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Genital syringocystadenocarcinoma papilliferum: An unusual location and review of the literature

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ABSTRACT

Syringocystadenocarcinoma papilliferum (SCACP) is an extremely rare adnexal neoplasm of the sweat glands. It is believed to arise from the malignant transformation of syringocystadenoma papilliferum (SCAP). The majority of cases present on the head and neck and up to 17% of cases show metastatic progression. These tumors seldom occur in the anogenital area and, to date, only one case has been reported on the penis. Here, we report a rare case of SCACP in a 65-year-old man who presented with an erythematous, non-healing, ulcerated lesion on the penis.

1. Introduction

Syringocystadenocarcinoma papilliferum (SCACP) is a rare malignant adnexal tumor that represents the malignant counterpart of syringocystadenoma papilliferum (SCAP).[1] Mostly, it is believed to arise from SCAP, nevus sebaceous, or linear nevus verrucosus lesions.[2] The majority of cases present on the head and neck and up to 17% of cases show metastatic progression.[1] Only 52 cases of this malignancy have been reported to date, with limited data being available about its origin and etiology. Moreover, these tumors seldom occur in the anogenital area; to date, only one case has been reported to appear on the penis.[2–4] Here, we report a case of SCACP in the anogenital area with penile involvement.

2. Case presentation

A 65-year-old Iranian man presented to the dermatology clinic with multiple papules, nodules, and exudative crust over the genitalia and an erythematous, non-healing, ulcerated lesion on the penis measuring 1 × 2 cm that had appeared two months earlier (Fig. 1). The patient complained of moderate irritating and obstructive urologic symptoms and had unintentionally lost approximately 10 kg of weight during the preceding 6 months. Severe lymphedema and subcutaneous edema were apparent in the scrotum. Skin biopsies were obtained from the skin ulcer and the adjacent papule. Sonography was performed, revealing irregularity in the bladder wall; the prostate volume was 45 ml, with bulging of 10 ml of the median lobe into the base of the bladder. No free fluid was detected in the abdominopelvic cavity. Testicular atrophy was prominent. The scrotal wall was thickened to about 19 mm with increased fat echogenicity in favor of soft tissue inflammation and edema. Bilateral complicated hydrocele was detected. Vascular flow was normal. Cystourethroscopy was performed under spinal anesthesia in the lithotomy position. There were some cotton-like, irregular, epithelial lesions in the distal part of the penile urethra, 2 cm proximal to the fossa navicularis; a cold cup biopsy was taken under vision. Other parts of the urethra had normal appearance and there was a prominent, enlarged prostate with kissing lobes. There was no visible abnormality in the bladder except for moderate trabeculation.

Histopathologic examination revealed hyperplastic epidermis and a crateriform lesion populated by papillary projections and lined by atypical epithelium, with the fibrovascular cores containing numerous plasma cells. Dermal invasion was seen, characterized by tubular

Abbreviations: FDG-PET, fluorodeoxyglucose-positron emission tomography; SCACP, Syringocystadenocarcinoma papilliferum.

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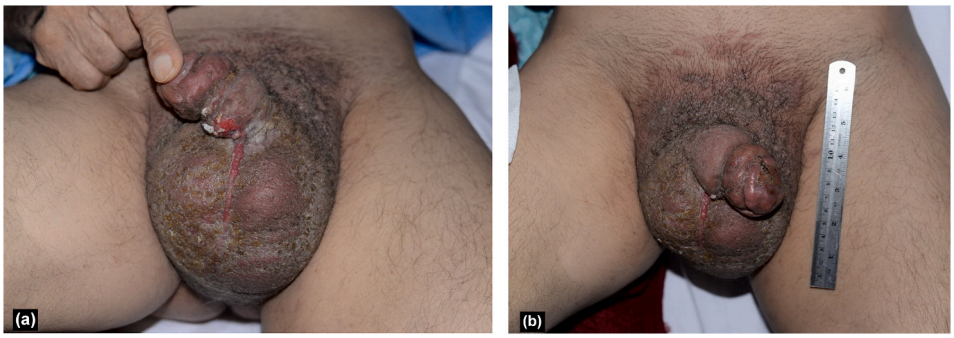

Fig. 1. Multiple papules, nodules, and exudative crust over the genitalia and an erythematous, non-healing, ulcerated lesion on the penis along with severe scrotal edema (a, b).

