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SYRINGOCYSTADENOMA PAPILLIFERUM: A CASE REPORT

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ABSTRACT

Syringocystadenoma papilliferum also known as naevus syringocystadenomatosus papilliferus is an exuberant proliferating lesion, commonly seen on the scalp in association with an organoid naevus, and showing predominantly apocrine differentiation. It is a benign adnexal skin tumour of the apocrine or the eccrine type. It is relatively a rare neoplasm.

INTRODUCTION

Syringocystadenoma papilliferum also known as naevus syringocystadenomatosus papilliferus is a proliferating lesion, commonly seen on the scalp in association with an organoid naevus, and showing predominantly apocrine differentiation. It is a benign adnexal skin tumour of the apocrine or the eccrine type. It is relatively a rare neoplasm [1]. Syringocystadenoma papilliferum occurs with equal frequency in both sexes [2]. The tumor has varied clinical presentation. It presents as a hairless area on the scalp [3] It is first noted at birth and presents as a solitary papule, several papules in a linear arrangement or a plaque.

CASE REPORT–

60 year old man came to the OPD with the complaints of ulcerative lesion on the scalp since 6-7 months which was gradually progessive and increased in size. The patient also had complaints of bleeding on touch and on combing hair. The history of tenderness and itching was also present. On presentation at the OPD in MGM Hospital the patient had maggots present at the scalp swelling. The patient underwent excision of the lesion under local anesthesia. The lesion was excised completely with a normal 1 cm margin around the lesion which was sent for histopathological examination.

Gross Examination :

Multiple grey white to grey brown firm tissue pieces were received . Largest measuring 1 x 0.5 cm and smallest measuring 0.8 x 0.5 cm.

Microscopic Examination:

H and E stained section showed tissue bit lined by keratinised stratified squamous epithelium showing papillomatous hyperplasia (FIG1). Multiple cystic invaginations were extending down from the epidermis into dermis comprising of papillary projections extending into the luminal aspect in the lower part (FIG2).

The papillae were lined by bilayered epithelium consisting of columnar cells and cuboidal cell with oval nuclei and pale eosinophilic cytoplasm. Some cells showed decapitation secretions (FIG3). The stroma was infiltrated with lymphomononuclear cells predominantly comprising of sheets of plasma cells. Deep dermis showed tubular glands having apocrine differentiation . Sebaceous glands were normal. Based on these features, the diagnosis of syringocystadenoma papilliferum was made.

The patient was followed up for seven months and showed no evidence of recurrence.

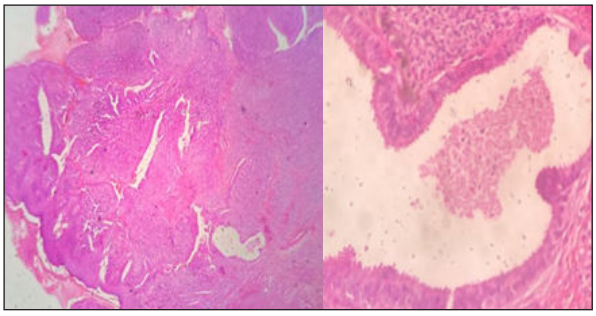

Fig1. Keratinised Stratified Squamous Epithelium Showing Papillomatous Hyperplasia

Fig2. Cystic invagination extending down from the epidermis into dermis comprising of papillary projections

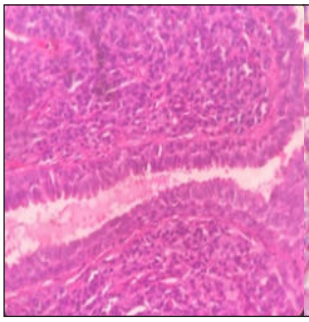

Fig3. Papillae lined by bilayered epithelium with cells showing decapitation secretions. Stroma infiltrated with lymphomononuclear cells predominantly comprising of sheets of plasma cells

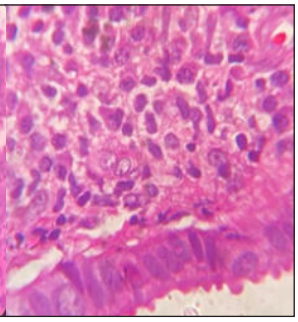

Fig4. Inset showing decapitatory secretions along with multiple plasma cells.

