A case series of three atypical cases of syringocystadenoma papilliferum with thigh, abdomen and axillary involvement, and review of literature

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Syringocystadenoma papilliferum is an uncommon benign apocrine gland tumour, mostly of congenital or early childhood onset, affecting head and neck. In addition, nevus sebaceous is commonly associated with this tumor. Here, we document three cases of syringocystadenoma papilliferum with many atypical features, such as the involvement of rare sites (abdomen, axilla and thigh), absence of nevus sebaceous and coexistence of tubular apocrine adenoma. The histopathologic characteristics were consistent with the diagnosis in all three cases.

Keywords: syringocystadenoma papilliferum, apocrine, hamartomatous

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INTRODUCTION

Syringocystadenoma Papilliferum (SCAP) is an uncommon skin appendageal benign tumour, predominantly of apocrine origin [1]. Most cases are either congenital or of childhood onset, where head and neck regions are the most commonly involved sites. SCAP might develop beyond these regions and involve trunk, genitalia or limbs. The coexistence with tubular apocrine adenoma (TAA) is also a rare phenomenon [1]. Here, we document three cases of SCAP, among whom, two are localized over abdomen and axilla, and the third one is associated with TAA located on the thigh.

CASE PRESENTATION

Case 1

A 14yearold boy presented with asymptomatic, solitary, pinkish plaque over the epigastric region of abdomen present since birth. The lesion was initially small, papular and started progressing gradually over the past 3-4 years. Upon local examination, mild papillomatous features and hemorrhagic foci were noted within the plaque and the size was approximately 2 cm × 4 cm (Figure 1a). Regional lymphadenopathy was absent and systemic examination was also unremarkable. The routine hematological and biochemical tests were within normal limits. Chest radiograph and abdominal ultrasonography were also normal.

Histopathological examination showed cystic invagination of epidermis and cystic spaces in the upper dermis. Papillary projections were further observed in these invaginations and spaces (Figure 1b). These projections were lined by two rows of cells: inner luminal columnar cells and outer cuboidal cells. Decapitation was also seen in the luminal layer (Figure 1c). Mixed inflammatory infiltrates consisting of lymphocytes, plasma cells and neutrophils were present in the fibrovascular

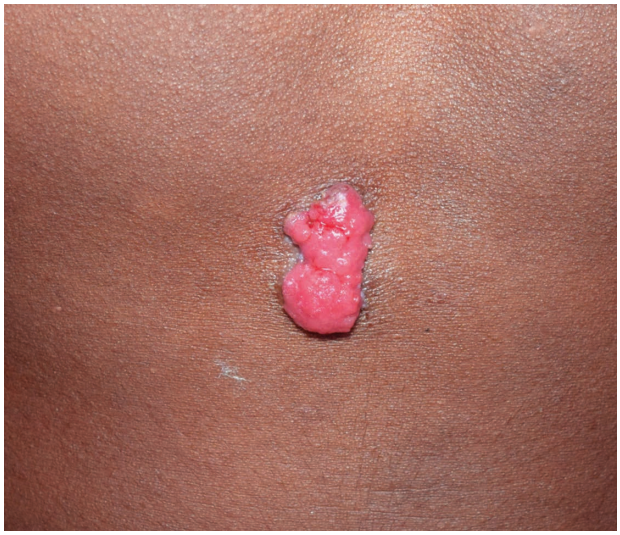

Figure 1a. A solitary, pinkish, plaque with mild papillomatous appearance and haemorrhages over the epigastric region of the abdomen

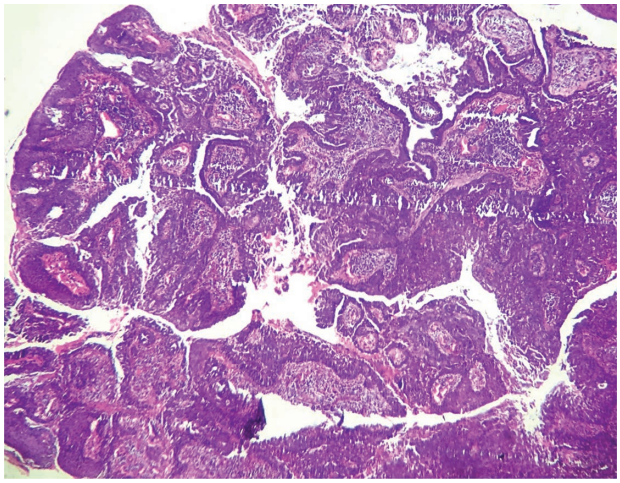

Figure 1b. Acanthosis and cystic invagination of epidermis with papillary projections, extending into the dermis (H & E, ×100)