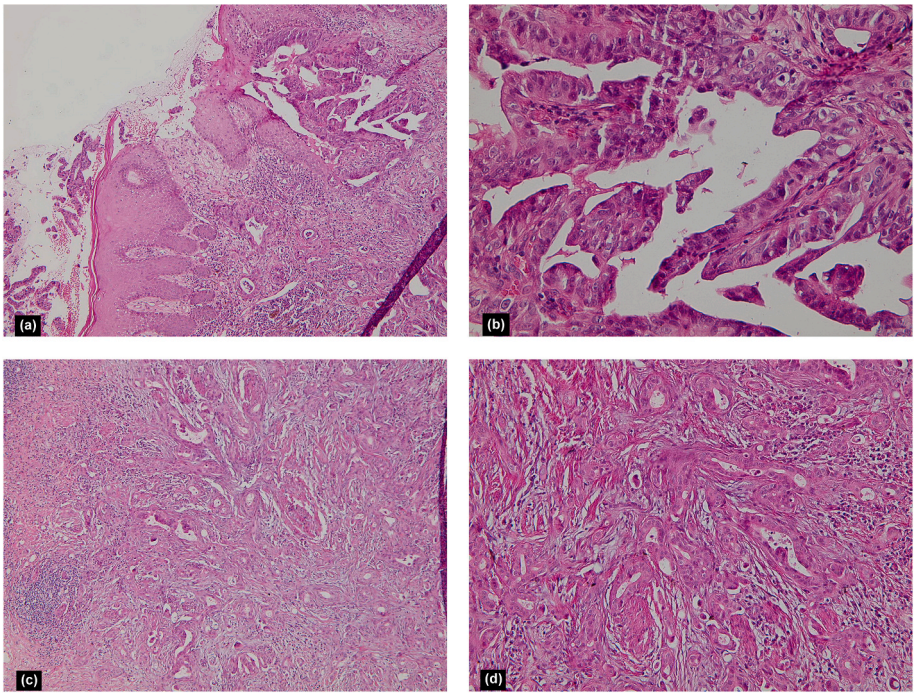

Fig. 2. Hyperplastic epidermis and a crateriform lesion populated by papillary projections and lined by atypical epithelium, with the fibrovascular cores containing numerous plasma cells (a, H&E × 100; b, H&E × 400) (a, b). Dermal invasion was seen, characterized by tubular structures, single cells, and small groups of cells that infiltrated the full thickness of the dermis and part of the subcutis (c, H&E × 100; d, H&E × 200) (c, d).

structures, single cells, and small groups of cells that infiltrated the full thickness of the dermis and part of the subcutis. These structures were surrounded by desmoplastic stroma with lymphoplasmacytic infiltration and perineural invasion. Mild neutrophilic infiltration in dermal nests was identified along with isolated tumor cell necrosis. Overall, the manifestations were characterized as a malignant epithelial neoplasm with papillary and tubular structures in favor of SCACP (Fig. 2). Histologic evaluation of the urethral lesion was also consistent with adenocarcinoma. No distant metastasis was identified via a fluorodeoxyglucose (FDG)-positron emission tomography (PET) scan.

3. Discussion

As an adnexal tumor, SCAP was first described by Stokes in 1917.[1] Dissanayake and Salm, in 1980, were the first to discuss SCACP, which is considered as the malignant counterpart of SCAP.[5] Since then, this is just the 53rd reported case of this malignancy, meaning that the clinical and pathological characteristics are not well defined.[4] We performed a literature review of the Medline, EMBASE, and Cochrane databases to characterize the cases previously listed in the literature (Table 1). Our results show that SCACP predominantly involves the head and neck of patients over 60 years of age with no gender predilection, but can also occur on the chest, arm, anogenital region, and back (Table 1).

These lesions clinically present as erythematous, skin-colored, brown or yellowish nodules, papules, and/or ulcerated lesions that may be associated with mild pain, pruritus, or easily induced bleedings (Table 1). In the present case, the initial differential diagnoses were angiosarcoma, Kaposi sarcoma, and squamous cell carcinoma; however, the histopathology findings were consistent with SCACP.

Currently, no treatment protocols for SCACP are available but surgery with wide margins is the most favorable option and Mohs surgery has been suggested.[1,4] The application of chemotherapy and radiotherapy is controversial.[4] Our therapeutic plan consisted of chemotherapy. Unfortunately, despite receiving five courses of chemotherapy, the patient died seven months after the diagnosis of SCACP.

There have been only eight cases reported with metastasis (Table 1); our patient did not show any signs of metastasis in the PET-FDG scan. On the other hand, there have been limited cases of SCACP associated with invasive adenocarcinoma (Table 1), as was the case for our patient.

