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Case Report

Syringocystadenocarcinoma Papilliferum: A rare Pathological entity

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ABSTRACT

Syringocystadenocarcinoma papilliferum is a malignant form of its benign tumour that is syringocystadenoma papilliferum. Regarding clinical morphology it mimics varied skin tumors so diagnosis is difficult. Histopathology is ultimate role in final clinical diagnosis of these tumours. We present an uncommon case of SCACP arising from back mass in 45 years male in view of only handful cases have been reported in the literature with little information is available on its clinical and morphological aspect.

1. Introduction

Syringocystadenocarcinoma papilliferum (SCACP) is uncommon cutaneous adenocarcinoma and neoplastic/malignant part i.e. syringocystadenoma papilliferum (SCAP) described 100 yrs ago.[1] It is a skin tumor arising from the apocrine/appendageal glands.[2] The causative agents are not known but some, causes include genetic predisposition, radiation, immunosuppression and trauma to the affected area.[3] Most common affection of these tumors in HNF areas but may also appear de novo on other parts of the body.[1] The clinical presentation of this tumors composed of large cauliflower like mass with ulceration and exudation.[4,5] It mostly has old age predilection.[5] A chronic duration and ignorance about these uncommon tumours pose diagnostic and therapeutic difficulties.[4] Hence ultimate final diagnosis is depends on histopathology.

2. Case Report

A 45-year-old male presented with a globular, large mass on his back since 10 years. The patient reported that the lesion had been rapidly growing over the past year. Physical exam demonstrated a grey brown to black colored, exophytic, verrucous 9x8 cms mass on the back with serosanguinous exudate. All the clinical, biochemical and pathological investigations were within normal limit. Clinically suspected as Verrucous carcinoma in view of its morphology and excisional biopsy was sent to histopathology.

2.1. Gross finding

We received excised mass (M) 10x9x3cms. External surface showed grey white to grey brown cauliflower like mass (M) 8x7x2.5 cm with multinodular and papillary appearance. Cut section was friable, grey white to grey brown and mass infiltrating into the deeper structure.(Figure 1)

2.2. Light microscopy

Microscopic examination revealed tissue lined by stratified squamous epithelium along with tumor. Epidermis showed varying degree of papillomatosis and downward cystic invaginations lined by squamous keratinizing cells. Papillary projections extend into lumina of invaginations and are lined by glandular epithelium having two rows of

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cells, the luminal or internal layer consisting of columnar cells showing secretion and external layer of small cuboidal cells.(Figure 2) Papillae are dense and diffusely infiltrated by mononuclear cells. Subepithelium showed tumor comprised of neoplastic cells arranged in glands, acini, tubules and cribriform pattern. (Figure 3) These neoplastic cells were round to oval and possessed hyperchromatic nuclei, scant to moderated eosinophilic cytoplasm and high N:C ratio.(Figure 4) The tumor component extended upto dermis and underneath fibrocollagenous and fibromuscular tissue was free from tumor.

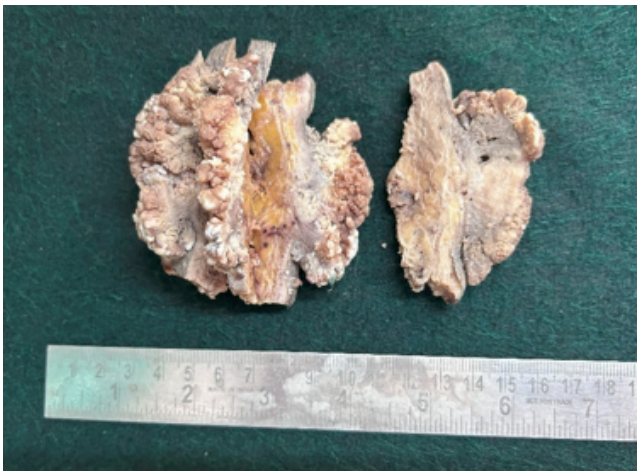

Figure 1: Gross of excised cauliflower mass with papillary appearance and foci of invasion on cut section.

