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

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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; 10x 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).
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
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 | -           | Premlatha 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 Biek 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  |

*(Continued next page)*
| 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 sebaceus; 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.

References

1. Chauhan A, Gupta L, Gautam RK, Bhardwaj M, Gopichandani K. Linear syringocystadenoma papilliferum: A case report with review of literature. Indian J Dermatol. 2013;58(5):409.
2. Karg E, Korom I, Varga E, Ban G, Turi S. Congenital syringocystadenoma papilliferum. Pediatr Dermatol. 2008;25(1):132-133.
3. Pahwa P, Kaushal S, Gupta S, Khaitan BK, Sharma VK, Serburman G. Linear syringocystadenoma papilliferum: an unusual location. Pediatr Dermatol. 2011;28(1):61-62.
4. Pinkus H. Life history of naevus syringocystadenomatous papilliferus. Arch Dermatol Syphil. 1954;69(3):305-322.
5. Rostan SE, Waller JD. Syringocystadenoma papilliferum in an unusual location: report of a case. Arch Dermatol. 1976;112(6):835-836.
6. Premalatha S, Rao NR, Yesudian P, Razack A, Zahra A. Segmental syringocystadenoma papilliferum in an unusual location. Int J Dermatol. 1985;24(8):520-521.
7. Goldberg NS, Esterly NB. Linear papules on the neck of a child: syringocystadenoma papilliferum. Arch Dermatol. 1985;121(9):1198-1201.
8. Epstein BA, Argenyi ZB, Goldstein G, Whitaker D. An unusual presentation of a congenital benign apocrine hamartoma. J Cutan Pathol. 1990;17(1):53-58.
9. de Bieck JP, Starink TM. Multiple linear syringocystadenoma papilliferum. J Eur Acad Dermatol Venereol. 1999;12(1):74-76.
10. Patterson JW, Straka B, Wick MR. Linear syringocystadenoma papilliferum of the thigh. J Am Acad Dermatol. 2001;45(1):139-141.
11. Dawn G, Gupta G. Linear warty papules on the neck of a young woman: syringocystadenoma papilliferum (SP) in a sebaceous nevus (SN). Arch Dermatol. 2002;138(8):1091-1096.
12. Laxmisha C, Thappa DM, Mishra MM, Verma SK. Linear syringocystadenoma papilliferum of the scalp. J Eur Acad Dermatol Venereol. 2007;21(2):275-276.
13. Narang T, De D, Dogra S, Saikia UN, Handa S. Linear papules and nodules on the neck. Syringocystadenoma papilliferum (SP). Arch Dermatol. 2008;144(11):1509-1514.
14. Gonul M, Soylu S, Gul U, Kaya I, Albayrak I, Unal T. Linear syringocystadenoma papilliferum of the arm: A rare localization of an uncommon tumour. Acta Derm Venereol. 2000;88(5):528-529.
15. Yaghooobi R, Zadeh SH, Zadeh AH. Giant linear syringocystadenoma papilliferum on scalp. Indian J Dermatol Venereol Leprol. 2009;75(3):318-319.
16. Yap FB, Lee BR, Baba R. Syringocystadenoma papilliferum in an unusual location beyond the head and neck region: A case report and review of literature. Dermatol Online J. 2010;16(10):4.
17. Martorell-Calatayud A, Sanz-Mottilla V, Garcia-Sales MA, Calatayud-Blas A. Linear syringocystadenoma papilliferum: an uncommon event with a favorable prognosis. Dermatol Online J. 2011;17(8):5.
18. Kar M, Kar JK, Maiti S. Giant linear syringocystadenoma papilliferum of the back. Indian J Dermatol Venereol Leprol. 2012;78(1):123.
19. Ghosh SK, Mandal RK, Bandypadhyay 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;18(11):16.
20. Bandyopadhyay D, Saha A, Kumar D. Linear syringocystadenoma papilliferum on female breast: a rare appendageal tumour on an unusual location. Dermatol Online J. 2014;21(2).
21. Ekinci AP, Buyukbabani N, Mehdi L, Yazganoglu KD, Baykal C. Linear syringocystadenoma papilliferum on the retroauricular area associated with nevus sebaceous. Dermatol Online J. 2016;22(11).
22. Bruno CB, Cordeiro FN, Soares Fdo E, Takano GH, Mendes LS. Dermoscopic aspects of syringocystadenoma papilliferum associated with nevus sebaceous. An Bras Dermatol. 2011;86(6):1213-1216.
23. Zabalous P, Serrano P, Flores G, et al. Dermoscopy of tumours arising in naevus sebaceous: a morphological study of 58 cases. J Eur Acad Dermatol Venereol. 2015;29(11):2231-2237.
24. Duman N, Ersoy-Evans S, Erkin Özaygen G, Gököz Ö. Syringocystadenoma papilliferum arising in naevus sebaceous: a morphological study of 58 cases. J Eur Acad Dermatol Venereol. 2015;29(11):2231-2237.
25. Duman N, Ersoy-Evans S, Erkin Özaygen G, Gököz Ö. Syringocystadenoma papilliferum arising in naevus sebaceous: A 6-year-old child case described with dermoscopic features. Australas J Dermatol. 2015;56(2):e53-54.
26. Zabalous P, Bańuls J, Medina C, Salsench E, Serrano P, Guionnet A. Linear syringocystadenoma papilliferum on the retroauricular area associated with nevus sebaceous. An Bras Dermatol. 2011;86(6):1213-1216.
27. Zabalous P, Bańuls J, Medina C, Salsench E, Serrano P, Guionnet A. Linear syringocystadenoma papilliferum on the retroauricular area associated with nevus sebaceous. An Bras Dermatol. 2011;86(6):1213-1216.
28. Zabalous P, Bańuls J, Medina C, Salsench E, Serrano P, Guionnet A. Linear syringocystadenoma papilliferum on the retroauricular area associated with nevus sebaceous. An Bras Dermatol. 2011;86(6):1213-1216.