To the best of our knowledge, the present case is the second case of

Table 1
Reported cases of syringocystadenocarcinoma papilliferum.

#	Reference	Clinical presentation	Sex/ Age (year)	Location	Size (mm)	Duration	Diagnosis	Association	Follow-up	Treatment
1	Dissanayake and Salm, 1980	Exophytic tumor with copious secretion	F/74	Scalp	65	30 years	SCACP in situ	SCAP	NED (6.75 years)	Surgery
2	Dissanayake and Salm, 1980	A lump	F/71	Back	30	N/A	SCACP invasive	N/A	NED (7 years)	Surgery
3	Seco Navedo et al., 1982	Tumor	F/50	Scalp	65	Congenital	SCACP invasive	Nevus sebaceous	3 Local lymph node, lymph node metastasis	Surgery + RTx + CTx (NED—2 years)
4	Numata et al., 1985	Nodular, partially cystic and solid tumor	F/52	Chest	130 × 80	20 years	SCACP invasive	N/A	1 Local lymph node, lymph node metastasis	Surgery NED (12 months)
5	Bondi and Urso, 1996	Ulcerated and crusted lesion.	M/47	Scalp	25	N/A	SCACP invasive	N/A	N/A	Surgery
6	Ishida-Yamamoto et al., 2001	Enlarging nodule as a black exophytic tumor with a granular surface	M/61	Perianal	60	10 years	SCACP in situ	N/A	NED (11 months)	Surgery
7	Arai et al., 2003	Enlarging tumor, bleed easily when pressed, surrounded by a bloody crust with macerated white papules on the surface	M/64	Scalp	35	2 years	SCACP in situ	SCAP	N/A	Surgery
8	Chi et al., 2004	Two ulcerated verrucous plaques coated with yellow crust, painful, pruritic	M/60	Auricle	40 × 10	Since childhood	SCACP invasive	SCAP	NED (72 months)	Surgery
9	Woestenborghs et al., 2006	A bleeding raised tumor	F/81	Scalp	15	N/A	SCACP in situ	SCAP	N/A	Surgery
10	Park et al., 2007	Single erythematous dome-shaped and firm nodule surrounded by bloody crust	M/65	Suprapubic region	35	2 years	SCACP in situ	N/A	NED (24 months)	Surgery
11	Langner and Ott, 2009	A nodule with partly cystic and partly solid appearance	M/83	Perianal	15	N/A	SCACP in situ	SCAP	N/A	Surgery
12	Sroa et al., 2010	Flesh-colored exophytic nodule with a peripheral crusted hyperpigmented border	M/77	Calf	25	9 years	SCACP invasive	N/A	NED (15 months)	Surgery
13	Kazakov et al., 2010	Verrucous ulcerated nodule	F/56	Neck	20	10 years	SCACP in situ	SCAP	NED (9 months)	Surgery
14	Kazakov et al., 2010	Clinical impression of a ruptured cyst or carcinoma	M/58	Forehead	25	25 years	SCACP invasive	SCAP	NED (4 years)	Surgery
15	Kazakov et al., 2010	Ulcerated smelling neoplasm	F/46	Scalp	35	N/A	SCACP invasive	SCAP	NED (6 years)	Surgery
16	Kazakov et al., 2010	Ulcerated nodule	M/67	Scalp	25	N/A	SCACP in situ	SCAP	NED (2 years)	Surgery
17	Kazakov et al., 2010	Ulcerated tumor with Recent rapid growth.	F/60	Scalp	30	>30 years	SCACP invasive	SCAP	N/A	Surgery
18	Kazakov et al., 2010	Inflammatory plaque	M/81	Scalp	20	N/A	SCACP invasive	SCAP	NED (21 months)	Surgery
19	Leeborg et al., 2010	Erythematous to violaceous asymmetric papule with lobulated contours	F/86	Neck	45	4 months	SCACP invasive	Invasive squamous cell carcinoma	Local recurrence (18 months)	Surgery + RTx
20		As described by the patient, a bead like,	M/62	Axilla	35	6 months	SCACP invasive	N/A	N/A	Surgery

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Table 1 (*continued*)

