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CASE REPORT

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Syringocystadenocarcinoma papilleferum: A very rare tumor of skin, thirteith case

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Abstract

Syringocystadenocarcinomapapilliferum (SCACP) is a very rare tumor of skin and only 29 cases have been reported in literature. Most of the cases arise from preexisting benign counter part of the tumor: Syringocystadenopapilliferum (SCAP). We report a case of SCACP in a 48 male patient who had a nodular lesion on the scalp for 30 years. The lesion which was initially small continued to grow overtime. He was diagnosed as SCAP on incisional biopsy four years back, and presented with ulceration and bleeding from the nodule for 1 year. After complete excision with clear margins, the case was diagnosed as SCACP based on characteristic papillomatous areas, in-situ component and invasive areas, all in the same tumor.

Keywords: *syringocystadenocarcinoma papilliferum, syringocystadenomapapilleferum, skin adnexal neoplasm, nodule*

Introduction

Syringocystadenocarcinoma papilliferum (SCACP) is a very rare, malignant skin adnexal neoplasm, of uncertain origin. Initially thought to be eccrine, later apocrine and now; a pleuripotent stem cell origin has been suggested [1]. SCACP arises from its benign counterpart Syringo Cystadenoma Papilleferum (SCAP). Only twenty-nine cases have been reported in literature, since it was initially reported in 1980 [2,3]. We report the thirtieth case of Syringocystadenocarcinoma in a 48 year old man, developing from Syringocystadenoma. No case has been reported from Pakistan yet.

Case report

A 48 year old man presented with a swelling on occipital region of scalp for 30 years. Initially the swelling was a black macule, slowly enlarged into a nodule and later into a nodulo-ulcerative lesion. He had a trucut biopsy of the lesion four years back on which a diagnosis of SCAP had been rendered. Patient now presented with complaint of discharge and bleeding from the swelling for a year. On clinical examination; an exophytic, nodular, ulcerated and hard swelling with irregular margins, was present on the occipital region of scalp. Regional lymph nodes were not enlarged. Patient was positive for Hepatitis B virus Antigen; however, no other significant physical findings were present. The lesion was excised completely with clear margins. Skin grafting was performed.

On gross examination; the size of the tumor was 4.5 X 3.8 X 2.2cm. The cut surface was pale white, lobulated, and tumor appeared to be invading into the dermis.

Histopathological examination revealed a tumor with irregular papillomatosis in the upper part, papillae lined by single to multiple layers of cuboidal to columnar cells. These papillae had funnel-shaped invaginations into the dermis that were connected to the in-situ component (Figure 1). The in-situ component resembled in-situ breast carcinoma with ducts showing cribriform, solid, comedo and papillary architectural patterns and central necrosis (Figure 2). In-situ component underwent transformation into Adenocarcinoma, deeper into the dermis, inciting a desmoplastic stromal response. Epithelial cells showed moderate nuclear atypia, prominent nucleoli, and frequent mitotic figures. Immunohistochemical stain Cytokeratin 7(DAKO envision) was positive in the tumor (Figure 3). Immunohistochemical stain p-63 highlighted the myoepithelial cells in the in-situ component and was absent in the invasive tumor, some positivity was also noted in the epithelial cells of in-situ tumor, indicating focal squamous differentiation (Figure 4).

Discussion and conclusion

Syringocystadenocarcinoma (SCACP) is a malignant skin adnexal neoplasm. The commonest site reported is scalp; others included neck, chest, supra-pubic, peri-anal, axilla and back. The size ranged from 2.5-13 cm [4]. It has been associated with nevus sebaceous of Jaddeson in 30-40 % of cases [1]. Traditionally patients have a long history of Syringocystadenomas and malignant conversion is associated with a rapid increase in size, bleeding and pain [5]. Three cases have reported regional lymph node metastasis and three cases have reported recurrence [2,6]. Some initially reported cases have in-situ component only [7,8].

Most of the cases have reported an in-situ as well as an invasive component. Malignant tumor may be Adenocarcinoma, Squamous cell carcinoma or poorly differentiated carcinoma [9]. Malignant lesions differ from benign counterparts by being asymmetric, deeply invasive, disruption of two layered organization, nuclear atypia and mitotic activity [2]. Morphology is characteristic; however, may not be completely represented in incisional biopsies, and invasive component may be missed due to its deeper location [2]. The differential diagnoses that could be considered are: Squamous cell carcinoma, Basal cell carcinoma, Malignant Melanoma and metastatic tumors from breast, colon, lung, thyroid and ovary [5]. Immunochemical studies performed in some studies showed positivity for p63, CK5/6, CEA, CK7 and GCDFP-15 [5,10].

