined and established for benign adnexal tumors thus averting the need for biopsy as a routine procedure in these patients.

## Conclusion

In this report, we describe clinical, histopathological features along with dermoscopic findings of SCAP in a 12-year-old girl. We wish to highlight the dermoscopic features of SCAP and emphasize that dermoscopy can contribute significantly to reaching a diagnosis of this rare entity. As far as we are aware, our patient represents the third case of de novo linear SCAP over the chest.

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