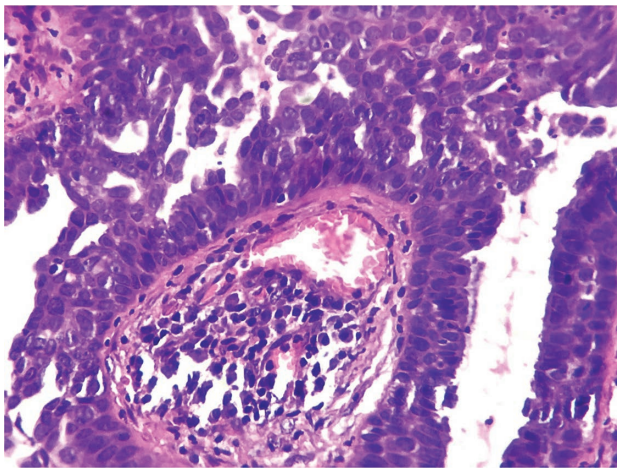

Figure 1c. Double layered sweat duct-like epithelium along with decapitation secretion. Plasma cells are present within the stroma of invagination (H & E, ×400)

core. These histopathologic features were consistent with the diagnosis of SCAP.

Case 2

A 32-year-old male referred to our dermatology department presenting with mildly painful nodular lesions on the right axilla, which he had since childhood. Upon local cutaneous examination, two sessile hyperpigmented nodules fused at their base were seen on the lower side of right axilla, of which the larger nodule was about 2 cm × 2 cm with eroded surface (Figure 2a). Other mucocutaneous

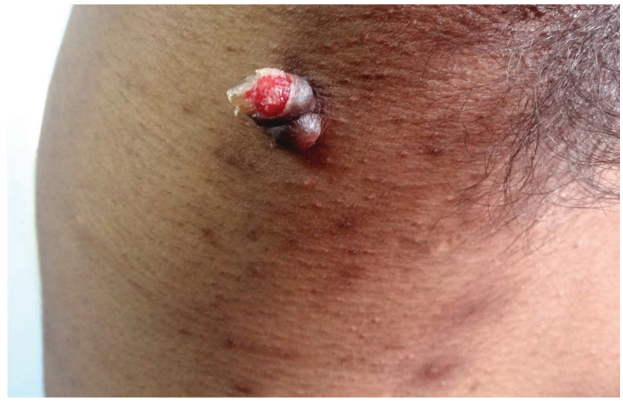

Figure 2a. Hyperpigmented, polypoid lesion with eroded top, observed in the right axilla.

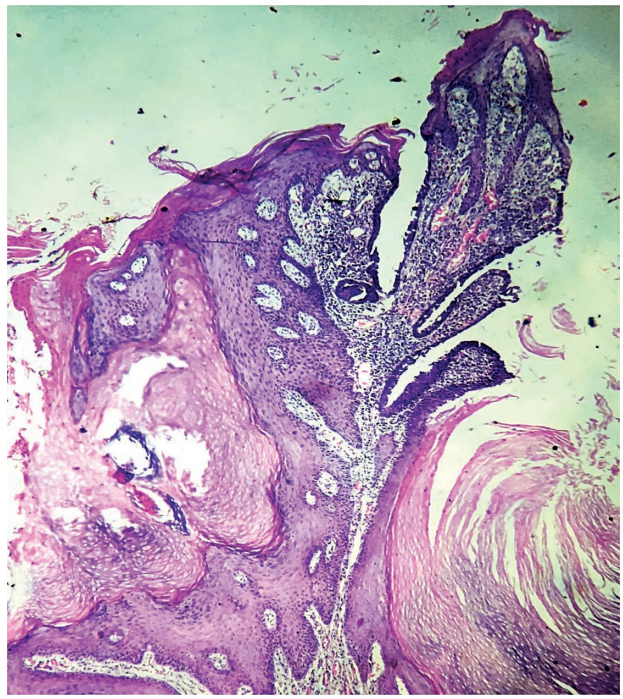

Figure 2b. Hyperkeratosis, acanthosis, papillomatosis, and duct-like epidermal invaginations extending into the upper dermis (H & E, ×400).

