Well-differentiated Squamous Cell Carcinoma Arising in Syringocystadenoma Papilliferum

Abstract

Syringocystadenoma papilliferum (SCAP) is a hamartoma of apocrine sweat gland origin, occurring predominantly in children, in the head and neck region. SCAP can arise or coexist with both benign and malignant tumors. We report herein an extremely rare case of squamous cell carcinoma (SCC) arising in SCAP growth located in unusual supragluteal region in an elderly Indian male. A wide surgical excision of the growth was performed which upon histopathological examination revealed the confirmatory diagnosis. SCC arising in SCAP is unusual, and only two histologically confirmed cases have been previously reported in the literature. This case report emphasizes about extremely rare malignant transformation of SCAP, which should be kept in mind while evaluating a suspicious skin lesion.

Keywords: *Apocrine, hamartoma, papilliferum, syringocystadenoma*

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Introduction

Syringocystadenoma papilliferum (SCAP) is a rare hamartomatous malformation of apocrine sweat gland which occurs predominantly in infancy or childhood usually involving the head and neck region.[1] SCAP may arise *de novo* without any preexisting lesion; however, it can arise or coexist with both benign and malignant tumors.[2] We report herein an extremely rare case of well-differentiated squamous cell carcinoma (SCC) arising within SCAP growth located in unusual supragluteal region in an elderly Indian male.

Case Report

A 61-year-old otherwise healthy elderly Indian male presented to our surgical outpatient department with a 4-year history of a painless, cutaneous lesion in the lower back which had shown a rapid increase in size for the past 6 months. On clinical examination, the supragluteal region showed a grey-brown color, large, cauliflower-like, polypoidal growth measuring approximately 6.5 × 4.5 × 2.5 cm with normal appearing surrounding skin and free from underlying structure [Figure 1a]. There was no associated lymphadenopathy. Imaging evaluation with magnetic resonance imaging demonstrated a well-defined mushroom-shaped lesion appearing intermediate in signal intensity on T1 and T2, with hypointense stalk limited to superficial skin with no evidence of deep invasion. The lesion appeared hyperintense in fat-suppressed sequence. The routine laboratory investigations were unremarkable. Clinical and radiological findings were suggestive of a malignant cutaneous tumor. A wide local excision was performed [Figure 1b]. Upon histopathological examination, cut surface of the specimen was friable, grey-white with cystic changes, and showed epidermis with papillomatosis with papillary projections extending and invaginating in a cystic space. These papillary invaginations were lined by glandular epithelium with apocrine differentiation, comprising two layers of epithelial cells: inner luminal surface columnar cells with decapitation secretions and basilar outer cuboidal cells [Figure 2]. The outer epidermis of the lesion extended into a growth showing hyperkeratosis, acanthosis with mild dysplasia and invasion in deep dermis by squamous nests, with surrounding dense mononuclear cell infiltrates and desmoplastic reaction, and evidence of keratin pearl formation [Figure 3]. The histopathological findings were consistent

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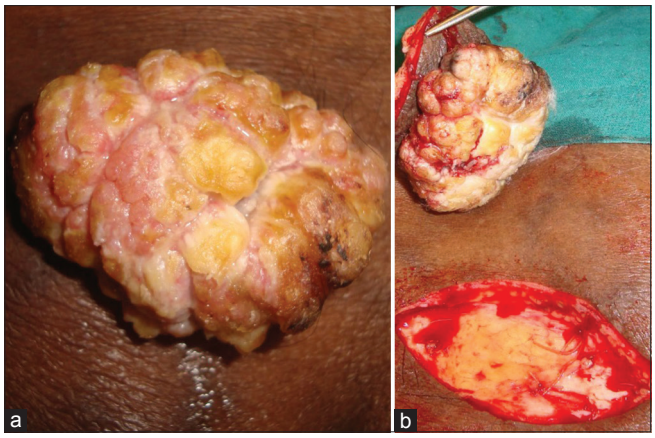

Figure 1: (a) Cauliflower-like ulcerative growth in supragluteal region. (b) Wide local excision for detailed diagnostic and therapeutic purpose