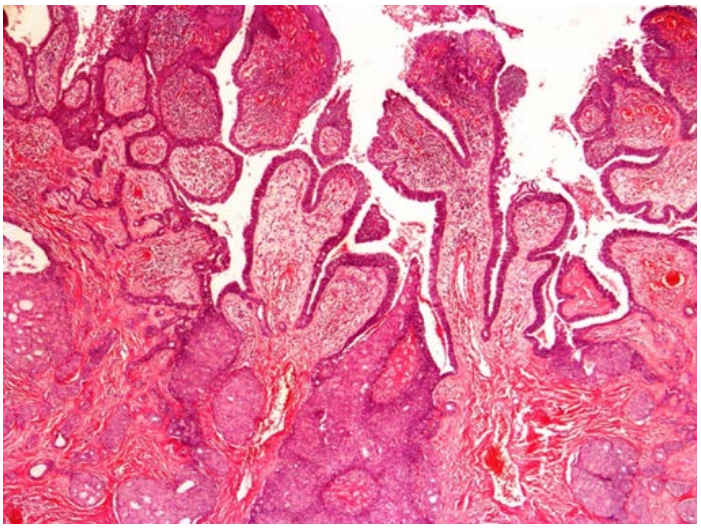

Figure 1. *Syrigocystadenocarcinoma: Surface of the tumor with papillomatosis and in-situ component.*

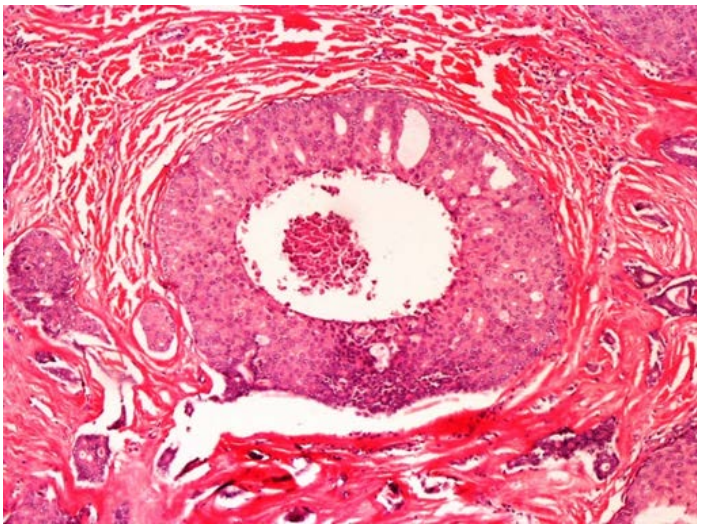

Figure 2. *In-situ resembling comedo in-situ carcinoma of breast, surrounded by the invasive component.*

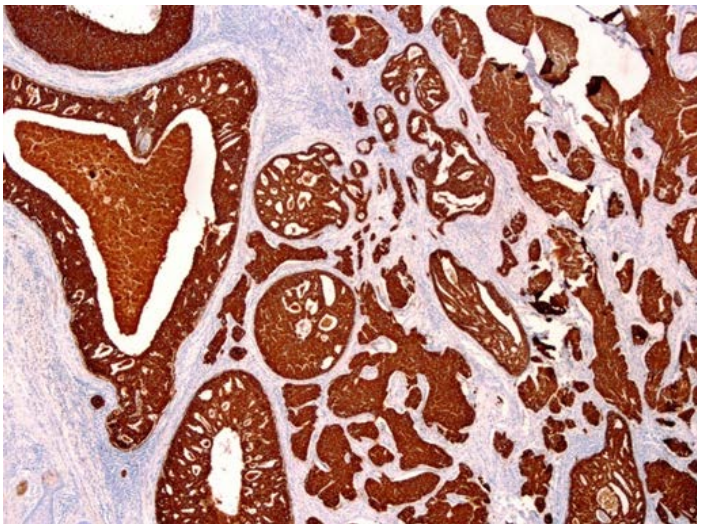

Figure 3. *Cytokeratin 7 positive in the in-situ and invasive carcinoma.*

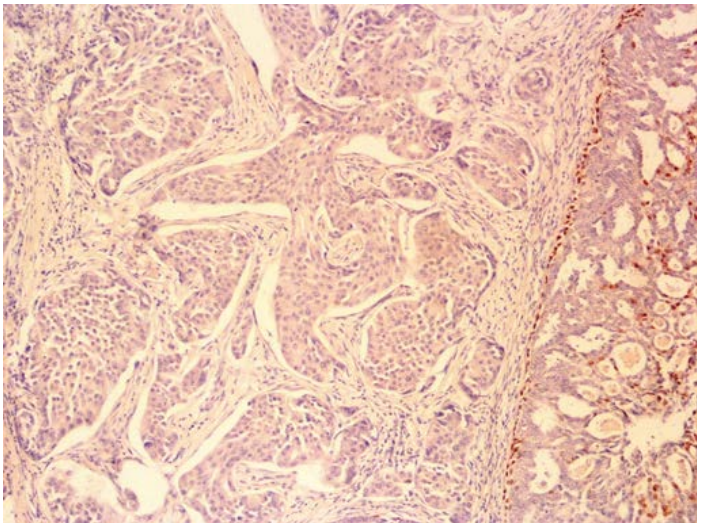

Figure 4. *P-63 positive in myoepithelial cells and some epithelial cells of the in-situ carcinoma and negative in the invasive tumor.*

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