Blaschko-linear Syringocystadenoma Papilliferum: A Peculiar Presentation

Sir,

Syringocystadenoma papilliferum (SCAP) is a benign adnexal neoplasm, mostly developing in a background of an organoid nevus over the head and neck region. Rarely, they can develop *de novo*. Here, we present the case of a young gentleman who presented with multiple papulo-nodules distributed in a linear pattern over the upper back and extending to the left arm. Histology was consistent with a diagnosis of SCAP. The unique features of our case are Blaschko-linear distribution over the upper trunk and *de novo* development in the absence of any organoid nevus in the background. The rarity of such a presentation in the English literature prompted us to report the case.

A 20-year-old otherwise healthy man was referred to our department for the evaluation of multiple asymptomatic, solid, elevated skin lesions over the back, present for the last 11 years. To begin with, there were a few pea-sized lesions on the midline of the back, which gradually increased in size and number to attain the present state. Cutaneous examination revealed numerous skin-colored to erythematous dome-shaped papulo-nodules coalescing to form plaques over the left side of the upper back and extending towards the extensor aspect of the left arm and chest with typical Blaschko-linear distribution. Few lesions were verrucous and eroded as well. Furthermore, there were similar lesions over the front of the chest [Figure 1]. The patient did not give history of any lesions prior to the development of the present nodules. Histopathological examination revealed hyperkeratosis, acanthosis, papillomatosis, and deep duct-like invaginations extending into the dermis. Besides, the papillary projections could be well appreciated. The ducts were lined by squamous epithelium in the upper portion and by two-layered columnar epithelium in the lower portion. Numerous plasma cells were present in the connective tissue stroma of the papillae. There was proliferation of eccrine and apocrine structures in the dermis, with evidence of decapitation secretion [Figures 2-4]. Based on the clinicopathological findings, a diagnosis of "*de novo* blaschko-linear syringocystadenoma papilliferum" was made. Immunohistochemical study could not be done due to lack of institutional facilities. The patient was referred to a plastic and reconstructive surgeon for surgical intervention.

SCAP is an uncommon neoplasm. It was earlier known as "adenoma cystoma intracanaliculare" and "nevus syringoadenomatous papilleferus." In 50% of the cases, it is discernible at birth, and in 15–30% of the cases, the lesions are evident in the peri-pubertal age. With age, the lesions tend to become more fleshy, verrucous, and larger. In most cases, it develops over an organoid nevus. Rarely, as in our case, the lesions develop as a primary benign neoplasm in the absence of any nevus.

SCAP has a varied clinical presentation ranging from a solitary skin-colored hairless nodule to verrucous plaque, and eroded and ulcerated mass. Unusual locations of SCAP include pinna, postoperative scar, breast, buttock, back, and knee.[1-6] Linear SCAP have been rarely reported over the arm, neck, scalp, pubic region, thigh, and abdomen.[7-12] Other adnexal tumors that show linear arrangement include nevus comedonicus, trichoepithelioma, basaloid follicular hamartoma, cylindroma, eccrine nevus, syringoma, eccrine poroma, eccrine spiradenoma, basal cell carcinoma, etc. However, the histology of SCAP is classical and consistent helping us to reach a diagnosis. SCAP has been reported to be positive for carcinoembryonic antigen (CEA), cytokeratin AE1 (CK AE1), and epithelial membrane antigen (EMA). Rarely, SCAP may become transformed into basal cell carcinoma, squamous cell carcinoma, sweat gland carcinoma, and syringocystadenocarcinoma papilliferum.[13] Treatment is surgical excision. Other options include CO_2 laser and Mohs' micrographic surgery.

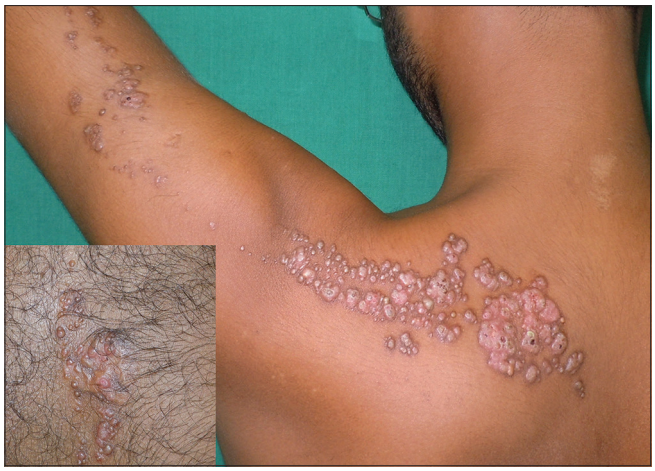

Figure 1: Linear distribution of dome-shaped papules and nodules over the left upper back, extending to the extensor aspect of the arm. Inset showing presence of similar lesions over the front of the chest