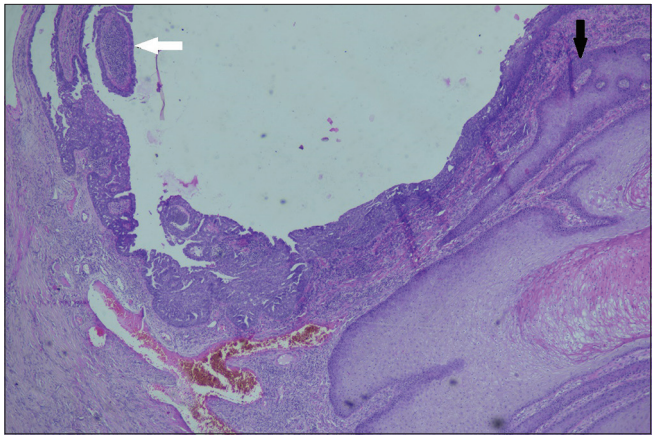

Figure 2: Papillary projections of syringocystadenoma papilliferum invaginating into cystic space (white arrow) and transition of outer epidermis to adjacent well-differentiated squamous cell carcinoma (black arrow) (H and E; ×40)

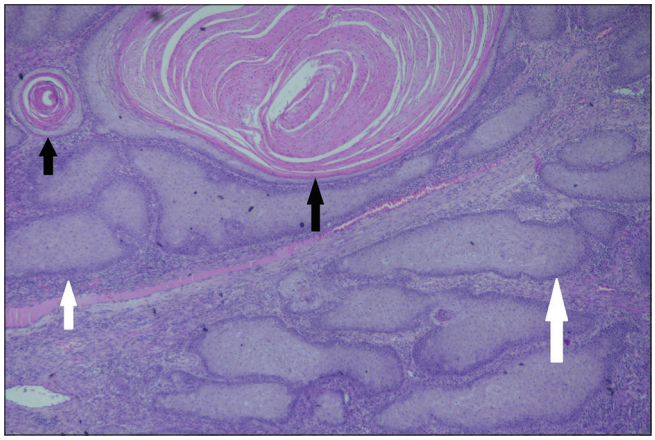

Figure 3: Infiltrating growth of well-differentiated squamous cell carcinoma (black arrow) with surrounding inflammation and keratin pearl (White arrow) formation (H and E; ×100)

with well-differentiated SCC arising within SCAP. The surgical margins of the specimen were clear. There was no evidence of lymphovascular or perineural invasion. The postoperative recovery was uneventful. On follow-up at 3 years, the patient showed no evidence of local recurrence or metastasis.

Discussion

The histopathological findings of excised lesion in the present case were consistent with well-differentiated SCC arising within SCAP. Only two histologically proven cases of SCC arising in SCAP have been described previously in the literature,[3,4] and a wide local excision and follow-up thereafter had shown result similar to the present case with no evidence of recurrence or metastasis. However, in the previous cases the growth was described located in the usual head and neck region in contrast to the present case with unusual supragluteal location. There are two more similar cases available in the literature, but both were not confirmed on histology.[5,6]

SCAP may arise *de novo* without any preexisting lesion as in present case, but more commonly SCAP arises in association with nevus sebaceous.[7] Rarely SCAP can transform into malignancy and the most common associated malignancy is basal cell carcinoma (BCC) occurring in approximately 10% of cases.[7] Grossly, the nodular form of SCAP may sometimes simulate BCC, but histopathologically BCC shows basaloid cells with peripheral palisading, which was lacking in the present case.[8]

Verrucous carcinoma (VC) has been reported arising rarely in association with SCAP.[9] Grossly, it may present as polypoidal growth; however, histopathologically it is characterized by epidermis displaying acanthosis, hyperkeratosis, papillomatosis, and rete ridges with bulbous ends, invading the dermis in a "pushing" pattern, in contrast to the "infiltrative" or invasive pattern, demonstrated in the present case. Unlike SCC, keratin pearl formation is also uncommon in VC.

Pseudoepitheliomatous hyperplasia may also present as nodular or polypoidal growth. Histopathologically, although it may superficially simulate an SCC as it shows elongated downward projections of epidermis with jagged borders, reaching up to the dermis, true invasion is always absent.[10]

SCAP may transform into its rare malignant counterpart syringocystadenocarcinoma papilliferum (SCACP).[11] SCACP with squamous differentiation is also reported.[12] Other less common lesions reported with SCAP include apocrine adenoma, condyloma acuminatum, and hidradenoma papilliferum.[2]

The histogenesis of SCAP is controversial.[13] It is believed to be derived from pleuripotent cells of apocrine sweat glands. Lack of abrupt transition from SCAP to SCC in the present case affirms that it was not a collision tumor and it was only SCAP which has possibly undergone mutations to give rise to SCC.

Conclusion

This is a report of histologically proven SCC arising in SCAP at supragluteal region remarkable for being extremely rare presentation with unusual location. The malignant transformation of SCAP should always be kept in mind while evaluating and complete surgical excision is to be recommended.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

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