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Syringocystadenocarcinoma papilliferum with orbital invasion: a case report with literature review

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Abstract: We present a case of Syringocystadenocarcinoma papilliferum that originated in the eyelid and extended into the orbit. These tumors are very rare and have the potential to metastasize. A literature review of all the previous cases has been compiled from the Medline, EMBASE, and PubMed databases. We found that the majority of cases present on the head and neck and up to 17% of cases showed metastatic progression. This is the first case to show orbital involvement and highlights the need to remain vigilant with such lesions, as they have a tendency to become aggressive.

Keywords: eyelid, orbit, Syringocystadenocarcinoma papilliferum

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Introduction

Syringocystadenocarcinoma papilliferum (SCACP) is a rare malignant sudoriferous gland tumor that is related to its more common, benign counterpart, syringocystadenoma papilliferum (SCAP). Since the original description of SCAP in 1917,[1] only 43 cases of SCACP have been described in the literature. To date, only one has appeared in the eyelid.[2] SCACP is thought to develop from SCAP, nevus sebaceous, and linear nevus verrucosus lesions.[3] However, due to the rarity of this tumor, little is known regarding its etiology and origin.[3]

In this study, we report the first case of SCACP with orbital involvement. Interestingly, it recurred following exenteration. An informed written consent was obtained from the patient for the publication of medical data and images.

Case report

A 63-year-old man presented with a lesion on the right upper eyelid that had been present for 7 years. The lesion was nodular, measuring 5.0 cm × 7.0 cm, ulcerated, indurated, and erythematous. It involved the lower eyelid (Figure 1). The patient had no light perception with the right eye and had a visual acuity of 20/20 on the left. Due to the presence of the tumor over the right eye, his intraocular pressure could not be measured, and it was found to be 18 mmHg on the left.

The left orbital examination did not reveal any abnormalities. A full examination of his local lymph nodes and lacrimal duct did not reveal any abnormalities. He explained that he did not have any previous therapy for this lesion. He was otherwise systemically well with no relevant family history. He did not have any history of trauma and informed us that he was a farmer by occupation.

A computed tomography (CT) scan of the orbit revealed right anterior orbital invasion with no bony or lacrimal gland involvement (Figure 2). A subsequent incisional biopsy revealed squamous cell invaginations extending from the epidermis into the dermis. The invaginations and papillary projections were lined with a bilayer epithelium: the luminal layer was composed of columnar cells with decapitation secretion and the outer layer was composed of small cuboidal cells. These cells had significant nuclear pleomorphism, prominent nucleoli, and increased mitotic activity (Figure 3). Immunohistochemical staining demonstrated positivity for epithelial membrane antigen (EMA),

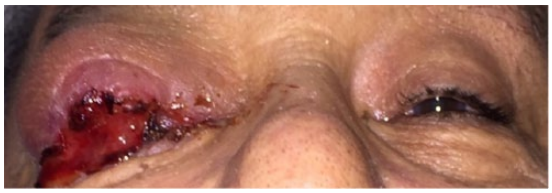

Figure 1. Lesion on presentation.

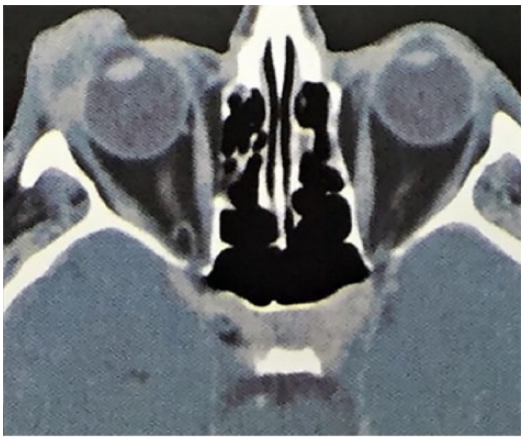

Figure 2. CT imaging of the lesion at presentation.

