Letter to Editor

Linear and Pedunculated Syringocystadenoma Papilliferum of the Lower Abdomen: Rare Localization, Clinical Presentation, and Histopathology

Sir,

Syringocystadenoma papilliferum (SCAP) occurs most commonly on the head and neck; however, in about one-fourth of cases, it may be seen elsewhere. It is usually first noted at birth or in early childhood and presents as a papule or several papules in a linear arrangement or as a plaque. The lesion increases in size at puberty, becoming papillomatous, and often crusted.[1] There is an associated organoid nevus in approximately one-third of cases; basal cell carcinoma may develop in 10% of lesions. Furthermore, SCAP is known to be associated with a variety of other skin adnexal benign and malignant tumors.[2] We are reporting a case of linear SCAP in the lower abdomen.

A 25-year-old man came to our dermatology clinic complaining of the skin lesion in the lower abdomen. He had this lesion since early childhood, describing it as three asymptomatic linearly arranged elevated skin lesions, which had enlarged during puberty but remained asymptomatic until a few months ago. The recently added pruritus made the patient seek medical advice. Clinical examination showed three separate linearly arranged lesions adjacent to the umbilicus. The first lesion being a 1 cm × 0.8 cm warty erythematous plaque, second as a 0.6 cm × 0.5 cm erythematous plaque with central umbilication, and the last (nearest to the umbilicus) as a 1.7 cm × 1 cm pedunculated erythematous lesion with verrucose changes on its surface [Figure 1].

The histopathological evaluation revealed skin tissue with papillomatosis, thick laminated orthokeratotic and parakeratotic hyperkeratosis, and acanthotic broad-based rete ridges in the epidermis. No cytopathic effect of viral infection was noted [Figure 2]. There was also a cystic invagination extending from the surface epidermis into the dermis, with multiple intra cystic papillary projections, lined by high columnar epithelial cells, and subjacent cuboidal cells [Figure 3]. The epithelial cells showed decapitation on the surface. The mitotic activity was nil, and there was no conspicuous cytologic atypia or necrosis indicating its benign nature. Numerous plasma cells were seen within the stroma of papillae [Figure 4]. There was no accompanying hyperplasia and/or abnormality in morphology or distribution of sebaceous, eccrine or apocrine glands arguing against the diagnosis of organoid nevus.

SCAP was first described as “nevus syringadenomatosus papilliferus” by Stokes in 1917. Though most cases occur on the head and neck region (75%), trunk and extremities may be involved in few cases. There are increasing reports of SCAP occurring in unusual locations. Yap et al. reviewed the literature for cases of SCAP outside the head and neck region and found 69 such cases, which include trunk (53.5%), extremities (33.8%), and genital SCAPs (12.5%).[1] Of the 38 cases on the trunk, 31 cases have not been more specified, three cases were on the breast, two on the back, one on the abdomen, and one on the chest.[1] Only four cases of SCAP occurring on the abdomen, of which only two were linear, have previously been documented in literature.[1,3]

SCAP presents as skin colored to dark brown or erythematous, solitary or multiple plaque, nodule, or linear lesion with various sizes. Linear lesions are the rare and only 17 cases with this

Figure 1: Clinical appearance of the lesions, approaching the midline, respectively, the first lesion was biopsied before the picture was taken, the second one is umbilicated, and the last one is pedunculated with verrucous surface

Figure 2: Whole mount view of the lesion. The surface epidermis presents church-siring (papillomatosis), and there is a cystic lesion with intracystic papillary structures in the dermis
presentation have been reported.\textsuperscript{[2,4-7]} The surface is papillomatous, verrucous, smooth, or flat. It is a clinically indistinct sweat gland tumor requiring biopsy for definite diagnosis. SCAP may enlarge and develop a papillomatous and crusted surface during puberty. SCAP is an adnexal skin tumor with ongoing discrepancy about its exact histogenesis. Features supporting its apocrine origin include epithelial lining composed of cells with prominent decapitation, staining of the luminal cells by Alcian blue, periodic acid–Schiff after diastase treatment, and colloidal iron methods.\textsuperscript{[3]} Positive immunohistochemical (IHC) staining for Gross Cystic Disease Fluid Protein 15, Leu-M1 antigen, and epithelial membrane antigen also support an apocrine origin. On the other hand, reports of positive staining for eccrine-specific marker IKH-4 along with some ultrastructural findings are in favor of an eccrine origin for these tumors. There is also an alternative theory that they originate from the pluripotent stem cells which could explain these divergent findings.\textsuperscript{[8]} Accordingly, SCAP is a hamartomatous lesion Mammino and Vidmar by reviewing 145 SCAP cases; based on microscopic, IHC, and ultrastructural findings; they observed that SCAP is a mixture of apocrine and eccrine elements.\textsuperscript{[6]} In conclusion, we are reporting a classic case of SCAP with unusual localization and clinical presentation.

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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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Conflicts of interest

There are no conflicts of interest.

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