#	Reference	Clinical presentation	Sex/ Age (year)	Location	Size (mm)	Duration	Diagnosis	Association	Follow-up	Treatment
	Abrari and Mukherjee, 2011	1.0 cm axillary swelling, had been self-discovered and this lesion had excoriated and attained a size of 3.5 cm in 6 months.								
21	Aydin et al., 2011	Ulcerative nodular lesion	M/67	Scalp	40	Since childhood	SCACP invasive	SCAP	NED (2 years)	Surgery
22	Hoekzema et al., 2011	Exophytic nodule with a moist surface	F/83	Arm	30	7 years	SCACP invasive	SCAP nevus verrucosus	N/A	Surgery
23	Hoguet et al., 2012	Erythematous, slightly elevated, centrally ulcerated and crusted nodule	M/86	Eyelid	4	N/A	SCACP invasive	N/A	NED (3 months)	Surgery
24	Plant et al., 2012	A non-healing ulcerated lesion	M/83	Penis	12	N/A	SCACP in situ	N/A	N/A	Surgery
25	Bakhshi et al., 2012	Hemispherical swelling was seen on the scalp which was firm in consistency with a granular surface and erosions and crustations	F/45	Scalp	60 × 30	12 months	N/A	SCAP	NED (12 months)	Surgery
26	Zhang et al., 2012	Solitary tender erythematous ulcerated nodule within a background of red patch	F/75	Arm	15	12 months	SCACP invasive	SCAP	NED (6 months)	Surgery
27	Peterson et al., 2013	Hairless flesh-colored exophytic tumor with serosanguinous exudate	M/65	Scalp	30 × 30	12 months	SCACP invasive	SCAP	NED	Surgery
28	Arslan et al., 2013	Multinodular ulcerated lesions	M/66	Scalp	N/A	20 years	SCACP invasive	SCAP	3 Local lymph node, lymph node metastasis	Surgery + RTx (NED—15 months)
29	Arslan et al., 2013	Well-defined erythematous ulcerated nodule	F/66	Scalp	30	>12 months	SCACP invasive	N/A	NED (2 years)	Surgery
30	Castillo et al., 2014	Well-delimited oval dermal nontender nodule with a solid and cystic appearance.	F/32	Scalp	22	N/A	SCACP in situ	N/A	Local recurrence (8 years)	Surgery
31	Paradiso et al., 2014	A single painful erythematous, dome-shaped, and firm nodule surrounded by normal skin	M/88	Shoulder	15 × 15	N/A	SCACP invasive	N/A	Died from other cause	N/A
32	Shan et al., 2014	A pink ulcerated nodule without tenderness nor pruritis	M/93	Popliteal fossa	20	>10 years	N/A	SCAP	NED	Surgery
33	Mohanty et al., 2014	Exophytic lobulated mass which was tan-pink to red in color with soft to firm consistency and was non-tender.	F/80	Scalp	50	8 years	SCACP in situ	N/A	NED (5 years)	Surgery
34	Satter et al., 2014	Focally ulcerated exophytic nodule associated with a few small satellite papules which bled with minor trauma	M/42	Scalp	45 × 40	>1 month	SCACP invasive	SCAP and Nevus sebaceous	Lymph node metastasis	Surgery
35	Parekh et al., 2016	A single well-demarcated red	M/74	Scalp	20	Since childhood	SCACP invasive	SCAP, nevus sebaceous of	Lymph node metastasis	Surgery

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Table 1 (*continued*)

