Case Report

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Pathology Section

# Syringocystadenocarcinoma Papilliferum of Arm: A Rare Case Report

S.V.SATYANARAYANA RAO, NEELIMA TIRUMALASETTI

## ABSTRACT

We report a rare case of syringocystadenocarcinoma papilliferum (SCACP) associated with syringocystadenoma papilliferum(SCAP). A 53-year-old female came with a complaint of swelling over medial aspect of right arm since 7yrs. The lesion was asymmetrical with numerous papillary projections covering the surface. Histologically, the tumour was similar to SCAP except for nuclear atypia and stratification. Our literature review shows that the histologic and immunohistochemical features of SCACP are not well defined with a clear morphologic overlap with SCAP. Wide surgical excision is the treatment of choice. These tumours show regional lymphnode metastases but no distant metastases.

**Keywords:** Apocrine differentiation, Syringocystadenoma papilliferum, Syringocystadenocarcinoma papilliferum

## INTRODUCTION

Syringocystadenocarcinoma papilliferum (SCACP) is the rare malignant counterpart of a more common, benign adnexal neoplasm known as syringocystadenoma papilliferum (SCAP) [1].

Most lesions seem to have arisen from long-standing SCAP [2]. They are most common on the head and neck of middle-aged or elderly individuals but is also reported in the skin of the scalp, back, chest, suprapubic area and perianal region [1].To our knowledge only 13 cases have been reported in the literature till now [1].

No case of SCACP has been reported in the arm. Herein, we report a case of SCACP occurring on the upper one third of right arm in a 53-year-old female patient.

## CASE REPORT

A 53-year-old female came with a complaint of swelling over medial aspect of right arm since 7yrs. The lesion began as a small papule and later gradually increased in size. She also complained of rapid increase in the size of the lesion in the past 2months along with some discharge from the lesion. On examination, the lesion was asymmetrical with numerous papillary projections covering the surface. Regional lymphnodes were not palapable.

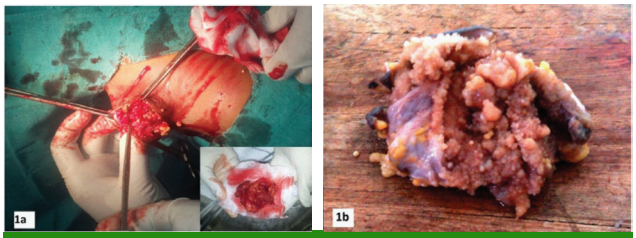
Ultrasound of the lesion showed a 4x2.5cm subcutaneous swelling with solid and cystic components in the medial aspect of right arm. Wide local excision was done [Table/Fig-1a] and the specimen was sent for histopathological examination [Table/Fig-1b].

Histopathology revealed a tumour exhibiting papillomatous growth on the epidermis. Papillary projections were lined by a two-layered epithelium upon a connective tissue core heavily infiltrated by lymphocytes and plasma cells. The outer layer was composed of cuboidal cells, and the inner luminal layer was composed of columnar cells [Table/Fig-2].

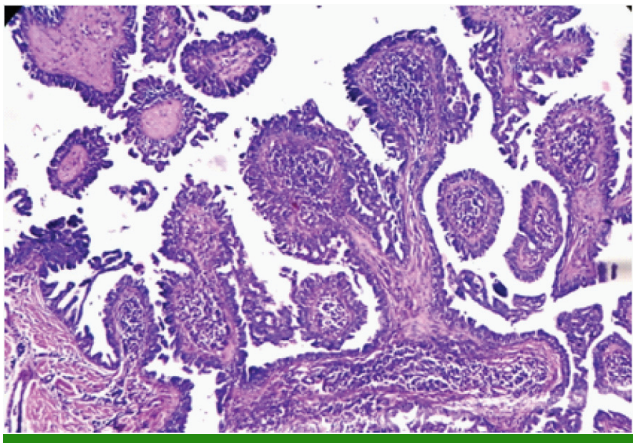
Rest of the papillae displayed a disorganised proliferation of atypical cells with eosinophilic cytoplasm, enlarged nuclei, prominent nucleoli and scattered mitoses [Table/Fig-3].

Upper parts of few papillae are lined by squamous epithelium. Apical snouting of cells lining the papillae were observed [Table/Fig-4]. Stroma showed few apocrine glands lined by atypical nuclei [Table/Fig-5]. From the above findings, a diagnosis of SCACP associated with SCAP was made.