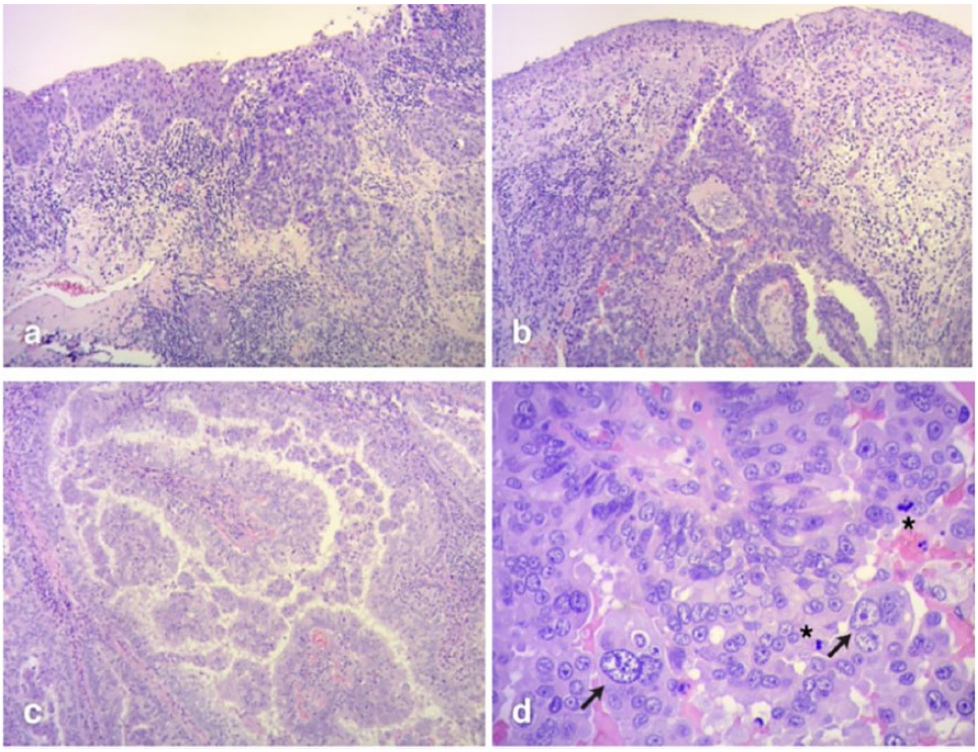

Figure 3. Hematoxylin and eosin staining (H&E): (a) the transition between squamous and glandular epithelium (100×). (b) Large areas of superficial epithelium were sphacelated. Glandular invaginations showed a characteristic funnel shape. Papillary structures can be identified inside a dermal cyst (100×). (c) The papillary structures are lined with a stratified atypical epithelium. Micropapillae and secretion by decapitation can be seen (100×). (d) At high power magnification, atypical nuclei are evident. Large atypical nuclear shapes are seen and increased mitotic activity is observed (*).

Table 1. Previous case reports on SCACP.

Reference	Age	Sex	Location	Size (mm)	Duration	Diagnosis	Association	Follow-up	Treatment
Dissanayake and Salm[5]	74	F	Scalp	65	30 years	SCACP in situ	SCAP	NED (6.75 years)	Surgery
	71	F	Back	30	N/A	SCACP invasive	N/A	NED (7 years)	Surgery
Seco Navedo and colleagues[6]	50	F	Scalp	65	Congenital	SCACP invasive	Nevus sebaceous	3 Local lymph node, lymph node metastasis	Surgery + Rt + Ct (NED—2 years)
Numata and colleagues[7]	52	F	Chest	130 × 80	20 years	SCACP invasive	N/A	1 Local lymph node, lymph node metastasis	Surgery NED (12 months)
Bondi and Urso[8]	47	M	Scalp	25	N/A	SCACP invasive	N/A	N/A	Surgery
Ishida-Yamamoto and colleagues[9]	61	M	Perianal	60	10 years	SCACP in situ	N/A	NED (11 months)	Surgery
Arai and colleagues[10]	64	M	Scalp	35	2 years	SCACP in situ	SCAP	N/A	Surgery
Chi and colleagues[11]	60	M	Auricle	40 × 10	Since childhood	SCACP invasive	SCAP	NED (72 months)	Surgery
Woestenborghs and colleagues[12]	81	F	Scalp	15	N/A	SCACP in situ	SCAP	N/A	Surgery
Park and colleagues[13]	65	M	Suprapubic region	35	2 years	SCACP in situ	N/A	NED (24 months)	Surgery
Langner and Ott[14]	83	M	Perianal	15	N/A	SCACP in situ	SCAP	N/A	Surgery
Sroa and colleagues[15]	77	M	Calf	25	9 years	SCACP invasive	N/A	NED (15 months)	Surgery
Kazakov and colleagues[16]	56	F	Neck	20	10 years	SCACP in situ	SCAP	NED (9 months)	Surgery
	58	M	Forehead	25	25 years	SCACP invasive	SCAP	NED (4 years)	Surgery
	46	F	Scalp	35	N/A	SCACP invasive	SCAP	NED (6 years)	Surgery
	67	M	Scalp	25	N/A	SCACP in situ	SCAP	NED (2 years)	Surgery
	60	F	Scalp	30	>30 years	SCACP invasive	SCAP	N/A	Surgery
	81	M	Scalp	20	N/A	SCACP invasive	SCAP	NED (21 months)	Surgery

