Case Series

Unusual sites with variable presentation of *de novo* syringocystadenoma papilliferum: A case series

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Abstract

Syringocystadenoma papilliferum (SCAP) is an uncommon, benign adnexal neoplasm that occurs *de novo* or in an organoid nevus. It usually presents as a skin-coloured to pink, solitary, smooth, hairless plaque, verruca or nodule frequently on the scalp and forehead. SCAP may be present at unusual sites including the arm, forearm, trunk and chest. Diagnosing SCAP arising on uncommon sites is difficult owing to its varied presentation. Mostly, they are wrongly diagnosed clinically and found to be SCAP only on histopathology. We present this study of cases of SCAP with unusual location and varied presentations, which were clinically misdiagnosed. The five cases included in this study were patients attending the dermatology outpatient department in a tertiary care centre in North India. The clinical presentation and the involved sites were noted by the dermatologist, and a clinical diagnosis was made. Biopsy of the lesions was sent for histopathological examination. There are five patients in the series – four are male and one female, with age ranging from 28 to 48 years. Locations included the forearm, arm, anterior chest wall and lateral abdominal wall. The lesions clinically appeared as warty papule or nodules and one lesion appeared within a plaque, with the average duration being 5.3 years. In all five patients, the lesions were clinically suspected to be either tuberculosis verruca cutis or nodular basal cell carcinoma or dermatofibroma sarcoma protuberans (DFSP) or verruca or fibroma or pyogenic granuloma. A confirmatory diagnosis of SCAP was made for all the patients on histopathology. We are presenting five cases which were misdiagnosed clinically due to the unusual location and varied presentation to emphasise the importance of histopathology in diagnosing SCAP arising *de novo,* which was clinically misdiagnosed. Also, we present this case series to alert the clinicians about the likelihood of SCAP on unusual locations with varied clinical presentation.

Keywords: *De Novo* SCAP, histopathological confirmation, misdiagnosis, uncommon sites, variable presentation

Introduction

Syringocystadenoma papilliferum (SCAP) is an uncommon benign adnexal neoplasm of childhood or adolescence that occurs *de novo* or in an organoid nevus and has variable clinical presentation.[1] It usually presents as a skin-coloured to pink, solitary, smooth, hairless plaque, verruca or nodule frequently on the scalp and forehead.[2] Nodular or verrucous transformation is commonly seen at puberty. Histopathologically, SCAP has a characteristic appearance showing ducts connecting to the surface, containing papillary processes and lined by two epithelial cell layers.[3]

Since 75% of the cases are reported in the head and neck region,[4] diagnosing SCAP arising *de novo* and on uncommon sites is difficult. Mostly, they are wrongly diagnosed clinically and are found to be SCAP only on histopathology. This case

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series, therefore, highlights the importance of histopathology in diagnosing SCAP arising *de novo*, which were clinically misdiagnosed due to the unusual location.

Case Presentation

The five cases that constitute this case series were patients attending the dermatology outpatient department in a tertiary care centre in North India from 2017 to 2021. The clinical presentation and the involved sites were noted by the dermatologist, and a clinical diagnosis was made [Table 1]. The lesions were present on unusual locations for SCAP, like arm, chest and trunk, and were clinically suspected to be either tuberculosis verruca cutis or nodular basal cell carcinoma or dermatofibroma sarcoma protuberans (DFSP) or verruca or fibroma or pyogenic granuloma. Biopsy samples of the lesions were sent for histopathological examination. Written consent was taken from the patients to publish this study.

Results

Four patients were male and one was a female, with age ranging from 28 to 48 years (mean = 36 years, median = 35 years) [Table 1]. Locations included the forearm (n = 1), arm (n = 1), elbow (n = 1) [Figure 1], anterior chest wall (n = 1) and lateral abdominal wall (n = 1) [Figure 2]. The lesions clinically appeared as warty papule or nodules and one lesion appeared within a plaque, with the average duration being 5.3 years. Progressive growth was reported in all lesions, with history of recurrence post excision in one lesion. Also, there was history of redness in one lesion. The sizes varied from 1.5 to 4.5 × 1 to 3 × 1 to 1.5 cm (mean = 2.7 × 1.7 × 1 cm) [Table 1]. In all five patients, the lesions were clinically suspected to be either tuberculosis verruca cutis or nodular basal cell carcinoma or DFSP or verruca or fibroma or pyogenic granuloma [Table 1]. The lesions were surgically excised and sent for histopathological examination. On histopathological evaluation, a confirmatory diagnosis of SCAP was made for all the five patients. There was no recurrence in any of the patients at 1 year follow-up.