DISCUSSION:

Syringocystadenoma papilliferum most commonly occurs in the head and neck region sometimes present within a nevus sebaceus.

There are three clinical types which have been described:

a. **Plaque Type:**

It presents as alopecic patch on the scalp that enlarges during

puberty to become verrucous, nodular, or crusted plaques.

b. **Linear Type:**
It consists of multiple reddish, fi rm papules, or umbilicated nodules 1-10 mm in size.

c. **Solitary Nodular Type:**
These are domed pedunculated nodules 5-10 mm in size and located near trunk shoulder and axilla[1].

It reportedly evolves within three stages:
a. **Infantile Stage:**
It appears as alopecic orange yellow plaque.

b. **Adolescent Stage:** Under androgenic influence the plaques undergoes various changes such as hyperkeratosis, hyperpigmentation, and sebaceous gland formation.

c. **Adult Stage:**
It is characterized by the presence of large sebaceous glands, ectopic apocrine glands, and epidermal hyperplasia. [4]

In about one-third of the case, syringocystadenoma papilliferum is associated with a nevus sebaceous. Multiple tumours of adnexal origin (such as trichoblastomas, aporine adenomas, hidradenoma papilliferum, porona follicular, trichilemmoma etc.) have been reported to arise on a sebaceous nevus, among which Syringocystadenoma papilliferum may be included [5]. Basal Cell Carcinoma (BCC) development has been reported in upto 10% of the cases[6]. Clinical diagnosis of SCAP is mostly not feasible due to the various presentations and possible differential diagnosis thus histopathology is the best option. Knowledge of this diagnosis would add a supplementary alternative to differential diagnosis. Treatment for such cases is surgical excision. SCAP should be excised given its future malignant transformation in adults. Thus, excision sufficiently eliminates this risk as well allows cosmetic relief for the patient[7]. The malignant tumors reported in association with syringocystadenoma papilliferum are squamous cell carcinoma, basal cell carcinoma, and ductal carcinoma. Basal cell carcinoma is the most common pathology and has been reported in 10% of the cases while only two cases of squamous carcinoma have been reported.

Histopathology typically shows varying degrees of papillomatosis along with cystic invaginations and malformed sebaceous glands. Immunohistochemistry helps in differentiating the origin of the tumor, i.e. either eccrine or apocrine, but is of no clinical significance. Positive immunoreactivity for proteins 15 and 24 and zinc-2 glycoprotein demonstrates evidence of apocrine differentiation, while positivity for Cytokeratins demonstrates eccrine differentiation[1] Immunohistochemistry was not done in the present case.

Smooth muscle actin (SMA) positivity is normally the feature of myoepithelial cells but syringocystadenoma papilliferum do not contain myoepithelial cells and hence the positivity indicates immaturity of the tumor.

Yamamoto et al. postulated an origin in pluripotent cells on immunohistochemical and ultrastructural grounds.[8] Böni et al. 10 showed mutations in PTCH or P16 tumour suppressor genes in syringocystadenoma papilliferum[9] . Kazakov et al. noted an overlap with tubular adenoma[10].

SCAP can exhibit both apocrine and eccrine differentiation . Studies have demonstrated loss of heterzygosity for patched and p16 negative regulator of cell cycle, in SCAP , suggesting that these molecules play a role in the pathogenesis of these lesions.

CONCLUSION

Syringocystadenoma Papilliferum is a rare neoplasm, and even though it is called as an adolescent tumour as it usually occurs at birth, during infancy or around the time of puberty, it quite rarely appears in adults. The nodular variety is seen most commonly in the trunk, but here, it presented on the scalp. In the present case, it was clinically diagnosed as benign adenexal tumour of the scalp, which later was histologically confirmed as Syringocystadenoma Papilliferum. There are many overlaps regarding differential diagnosis and its association with other benign pathologies as well as developing into various malignancies, it is mandatory to confirm with histopathological examination for early diagnosis and cure by surgical excision.

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