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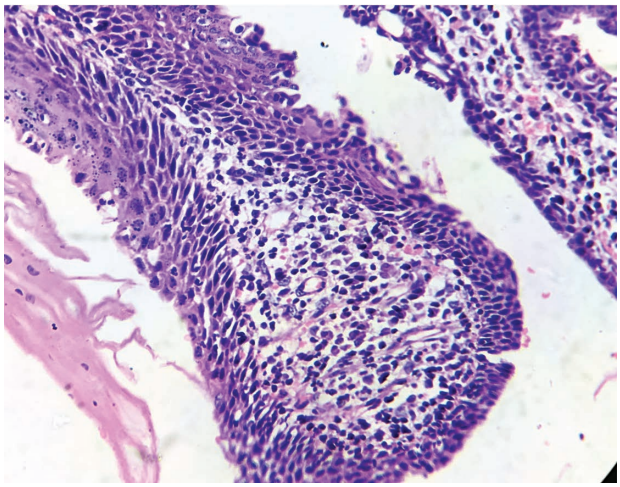

Figure 2c. Double-layered sweat duct-like epithelium showing decapitation secretion. Plasma cell infiltration within stroma (H & E, ×400).

and systemic examinations were not remarkable. The routine haematological and laboratory parameters were also within normal limits.

Histopathologic examination revealed cystic epidermal invagination with papillary projections (Figure 2b), lined by inner luminal columnar cells showing decapitation and outer cuboidal cells (Figure 2c). These features were characteristic of SCAP.

Case 3

A 15-year-old girl presented with asymptomatic single plaque on the left thigh since her birth. Of note, it started growing rapidly in the past 2 years. Upon local cutaneous examination, it was seen as a whitish firm rubbery plaque of 2.5 cm × 1.5 cm size with blood tinged and crusted surface at certain places (Figure 3a). No other mucocutaneous and systemic examinations were found remarkable.

Histopathologic examination revealed the upper part of the tumor with cystic epidermal invagination and papillary projections (Figure 3b), lined by inner luminal columnar cells showing decapitation, outer cuboidal cells and plasma cell rich stroma (figure 3c). These features are characteristic of SCAP. The deeper portion showed tubular structuress without any epidermal connection or papillary projections, yet lined by the similar two rows of cells, suggestive of tubular apocrine adenoma (Figure 3b).

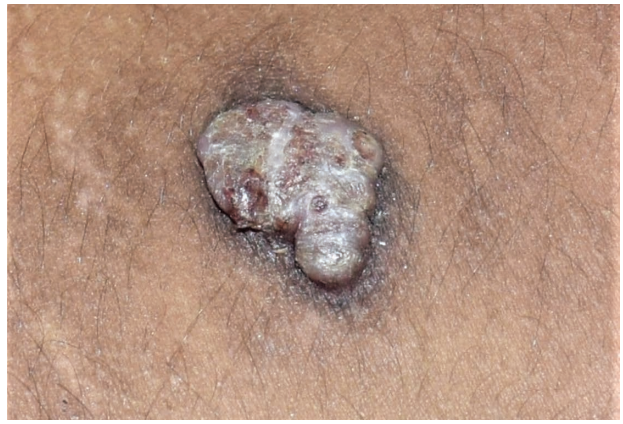

Figure 3a. Solitary whitish firm rubbery plaque with erosion and crusting at certain places

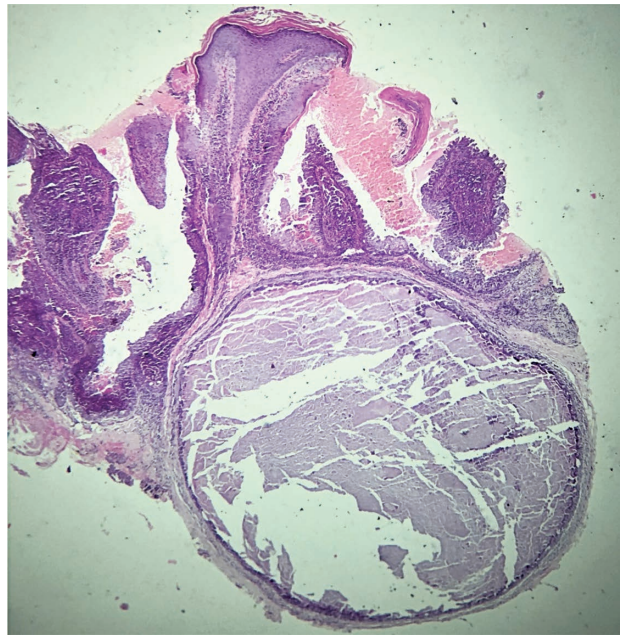

Figure 3b. Upper part of the tumor showing epidermal invaginations with papillary projections. The deeper portion showed tubular structures without any epidermal connection or papillary projections (H & E, ×100)