Histopathological findings

On serial sections, all tumours showed irregular papillary projections protruding as invagination of the surface epithelium [Figures 3a, 4–6]. Papillary projections were seen to be covered by an inner layer of columnar epithelium and an outer layer of cuboidal cells. The stroma of papillary projections showed connective tissue along with numerous plasma cells [Figure 3b].

Discussion

SCAP is a rare benign neoplasm of childhood classically[1,5] and some arising in later life as the patients in our case series.[6]

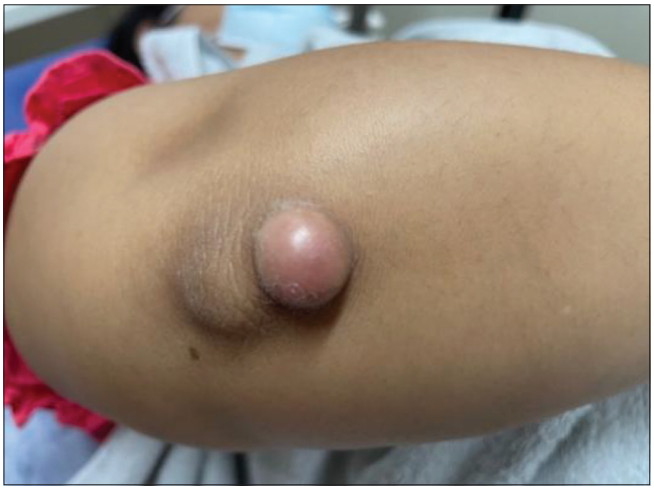

Figure 1: Nodular lesion on the right elbow

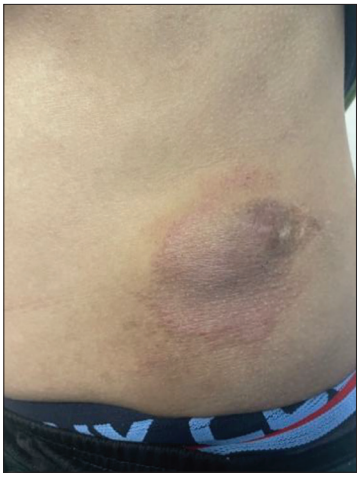

Figure 2: Swelling arising within an erythematous plaque on the left lateral abdominal wall

Table 1: Summary of the cases

Age/gender (years)	Duration (years)	Location	Examination findings	Size (cm)	Differential diagnosis (based on clinical findings)	Diagnosis on histopathology
28/M	5	Forearm	Partially eroded nodule	2×1 × 0.5	Pyogenic granuloma, nodular BCC	SCAP
30/M	4.5	Anterior chest wall	Exophytic papillary growth	1.5×1 × 1	Verruca, papilloma	SCAP
35/F	2	Elbow	Nodule [Figure 1]	1.5×1 × 1	Fibroma, desmoid tumour	SCAP
39/M	5	Lateral abdominal wall (lt)	Swelling arising within erythematous plaque [Figure 2]	4×3 × 1.5	DFSP	SCAP
48/M	10	Arm (rt)	Eroded verrucous nodule, cystic swelling with haemorrhagic fluid	4.5×2.5×2	Haemangioma	SCAP

BC=basal cell carcinoma, DFSP=dermatofibroma sarcoma protuberans, SCAP=syringocystadenoma papilliferum

