

Giant syringocystadenoma papilliferum along the Blaschko's lines

Dear Editor,

Syringocystadenoma papilliferum (SCAP) is a benign adnexal tumour originating from both apocrine and eccrine histogenesis. It was first reported as 'naevus syringadenomatosus papilliferus' by Stokes in 1917.¹ SCAP typically presents as a papillary or solitary lesion in the head or neck region. On rare occasions, it presents as multiple small papules, occasionally along Blaschko lines. We report an unusual case of giant SCAP presenting along Blaschko lines on the thigh. We also reviewed the relevant literature on giant SCAP to further comprehend this disease.

A 24-year-old woman presented with red giant tumours over the right outer thigh in a linear configuration in 2023. The lesions began as several small papules shortly after birth, which then gradually increased in size and number without showing any other symptoms. However, the lesions progressed significantly with traumatic bleeding in 2022. The patient has no history of trauma or any notable medical issues. A physical examination revealed several red, dark-brown, moist, cauliflower-like papules measuring 1–5 mm in diameter and plaques of sizes 4 x 5 cm, 5 x 5 cm and 9 x 3 cm on the right thigh along the Blaschko lines [Figure 1]. All laboratory data were within the normal ranges. Squamous cell carcinoma, giant warts, tubular apocrine adenoma, and verrucous naevus were initially considered as differential diagnoses. Thereafter, a punch biopsy of the largest lesion revealed an exophytic configuration composed of epithelium-covered papillae [Figure 2a]. The inner layer of the papillae showed small cuboidal myoepithelial cells with hyperchromatic nuclei, while the outer layer was composed of tall columnar cells with eosinophilic cytoplasm, large vesicular nuclei and decapitation secretion [Figure 2b]. The stroma contained numerous plasma cells. Immunohistochemistry results indicated that the columnar cells were positive for AE1/AE3 [Figure 2c], CK-7 [Figure 2d] and carcinoembryonic antigen (CEA) [Figure 2e], while the cuboidal cells were positive for P63 [Figure 2f]. Plasma cells of the stroma were positive for CD38 [Figure 2g]. SCAP was diagnosed based on the



Figure 1: Multiple red, dark-brown, moist, cauliflower-like papules and plaques on the right thigh along Blaschko lines.

clinical findings and characteristic histological features of the present case. All lesions were completely excised and closed with a tension-reducing sutures. The excised lesions were reconfirmed as SCAP by histopathological examination without any malignant transformation. The wound healed completely eight weeks after the surgery.

SCAP is a benign cutaneous neuroma derived from both apocrine and eccrine histogenesis and composed of undifferentiated pluripotential cells.² Clinically, it typically presents as a dark-brown or grey papillary or rather warty, sometimes crusty, excrescence with a moist appearance

How to cite this article: Han B, Zhang W, Li S, Wang H, Tian Y. Giant syringocystadenoma papilliferum along the Blaschko's lines. *Indian J Dermatol Venereol Leprol*. doi: 10.25259/IJDVL_659_2023

Received: June, 2023 **Accepted:** April, 2024 **Epub Ahead of Print:** July, 2024

DOI: 10.25259/IJDVL_659_2023

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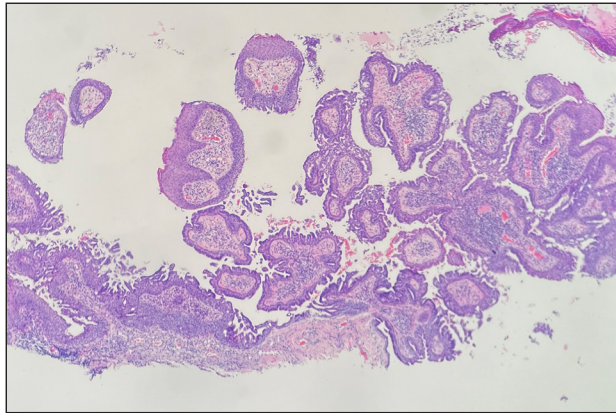


Figure 2a: Exophytic configuration composed of epithelium-covered papillae (Haematoxylin and Eosin, 40x).