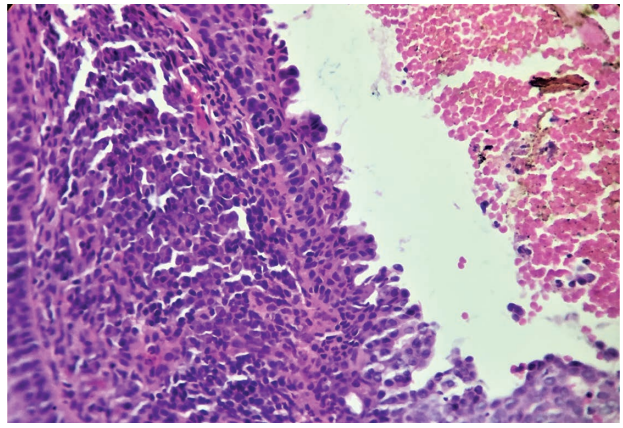

Figure 3c. Two rows of cells: inner luminal columnar and outer cuboidal cells (H & E, ×400)

DISCUSSION

SCAP is an uncommon, benign hamartomatous sweat gland tumour, also known as adenoma cystoma intracanaliculare and nevus syringoadenomatous papilleferus [2].

The fact that SCAP is considered more of apocrine origin is still disputed. Apocrine and eccrine, both lineages of differentiation have been reported on immunohistochemical and ultrastructural studies in literature. Therefore, it is becoming more and more acceptable to consider it of hamartomatous origin from undifferentiated pleuripotential cells capable to differentiate both lineages [3].

About half of the cases occur at birth or in childhood, and 15–30% develop in the peripubertal age. It more frequently than not presents as one of the three well recognized morphologic forms: solitary papule or nodule, smooth hairless plaque, and, less commonly, as linearly arranged multiple papules and nodules [3]. They are mostly asymptomatic and brownish or erythematous in colour. During peripubertal age, probably under androgenic hormonal stimulation, lesions tend to become larger, verrucous/papillomatous and crusted sometimes [4].

SCAP mostly occurs on the head and neck, although it may also develop on the back, abdomen, arms, breast, axilla, lower limb, inguinal, vulva and scrotum [4]. To our knowledge, only seven cases of SCAP have been associated with axilla, and five cases with abdomen [4-10]. SCAP on thigh has been reported in only 10 cases so far [2,3,11-18].

Nevus sebaceous on scalp is the most frequent association, but there are many others such as linear naevus verrucosus, naevus comedonicus, tubular apocrine adenoma (TAC), apocrine poroma, apocrine hidrocystoma, hidradenoma papilliferum, papillary eccrine adenoma, warts, verrucous carcinoma, syringoma and basal cell epithelioma [3]. Of note, TAC has been associated with SCAP in only 13 cases so far, most of which were present over scalp with pre-existing nevus sebaceous. The first of such associations was reported on the scalp by Toribio et al. in 1987 in a 33-year-old male [19]. In the following three decades, more cases were seen, with the most recent one published in 2017 by Leda et al, where a 14-year-old male had a plaque on his back [1]. In our third case, lesions of SCAP with TAC, present on thigh, and the absence of sebaceous nevus were the unique features which encouraged the present report.

Histopathologically, SCAP is characterized by several cystic invaginations of the epithelium into the dermis leading to the dermal cystic spaces. In the invaginations and cystic spaces, papillary projections are seen protruding into their lumen, lined by two rows of epithelial cells; outer cuboidal and inner luminal high-columnar epithelium. Occasionally, some of these cells show active decapitation secretion, and cellular debris is found in the lumina. Invariably, dilated capillaries and a dense plasma cell infiltrate are present in the stroma of the papillary projections [1].

SCAP should be differentiated from hidradenoma papilliferum, papillary eccrine adenoma, eccrine poroma, eccrine spiradenoma, trichoepithelioma, basaloid follicular hamartoma, cylindroma, warty dyskeratoma, basal cell carcinoma and squamous cell carcinoma [3].

CONCLUSION

Syringocystadenoma papilliferum is an uncommon benign apocrine sweat gland tumor usually involving head and neck. Involvements with axilla, abdomen,lower limb, and tubular apocrine adenoma are very rare. We reported three cases of syringocystadenoma papilliferum located on axilla, abdomen and thigh. Moreover, the case with plaque on thigh had also the histopathologic features of tubular apocrine adenoma. Accordingly, although rare, occurrence of syringocystadenoma papilliferum is not limited to head and neck regions, hence the necessity of the histopathologic examination of all such cases.

Conflict of Interest: None declared.

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