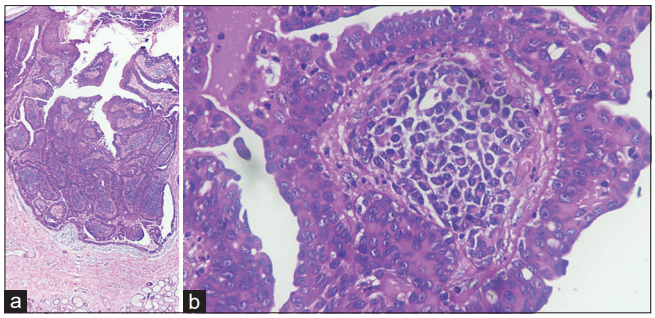

Figure 3: (a) Photomicrograph of the first case showing epidermal invaginations into the dermis, which are lined by stratified squamous epithelium in the upper part and double-layered rows of outer cuboidal and luminal columnar cells in the lower portions (haematoxylin and eosin, 200×). (b). The inner layer is composed of columnar cells, where decapitation secretion is observed, and the outer layer is formed by cuboidal cells. Stroma with infiltrate rich in inflammatory cells, mainly plasma cells (haematoxylin and eosin, 400×)

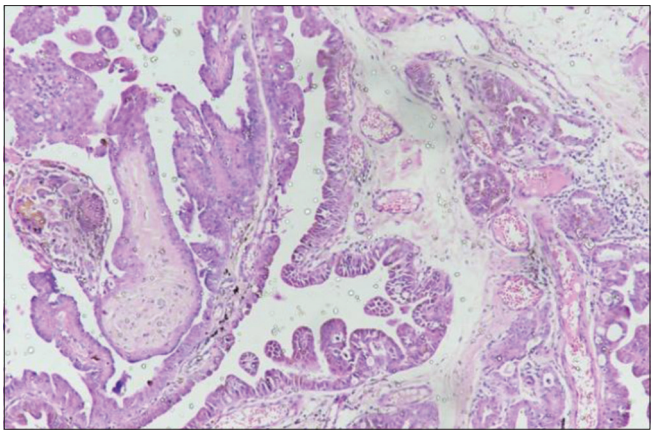

Figure 4: Photomicrograph of the second case showing papillary and cystic structures lined by two layers of cells (haematoxylin and eosin, 40×, 100×)

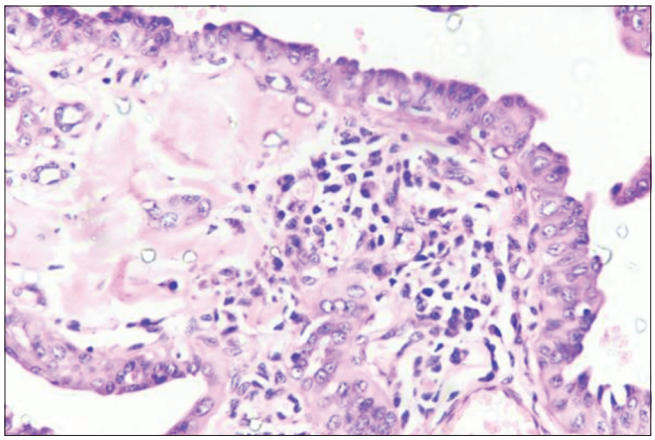

Figure 5: Photomicrograph of the third case shows papillae lined by an inner layer composed of columnar cells and an outer layer formed by cuboidal cells (haematoxylin and eosin, 400×)

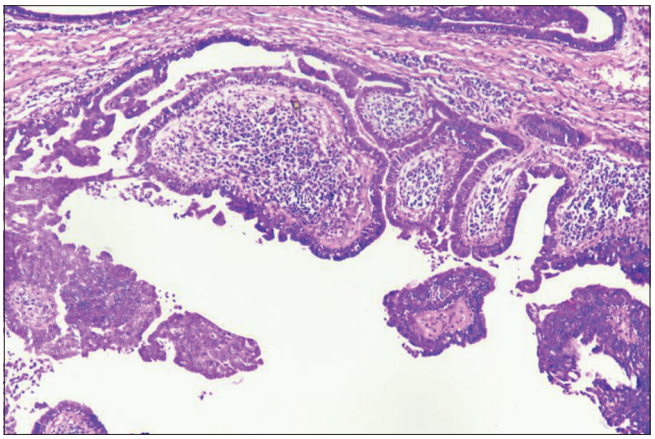

Figure 6: Photomicrograph of the fourth case shows cystic invagination into dermis comprising papillae lined by an inner layer composed of columnar cells and an outer layer formed by cuboidal cells (haematoxylin and eosin, 100×)