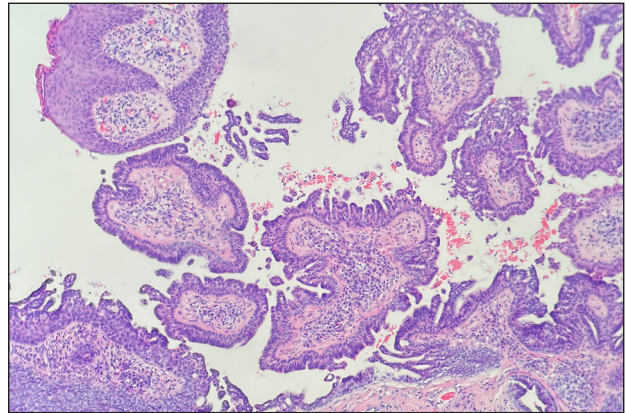


Figure 2b: The inner layer of the papillae included small cuboidal myoepithelial cells, while the outer layer was composed of tall columnar cells (Haematoxylin and Eosin, 100x).

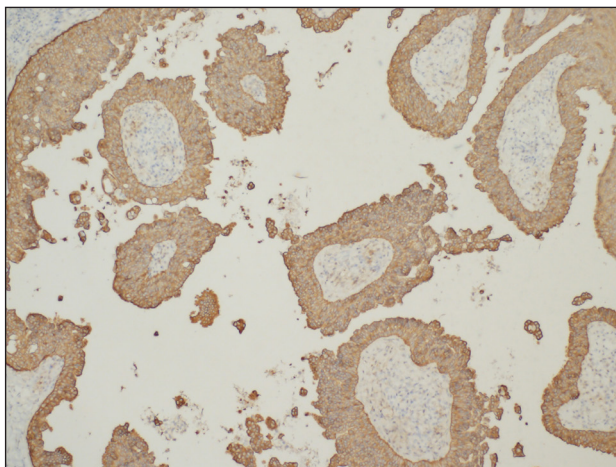


Figure 2c: Columnar cells were positive for CK cocktail AE1/AE3 (AE1/AE3 Immunohistochemistry, 100x).

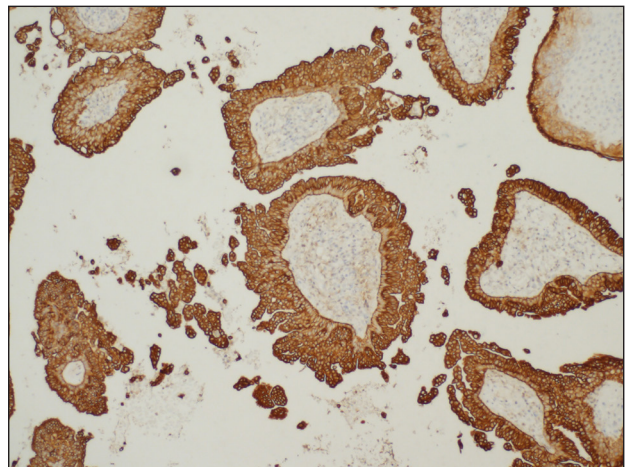


Figure 2d: Columnar cells were positive for CK-7 (CK-7 Immunohistochemistry, 100x).

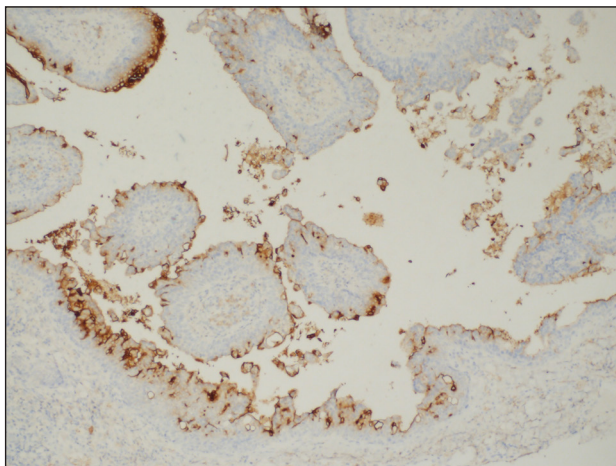


Figure 2e: Columnar cells were positive for CEA (CEA Immunohistochemistry, 100x).

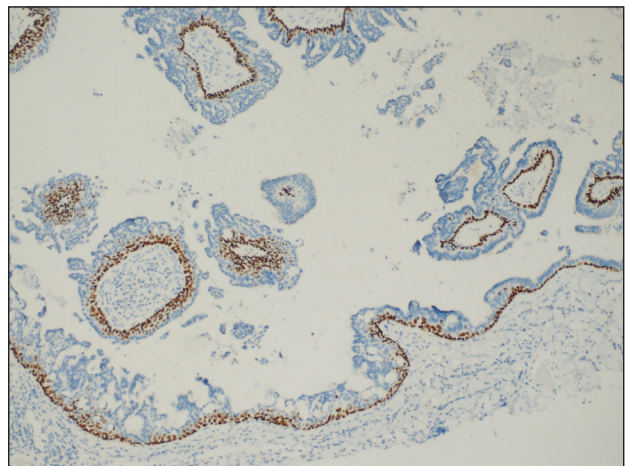


Figure 2f: Cuboidal cells were positive for P63 (P63 Immunohistochemistry, 100x).