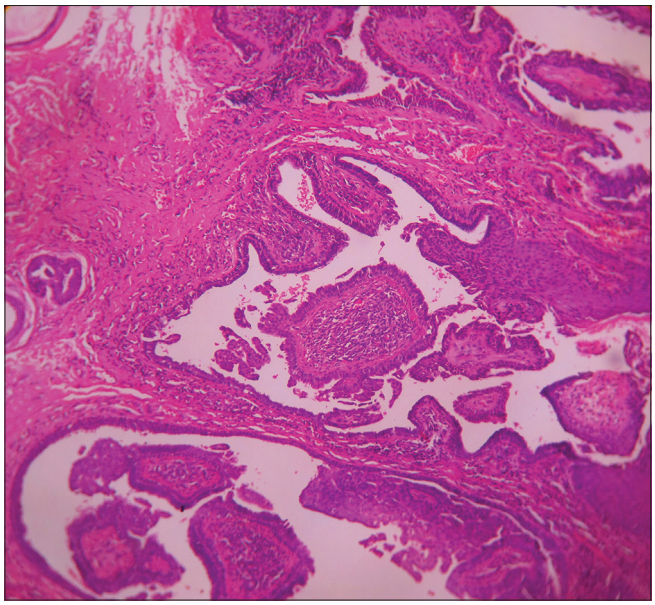

Figure 3: Photomicrograph showing ducts lined by squamous epithelium in the upper portion and by two-layered sweat duct-like epithelium in the lower portion. (H and E, ×100)

Features such as Blaschko-linear distribution over the upper trunk and *de novo* development made our case unique and encouraged us to report for its rarity.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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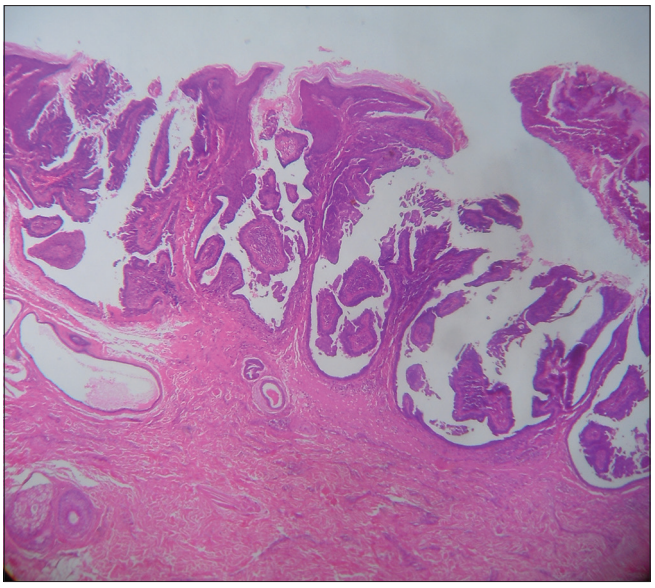

Figure 2: Photomicrograph showing hyperkeratosis, acanthosis, papillomatosis, and deep duct-like invaginations extending into the dermis. (H and E, scanner view)

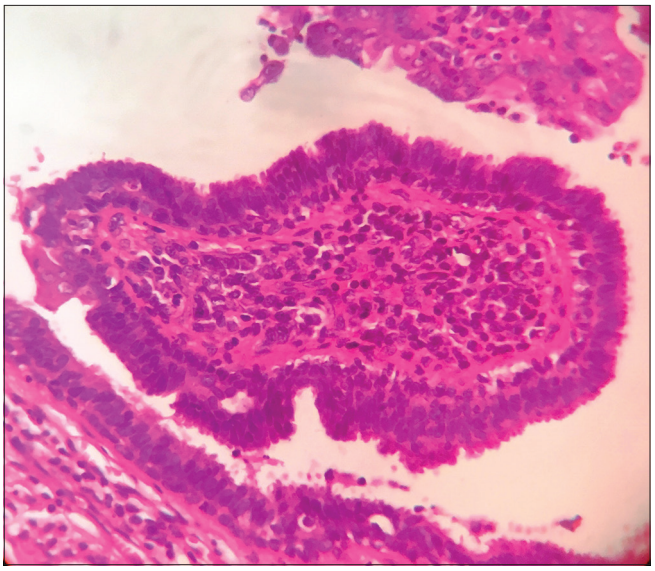

Figure 4: Photomicrograph showing two-layered sweat duct-like epithelium with evidence of decapitation secretion. (H and E, ×400)

Conflicts of interest

There are no conflicts of interest.

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