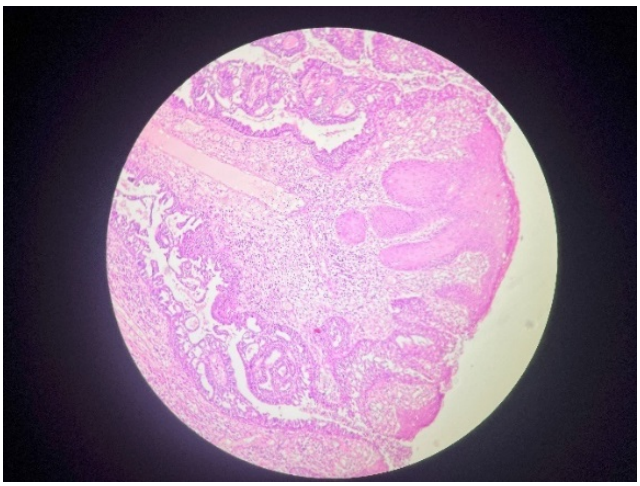

Figure 2: Light microscopic of SCAP-Papillary projections extend into lumina of invaginations and are lined by glandular epithelium having two rows of cells. (H &E stain, x100)

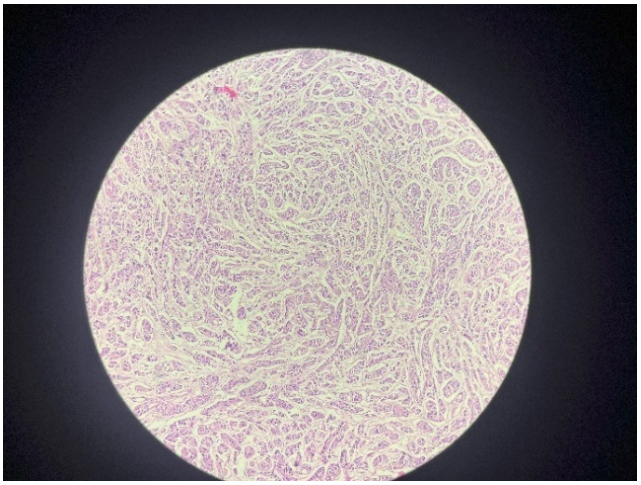

Figure 3: Subepithelium showed tumor comprised of neoplastic cells arranged in glands, acini, tubules and cribriform pattern. (H &E stain, x100)

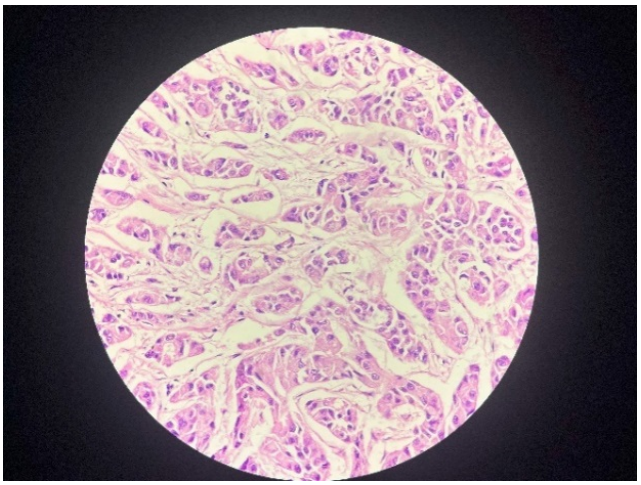

Figure 4: Individualtumour cells are round to oval with hyperchromatic nuclei , scant to moderated eosinophilic cytoplasm and high N:C ratio (H &E stain, x400) .

3. Discussion

In early 1917, first case of SCACP was designated.[6] SCACP is a rare skin adenocarcinoma, adenexal origin from apocrine glands with prolonged course of disease.[7,8] The aetiology of the tumor is obscure, but genetic predisposition seems to play a main role in the carcinogenesis of the SCACP.[7] Grossly the tumor appeared as grey tan cauliflower mass with surface ulcerations and exudations leading to verrucous appearance of the mass.[9]

Histologically, SCACP can present as in situ adenocarcinoma, invasive tumor and rare instances invasive sqamous cell carcinoma.[10] Histologically, SCACP is similar to its benign component as cystic and papillary pattern of epithelial elements composed of

an inner columnar and outer cuboidal layer, projecting downwards into the dermis. Innermost layer showed columnar epithelium with secretions. The stroma of the tumor are infiltrated with momonuclear inflammatory cells.[11] SCACP is the rare malignant counterpart of SCAP. Clinically rapid enlargement and ulceration and bleeding are indicative of malignant transformation. The diagnosis is clinically suspected and histologically confirmed by the presence of asymmetry, poor circumscription, marked cytological atypia, mitotic figures and invasion into dermis.[11] Differential diagnosis as SCC,BCC ,melanoma, lymphoma, porocarcinoma can easily differentiate on light microscopy from SCACP.