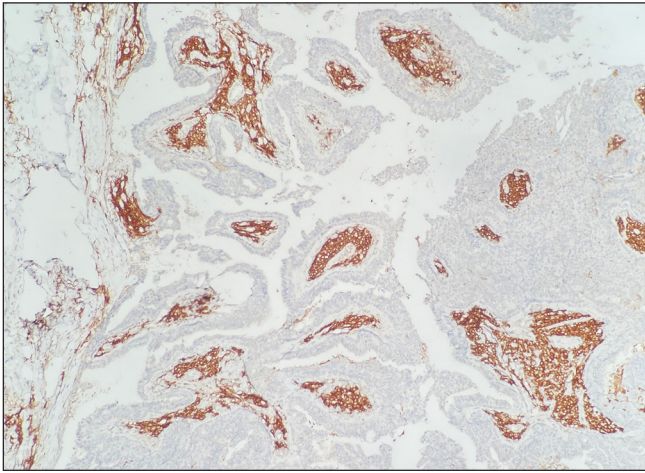


Figure 2g: Plasma cells of the stroma were positive for CD38 (CD38 Immunohistochemistry, 100×).

and positive for P63 in cuboidal cells.² The clinical and histological features of the present case were found to be consistent with those of SCAP.

SCAP has recently been reported to be associated with somatic activating mutations in the *HRAS*, *KRAS* or *BRAF* oncogenes.⁷ Owing to financial reasons, the patient refused genetic testing. Surgical excision is the first-line treatment option for SCAP and CO₂ laser may be considered for anatomic sites unfavourable to surgical excision.¹ All lesions of the present patient were removed surgically.

In summary, we report a case of Blaschko-linear SCAP located on the thigh with a giant appearance. Our case findings highlight the important role played by histopathological examination to rule out other cutaneous tumours demonstrating similar characteristics.

Table 1: The published reports on cases of giant syringocystadenoma papilliferum (SCAP)

Author (date)	Age (year)/sex	Age of onset (year)	Location	Appearance	Clinical features	Accompanying diseases
Kar <i>et al.</i> ¹ (2012)	12/M	Since birth	Lower back	A 20-cm long and 7-cm wide fleshy cauliflower-like erythematous papulonodular lesion arranged in a linear array	No	No
Yaghoobi <i>et al.</i> ⁴ (2009)	20/F	19	Left-sided occipital part of the scalp	A linear lesion, highly elevated, moist, fetid, vegetated and pinkish, measuring 8 × 3 × 2 cm in diameter	No	Hypochromic, microcytic anaemia
Sardesai <i>et al.</i> ⁵ (2009)	30/F	25	The medial aspect of the right thigh	Two skin lesions: (large) 6 × 7 cm and (small) 1 × 3 cm	No	Condyloma acuminata
Kaehler <i>et al.</i> ⁶ (2005)	43/M	Since birth	The left side of the ventral thorax	A giant, 4 × 6 cm brownish, verrucous tumour with several scars	No	No
Our case (2023)	24/F	Since birth	Right thigh along Blaschko line	Several red, dark-brown, moist, cauliflower-like papules, measuring 1–5 mm in diameter, and plaques of size 4 × 5 cm, 5 × 5 cm, and 9 × 3 cm, respectively, on the right thigh along the Blaschko lines	Traumatic bleeding	No

that may manifest at birth or develop during childhood.² Head and neck regions are most commonly involved sites.² The following three clinical types of SCAP have been described so far: solitary nodular type, plaque type and linear type, of which, the linear type is rare.³ So far, there have been only four published cases of giant SCAP reported in the English literature,^{1,4-6} and none has been described to have presented on the thigh as a linear type. Based on the literature review, the age at the time of diagnosis, sex, the age of onset, location, appearance, clinical features, and any accompanying diseases are summarised in Table 1. Histologically, SCAP has a characteristic that makes it readily recognisable owing to its papillary projection consisting of two rows of cells – columnar and cuboidal, and the stroma contains numerous plasma cells.² SCAP has been reported to be positive for carcinoembryonic antigen, cytokeratin AE1 (CK AE1), and epithelial membrane antigen (EMA) in columnar cells

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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