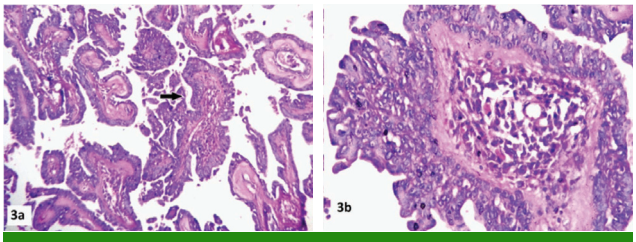
Mammography of both the breasts, ultrasound of neck,




**[Table/Fig-1a]:** Shows peroperative excision of tumor from medial aspect of right upper arm. Inset showing the excised tumour
**[Table/Fig-1b]:** Shows tumour with numerous papillary projections on the skin surface



**[Table/Fig-2]:** Shows a tumor exhibiting papillomatous growth with papillary projections lined by a two-layered epithelium upon a connective tissue core heavily infiltrated by lymphocytes and plasma cells.(H&E,100x)




**[Table/Fig-3a&b]:** Low and high power views showing papillae with disorganized proliferation of atypical cells with eosinophilic cytoplasm, enlarged nuclei, prominent nucleoli and scattered mitoses.(H&E,100x &400x)

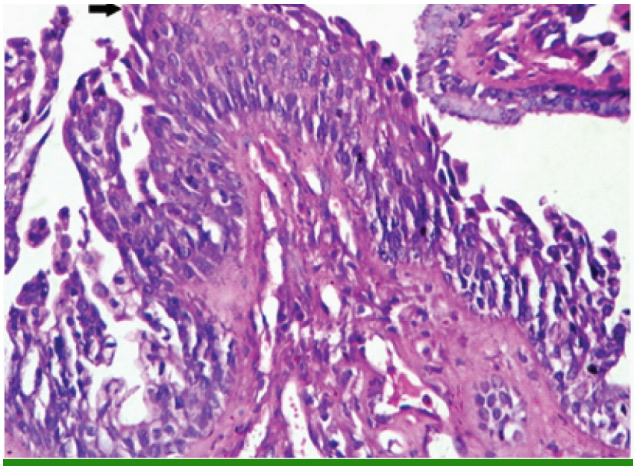
abdomen and pelvis were normal.

## DISCUSSION

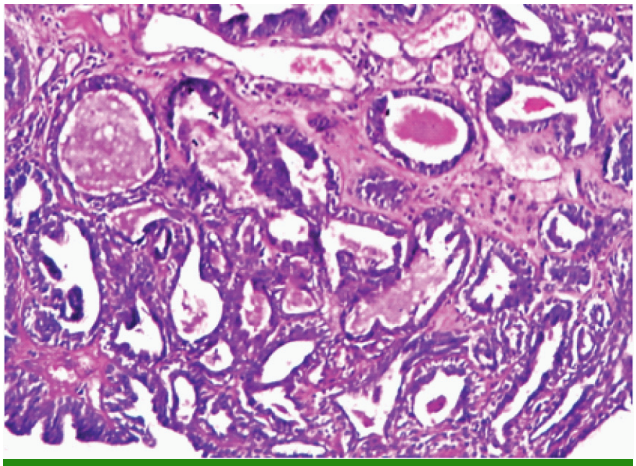
SCACP is a rare neoplasm of apocrine glands and is the malignant counterpart of the more common benign syringocystadenoma papilliferum [1]. The period from the formation of SCAP to the formation of SCACP ranges from 20 to 50 years [3]. Clinically, the lesion shows skin-colored or yellowish papules or nodules that remain unchanged for many years but then begin to enlarge suddenly with bleeding or ulceration [4]. Usually the lesion is covered with crusts as a consequence of secretion of apocrine epithelial cells[4]. As in the present case, the skin surface may be papillary or exophytic as the name papilliferum implies [5]. Its size varies from 2.5 to 13 cm [4].

Histopathologically, SCACP has many structural similarities with SCAP, but it can be differentiated from SCAP in that it has an asymmetric and poorly circumscribed structure of tumour, often extending deep into the subcutaneous fat, and atypical cells, many of which are in mitosis [2].