It presents usually as skin-coloured to pink, hairless, firm plaque of grouped nodules or as a solitary nodule,[7] most frequently located on the scalp, neck or face.[4] Cauliflower-like, verrucous, papillary, hyperkeratotic or sometimes moist fleshy excrescences have also been described. Some tumours may show central umbilication.[8]

Rare clinical patterns include linear and segmental variants.[9-11] The lesions are often dark brown or red. Unusual locations include the eyelid,[12,13] outer ear canal, pinna, chest, arms,[11] trunk,[14] lower limb,[15,16] thigh,[9] back,[17] gluteal, inguinal,[18] perianal,[19] perineal region, scrotum,[4,20,21] axilla and nipple.[22] Nodular variety is said to occur more frequently on the trunk.[23]

Histogenesis

SCAP occurs *de novo* or in an organoid nevus such as nevus sebaceous of Jadassohn (30%–40% of cases).[24] It is largely considered to be a hamartomatous tumour arising from pluripotent cells, although the histogenesis of this tumour remains disputed as the constituent cells of this tumour have not been fully characterised.[25] The concept of apoeccrine gland has been nurtured as it exhibits microscopic, immunohistochemical and ultrastructural features of both eccrine and apocrine glands.[26,27]

Histopathology findings

It is histologically characterised by the presence of endophytic villous or papillary invaginations of the epithelium into the dermis, forming cystic spaces. These are lined by double-layered cells: outer cuboidal and luminal high columnar epithelium.[28] Stroma of these papillary projections is rich in dilated capillaries and dense plasma cell infiltrates.[29] Similar histopathological findings were noted in all our five cases. Tumour cells show a positive staining reaction with carcinoembryonic antigen, gross cystic disease fluid protein-15, cytokeratin 7, epithelial membrane antigen, androgen receptors and, sometimes, smooth muscle actin-positive myoepithelial layer.[30]

Dermoscopy

Dermoscopy of SCAP arising in nevus sebaceous reveals polymorphous vessels as a prominent feature along with milky

red papillomatous projections.[31-33] There is a single case report of dermoscopy of SCAP arising *de novo* revealing central crater surrounded by white structureless areas and or shiny white lines and an outer rim of brown structureless area. Occasional rosettes and dotted vessels were also mentioned,[34] making it difficult to differentiate from molluscum contagiosum, and hence, there was a need to rely on histopathology for diagnosis confirmation.

Excision biopsy serves the dual role of diagnostic and therapeutic modality in cases of SCAP. CO_2 laser excision of SCAP of the head and neck is another treatment option, which is especially useful in anatomic areas of cosmetic significance or in areas unfavourable to excision and grafting.[2,4] SCAP has also been reported to be treated with Moh's micrographic surgery.[35] Recurrences have been associated with incomplete excision, which was also seen in one of our cases.

Solitary tumours in unusual locations generate multiple differential diagnoses and must be sent for histopathological examination as the clinical presentation can be diverse in morphology, often leading to misdiagnosis as in this case series. The trunk and upper extremity being an unusual site, SCAP was not considered in our cases. Viral wart was considered based on the appearance of the lesion and the long clinical history. Based on the high incidence of tuberculosis in our country, warty tuberculosis was considered as a second possibility. Eroded nodules were considered as pyogenic granuloma; however, they did not bleed on touching. In the case arising within a plaque, DFSP was thought as a possibility. Onset at puberty should alert one about the possibility of unusual location of SCAP. This case series is unique as the cases are all *de novo* in origin and without any other tumour in association.

Conclusion

We have presented five cases which were misdiagnosed clinically owing to the unusual location and variable presentation. These cases may provide further insight into the likelihood of SCAP on unusual locations. This case series may alert the clinicians to consider variable presentations for SCAP and also emphasise the importance of histopathology in diagnosing SCAP arising *de novo* to avoid misdiagnosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for 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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