(Continued)

Table 1. (Continued)

Reference	Age	Sex	Location	Size (mm)	Duration	Diagnosis	Association	Follow-up	Treatment
Leeborg and colleagues[17]	86	F	Neck	45	4 months	SCACP invasive	Invasive squamous cell carcinoma	Local recurrence (18 months)	Surgery + Rt
Abrari and Mukherjee[18]	62	M	Axilla	35	6 months	SCACP invasive	N/A	N/A	Surgery
Aydin and colleagues[19]	67	M	Scalp	40	Since childhood	SCACP invasive	SCAP	NED (2 years)	Surgery
Hoekzema and colleagues[3]	83	F	Arm	30	7 years	SCACP invasive	SCAP nevus verrucosus	N/A	Surgery
Hoguet and colleagues[20]	86	M	Eyelid	4	N/A	SCACP invasive	N/A	NED (3 months)	Surgery
Plant and colleagues[21]	83	M	Penis	12	N/A	SCACP in situ	N/A	N/A	Surgery
Bakhshi and colleagues[22]	45	F	Scalp	60 × 30	12 months	N/A	SCAP	NED (12 months)	Surgery in situ
Zhang and colleagues[23]	75	F	Arm	15	12 months	SCACP invasive	SCAP	NED (6 months)	Surgery
Peterson and colleagues[24]	65	M	Scalp	30x30	12 months	SCACP invasive	SCAP	NED	Surgery
Arslan and colleagues[2]	66	M	Scalp	N/A	20 years	SCACP invasive	SCAP	3 Local lymph node, lymph node metastasis	Surgery + Rt (NED—15 months)
	66	F	Scalp	30	>12 months	SCACP invasive	N/A	NED (2 years)	Surgery
Castillo and colleagues[25]	32	F	Scalp	22	N/A	SCACP in situ	N/A	Local recurrence (8 years)	Surgery
Paradiso and colleagues[26]	88	M	Shoulder	15 × 15	N/A	SCACP invasive	N/A	Died from other cause	N/A
Shan and colleagues[27]	93	M	Popliteal fossa	20	>10 years	N/A	SCAP	NED	Surgery
Mohanty and colleagues[28]	80	F	Scalp	50	8 years	SCACP in situ	N/A	NED (5 years)	Surgery
Satter and colleagues[29]	42	M	Scalp	45 × 40	>1 month	SCACP invasive	SCAP and Nevus sebaceous	Lymph node metastasis	Surgery
Parekh and colleagues[4]	74	M	Scalp	20	Since childhood	SCACP invasive	SCAP, nevus sebaceous of Jadassohn, trichoblastoma	Lymph node metastasis	Surgery

Table 1. (Continued)

Reference	Age	Sex	Location	Size (mm)	Duration	Diagnosis	Association	Follow-up	Treatment
Chen and colleagues[30]	60	F	Scalp	28 × 20	12 months	SCACP in situ	Nevus sebaceous	N/A	Surgery
Singh and colleagues[31]	60	F	Back	15 × 10	>10 years	SCACP in situ	SCACP in situ with macular amyloidosis	N/A	Surgery
Zhang and colleagues[32]	26	M	Chest	50	22 years	SCACP in situ	Invasive adenocarcinoma subcutis	Left axillary lymph node and bilateral lung metastases, DOD 2 months after diagnosis	Surgery + Ct
	47	M	Abdomen	15	23 years	SCACP in situ	N/A	NED (9 years)	Surgery
	67	M	Left Axilla	20	6 years	SCACP in situ	Invasive adenocarcinoma subcutis	N/A	Surgery + left axilla lymphadenectomy
	64	M	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 + Rt
	63	M	Chest	10	10 years	SCACP in situ	Invasive adenocarcinoma in dermis	NED (36 months)	Surgery
	74	M	Chest	20	6 years	SCACP in situ	Invasive adenocarcinoma subcutis	NED (30 months)	Surgery
	63	F	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
	40	M	Chest	50	5 years	SCACP in situ	Invasive adenocarcinoma subcutis	NED (14 months)	Surgery + bilateral lymphadenectomy + Ct
	29	F	Forehead	15	2 years	SCACP in situ	Invasive squamous cell carcinoma	NED (10 months)	Surgery
	64	M	Axilla	22	10 years	SCACP in situ	Invasive adenocarcinoma subcutis	NED (3 months)	Surgery + right axilla lymphadenectomy + Ct
Present case	63	M	Eyelid	50 × 70	>6 years	SCACP invasive	SCAP	Local recurrence	Surgery