#	Reference	Clinical presentation	Sex/ Age (year)	Location	Size (mm)	Duration	Diagnosis	Association	Follow-up	Treatment
		exophytic nodule with small foci of ulceration						Jadassohn, trichoblastoma		
36	Chen et al., 2016	A hairless, rough, ill-defined erythematous erosive warty plaque on the right parietal scalp with serosanguinous exudate	F/60	Scalp	28 × 20	12 months	SCACP in situ	Nevus sebaceous	N/A	Surgery
37	Singh et al., 2017	Well-defined erythematous plaque with overlying ulceration on the midback, 1 cm left of the spine. Adjoining skin showed a horseshoe-shaped oval area of lichenification	F/60	Back	15 × 10	>10 years	SCACP in situ	SCACP in situ with macular amyloidosis	N/A	Surgery
38	Zhang et al., 2017	Solitary red plaque	M/26	Chest	50	22 years	SCACP in situ	Invasive adenocarcinoma subcutis	Left axillary lymph node and bilateral lung metastases, DoD 2 months after diagnosis	Surgery + CTx
39	Zhang et al., 2017	Solitary nodule with exudate	M/47	Abdomen	15	23 years	SCACP in situ	N/A	NED (9 years)	Surgery
40	Zhang et al., 2017	Erythematous nodule	M/67	Left Axilla	20	6 years	SCACP in situ	Invasive adenocarcinoma subcutis	N/A	Surgery + left axilla lymphadenectomy
41	Zhang et al., 2017	Flat verrucous neoplasm	M/64	Scalp	20	1 years	SCACP in situ	Invasive adenocarcinoma in dermis + mucinous metaplasia	Metastases to multiple distant lymph nodes and lung metastases, DoD 34 months after diagnosis	Surgery + RTx
42	Zhang et al., 2017	Exophytic ulcerated nodule with bleeding	M/63	Chest	10	10 years	SCACP in situ	Invasive adenocarcinoma in dermis	NED (36 months)	Surgery
43	Zhang et al., 2017	Exophytic pinkish nodule	M/74	Chest	20	6 years	SCACP in situ	Invasive adenocarcinoma subcutis	NED (30 months)	Surgery
44	Zhang et al., 2017	Flat mass with granular surface	F/63	Axilla	50	3 months	SCACP in situ	Invasive adenocarcinoma + invasive squamous cell carcinoma	Widespread subcutaneous metastases, DoD 20 months after diagnosis	Surgery + right axilla lymphadenectomy
45	Zhang et al., 2017	Keloid plaque	M/40	Chest	50	5 years	SCACP in situ	Invasive adenocarcinoma subcutis	NED (14 months)	Surgery + bilateral lymphadenectomy + CTx
46	Zhang et al., 2017	Subcutaneous nodule	F/29	Forehead	15	2 years	SCACP in situ	Invasive squamous cell carcinoma	NED (10 months)	Surgery
47	Zhang et al., 2017	Verrucous ulcerated mass	M/64	Axilla	22	10 years	SCACP in situ	Invasive adenocarcinoma subcutis	NED (3 months)	Surgery + right axilla lymphadenectomy + CTx
48	Muthusamy et al., 2017	Ulcerated nodule with mild pain	F/78	Scalp	45 × 35	6 months	SCACP invasive	SCAP and Trichoblastoma	N/A	N/A
49*	Yao et al., 2018	Erythematous friable mass which bleed intermittently	F/73	Scalp	30 × 27	Since birth	N/A	N/A	N/A	N/A
50	Agulló Pérez et al., 2018	Pediculated erythematous-brownish papule which occasional bleeding caused by	M/40	Chest	10	1 year	SCACP invasive	N/A	NED (8 months)	Surgery

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Table 1 (*continued*)

#	Reference	Clinical presentation	Sex/ Age (year)	Location	Size (mm)	Duration	Diagnosis	Association	Follow-up	Treatment
		friction and exudative crust								
51	Pagano Boza et al., 2019	Ulcerated, indurated, and erythematous nodule	M/63	Eyelid	50 × 70	>6 years	SCACP invasive	SCAP	Local recurrence	Surgery
52	Alegría-Landa et al., 2019	Eroded easily-bleeding nodule	F/90	Scalp	N/A	10 months	SCACP in situ with sarcomatoid appearance	N/A	Died from other cause 1 year after diagnosis	N/A
53	Present case	Non-healing erythematous ulcerated lesion, multiple papules and nodules, crustations and hyperpigmented patches. Scrotal Tense edema.	M/65	Genitalia	10× 20	2 months	SCACP invasive	Invasive Adenocarcinoma	Died from other cause 7 months after diagnosis	CTx

Abbreviations: CTx, chemotherapy; DoD, died of disease; F, Female; M, Male; N/A, not available; NED, no evidence of disease; RTx, radiation therapy.
*Full text was not accessible online

SCACP with involvement of the penis, the first involving the scrotum, and one of the limited cases associated with invasive behavior of SCACP.

In conclusion, regarding the variable course of SCACP, in case of encountering chronic nodular and ulcerative lesions in the genital area, despite the rarity, clinicians should be aware of its possibility.

Ethics approval and consent to participate

Ethical approval was not required for this study in accordance with national guidelines.

Consent for publication

Written informed consent was obtained from the patient's family for publication of this case report and any accompanying images.

Declaration of competing interest

The authors have no conflicts of interest to declare.

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Author contributions

MSD, FA and AA, were involved in the diagnosis and management of the patients and have been responsible for the clinical part of the manuscript. AR reported the result of histopathological evaluation. FA and MB did literature review and drafted the manuscript. MSD, FA, AR and AA were responsible for final editing of the manuscript, and coordinated the study. All authors have read and approved the final manuscript.

Availability of data and material

Not applicable.

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