Arai et al., stated various findings supporting the origin of SCACP from the apocrine glands. They are: decapitation on



**[Table/Fig-4]:** Showing upper part of papilla lined by squamous epithelium. Apical snouting of cells lining the papilla are noted.(H&E, 400x)



**[Table/Fig-5]:** Stroma showed apocrine glands lined by atypical nuclei (H&E, 100x)

the luminal surface of inner layer cells, association with follicles or sebaceous glands and presence of apocrine glands in the underlying tissue. In the present case, decapitation/ apical snouting and presence of apocrine glands in the underlying tissue with discharge from the lesion support the hypothesis that SCACP originates from apocrine glands [3]. SCACP may mimic a metastatic malignancy to the skin. A metastatic lesion is far more likely, and the differential diagnosis includes metastatic tumours from breast, gastrointestinal tract and thyroid [2,6,7].

There is no definitive immunoprofile that can discriminate between a primary cutaneous and a metastatic adenocarcinoma [1]. Loss of immunoreactivities of Carcino embrogenic antigen (CEA) and gross cystic disease fluid protein (GCDFP) is associated with the malignant de-differentiation. Immunohistochemically, positivity for GCDFP, lysozyme, and Leu M1 are characteristics of apocrine differentiation. Further

study is necessary to determine a definitive marker of apocrine differentiation [8].

The present case is a rare tumour peculiar in its site, arising from medial aspect of arm. This is the first reported case of SCACP in the arm [3].

The surgical pathologist should recognise this rare neoplasm because a correct diagnosis may affect patient treatment and prognosis. The reported patients with SCACP have done well with surgical excision alone, with only two reported cases of regional lymph node metastases and no distant metastases.

## CONCLUSION

SCACP is a rare neoplasm of apocrine glands commonly present in the middle aged/ elderly. Surgical excision is the treatment of choice with rarely reported regional lymph node metastases.

## REFERENCES

[1] Nicky Leeborg, Michele Thompson, Sarah Rossmiller, Neil Gross, Clifton White, Ken Gatter. Diagnostic Pitfalls in Syringocystadenocarcinoma Papilliferum- Case Report and Review of the Literature. *Arch Pathol Lab Med*. 2010;134:1205-09.

[2] Park SH, Shin YM, Shin DH, Choi JS, Kim KH. Syringocystadenocarcinoma papilliferum: a case report. *J Korean Med Sci.* 2007;22:762–65.

[3] Arai Y, Kusakabe H, Kiyokane K. A case of syringocystadenocarcinoma papilliferum in situ occurring partially in syringocystadenoma papilliferum. *J Dermatol*. 2003;30:146-50.

[4] Requena L, Kiryu H, Ackerman AB. Ackerman's Histologic Diagnosis of Neoplastic Skin Disease: A Method by Pattern Analysis. Neoplasms with Apocrine Differentiation. *Philadelphia, PA: Lippincott-Raven* 1998: 665-75.

[5] Ishida-Yamamoto A, Sato K, Wada T, Takahashi H, Iizuka H. Syringocystadenocarcinoma papilliferum: case report and immunohistochemical comparison with its benign counterpart. *J Am Acad Dermatol*. 2001;45:755–59.

[6] Numata M, Hosoe S, Itoh N, Munakata Y, Hayashi S, Maruyama Y. Syringadenocarcinoma papilliferum. *J Cutan Pathol.* 1985;12:3–7.

[7] Chi CC, Tsai RY, Wang SH. Syringocystadenocarcinoma papilliferum: successfully treated with Mohs micrographic surgery. *Dermatol Surg.* 2004;30:468–71.

[8] Seco Navedo MA, Fresno Forcelledo M, Orduna Domingo A, Junco Petrement P, Soler Sanchez T. Syringocystadenoma papilliferum with malignant evolution: presentation of a case. *Ann Dermatol Venereol*. 1982;109:685–89.

**AUTHOR(S):**

1. Dr. S.V.Satyanarayana Rao
2. Dr. Neelima Tirumalasetti

**PARTICULARS OF CONTRIBUTORS:**

1. Associate Professor, Department of Genral Surgery, Katuri Medical College and Hospital, Guntur, India.
2. Assistant Professor, Department of Pathology, Katuri Medical College and Hospital, Guntur, India.

**NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:**

Dr. Neelima Tirumalasetti,
D/O T.Lenin,
Door No: 23-34-39A, Manepallivari street,
Satyanarayanapuram, Vijayawada- 520011, India.
Ph. : 9642089268
E-mail : nilu_medico@yahoo.co.in

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