Ct, chemotherapy; Rt, radiation therapy; N/A, not available; NED, no evidence of disease; DOD, died of disease.

Cytokeratin 8/18, and a Cytokeratin cocktail of high and low density (Figure 3). It was negative for GCDFP-15 (protein 15 of the fibrocystic disease of the breast), which excluded a lesion of breast origin and carcinoembryonic antigen (CEA). The diagnosis of SCACP was therefore confirmed. A positron emission tomography (PET) scan did not reveal any metastatic spread.

The patient was treated with exenteration of the right orbit to remove the tumor. After 11 months of follow-up, we noted local recurrence of the original tumor (confirmed with biopsy) in the anophthalmic orbit. There was no associated lymph node enlargement on examination, though the patient refused any further imaging. Radical exenteration with adjuvant radiotherapy has been planned for the patient.

Discussion

SCACP is an extremely rare adnexal neoplasm of the sweat glands and has only been documented 43 times in the literature. It is believed to arise from a malignant transformation of SCAP lesions.[4] Clinically, it may present as an asymptomatic long-standing lesion, which may be flat or nodular, cystic, or ulcerated. We performed a literature review of the Medline, EMBASE, and Cochrane databases to characterize the cases previously listed in the literature (Table 1).

The tumor appears to affect middle-aged or elderly individuals[15] and does not seem to have a gender bias. The most frequent location is the head and neck (53%), with only one case in the eyelid. Other locations where these lesions occur frequently are the back, chest, suprapubic, and perianal regions.

Treatment is based on a complete tumor resection with oncological margins, which is essential for a better prognosis. Mohs surgery has also been successfully used for this purpose.[11] Sentinel lymph node biopsy may be feasible in some cases when there is suspicion of lymph spread, although lymphatic spread has been shown to be rare with this tumor (6 of the 42 documented cases; Table 1). Radiotherapy and chemotherapy have also been used rarely, but the experience with these treatments is scarce due to the rarity of the lesion.[25]

SCACP characteristically presents with squamous cell invaginations extending from the epidermis into the dermis. The invaginations and papillary projections are lined by two-layer epithelium: the luminal layer composed of columnar cells with decapitation secretions and the outer layer composed of small cuboidal cells. The immunohistochemical features of SCACP are still under study, but the most frequently reported markers are CEA,[15,20,28] followed by EMA,[9,28] GDFP-15,[20,28,32] and cytokeratin.[11,28,32]

Due to its appearance, the differential diagnosis includes other skin tumors such as basal cell carcinoma, squamous cell carcinoma, sebaceous carcinoma, metastatic breast or gastrointestinal adenocarcinomas, and other sweat gland neoplasms.[2,20]

Of the cases that reported head and neck involvement, 16 (72.72%) were in remission following therapy, 2 (9.09%) had local recurrence, 3 (13.63%) had regional lymphatic invasion, and 1 (4.54%) had distant metastases. Of the reports describing involvement of the thorax, abdomen, and pelvis, 17 (85%) went into remission following therapy, none had local recurrence, 1 (5%) had regional lymphatic invasion and 2 (10%) had distant metastases.

This is the first reported case of SCACP with extension into the anterior orbit. While SCACP is an exceedingly rare tumor, we found that of the reported cases, 16% showed signs of metastasis. It is therefore an important diagnosis to consider when reviewing skin lesions around the orbit. It also encourages us to monitor patients with SCAP more closely as our literature review suggests that SCACP may be more aggressive than previously considered.

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Conflict of interest statement

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