In immunohistochemistry, no specific marker is useful that differentiate primary from metastatic adenocarcinoma. But, positive results of cytokeratin 5/6, p63, and D2-40 staining is usually associated with cutaneous neoplasms rather than metastatic tumors like adenocarcinoma.[11]

In view of its varied and prolong presentation, treatment modalities are not well established. However, complete surgical excision of the lesion is the main modality of the treatment and monitoring and follow up is required for the patients with SCACP.[12] Recurrence of these lesions is not uncommon and has been reported in less than 20% of cases.

4. Conclusion

SCACP is extremely rare cutaneous adnexal tumours as only few (less than 50) of case reports are available in the medical literature. Behaviour, clinical features and morphology of this tumor can be easily confused with the other skin cancers and cutaneous metastases. Hence, histopathological confirmation is crucial for the accurate diagnosis of SCACP. The most important thing is to differentiate these tumours from other cutaneous neoplasms due to its unusual clinical presentation and varied morphology and different treatment modalities for the benefit of the patients.

5. Source of Funding

None.

6. Conflict of Interest

None.

References

1. Hoekzema R, Leenarts MF, Nijhuis EW. Syringocystadenocarcinoma papilliferum in a linear nevus verrucosus. *J Cutan Pathol*. 2011;38(2):246–50.
2. Park SH, Shin YM, Shin DH, Choi JS, Kim KH. Syringocystadenocarcinoma papilliferum: a case report. *J Korean Med Sci*. 2007;22(4):762–5.
3. Bell D, Aung PP, Prieto VG, Ivan D. Next-generation sequencing reveals rare genomic alterations in aggressive digital papillary adenocarcinoma. *Ann Diagn Pathol*. 2015;19(6):381–4.
4. Zhang Y, Kong YY, Cai X, Shen XX, Kong JC. Syringocystadenocarcinoma papilliferum: clinicopathologic analysis of 10 cases. *J Cutan Pathol*. 2017;44(6):538–43.
5. Peterson J, Tefft K, Blackmon J, Rajpara A, Fraga G. Syringocystadenocarcinoma papilliferum: a rare tumor with a favorable prognosis. *Dermatol Online J*. 2013;19(9):230–4.
6. Dadras MS, Baghani M, Rakhshan A, Djafari AA, Abdollahimajd F. Genital Syringocystadenocarcinoma papilliferum: An unusual location and review of the literature. *Urol Case Rep*. 2022;40:101934. doi:10.1016/j.eucr.2021.101934.
7. Halsey JN, Faith EF, Logan SJ, Shenoy A, Schieffer KM, Cottrell CE, et al. Syringocystadenocarcinoma Papilliferum in a Fifteen-Year-Old Girl: A Case Report and Review of the Literature. *Case Rep Dermatol Med*. 2022;p. 8076649. doi:10.1155/2022/8076649.
8. Lee KG, Choi W, Lim JS, Hahn HJ, Myung KB, Cheong SH, et al. Syringocystadenocarcinoma papilliferum: a case report and review of the literature. *Ann Dermatol*. 2019;31(5):559–62.
9. Muthusamy RK, Mehta SS. Syringocystadenocarcinoma papilliferum with coexisting trichoblastoma: A case report with review of literature. *Indian J Dermatol Venereol Leprol*. 2017;83(5):574–6.
10. Parekh V, Guerrero CE, Knapp CF, Elmets CA, Mckay KM. A Histological Snapshot of Hypothetical Multistep Progression From Nevus Sebaceus to Invasive Syringocystadenocarcinoma Papilliferum. *Am J Dermatopathol*. 2016;38(1):56–62.
11. Kim MS, Lee JH, Lee WM, Son SJ. A case of tubular apocrine adenoma with syringocystadenomapapilliferum that developed in a nevus sebaceus. *Ann Dermatol*. 2010;22(3):319–22.
12. Hoguet AS, Dolphin K, Mccormick SA, Milman T. Syringocystadenocarcinoma papilliferum of the eyelid. *Ophthalmic Plast Reconstr Surg*. 2012;28(1):27–9.

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