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Syringocystadenoma Papilliferum-Case Report.

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Abstract: *Syringocystadenoma papilliferum is an exceedingly rare skin adnexal neoplasm of apocrine gland origin located primarily on the scalp and appearing as a hairless nodular plaque lesion. In one third of the cases Syringocystadenoma papilliferum is associated with Naevus Sebaceous of Jadassohn. We report the case of a 15 year old boy with history of an alopecic patch over scalp since childhood with the secondary development of multiple soft nodules over the last 2 years. Various malignancies like basal cell carcinoma, squamous cell carcinoma, ductal carcinoma, apocrine carcinoma have been documented to arise in Syringocystadenoma papilliferum. A strong clinical acumen, prompt excision and confirmation by histopathology underlie the treatment of this deceptively docile neoplasm. We report this case because of its rarity, tendency for vagrant behavior and their implied management ramifications.*

Keywords- *Syringocystadenoma papilliferum, Naevus Sebaceous of Jadassohn.*

I. Introduction

Skin is the largest organ of the body, a complex organ with varied protective, metabolic, homeostatic and excretory function; the pathologies afflicting the skin span a bewildering spectrum and a specialty subject in its own.

The skin is commonly affected by a multitude of congenital, developmental and neoplastic aberrations and lesions, some of which are so rare and find mention in aeons in medical literature.

II. Case Report

15 year old boy came to pathology OPD with history of alopecic patch over scalp since childhood, now presenting with the chief complains of development of multiple soft nodules over last two years and associated itching and bleeding.

Examination revealed a yellowish white alopecic plaque of 3x1 cm over the right parietal area of scalp. Also noted were 4-5 soft nodular crusted lesions of varying dimensions. There were no active bleeding, discharge or satellite nodules; skin sensations were normal. A presumptive clinical diagnosis of squamous cell carcinoma or angiokeratoma was reached. The patient was worked up and posted for excision reconstruction of the lesion.

Figure1: Gross excised specimen showing verrucous nodular growth.

Histopathological examination of the lesion confirmed the diagnosis of Syringocystadenoma papilliferum arising in Naevus Sebaceous of Jadassohn. The post operative period was uneventful and patient is on regular follow up.

III. Histopathological Examination

Sections reveal tissue lined by hyperkeratotic, parakeratotic squamous epithelium with associated papillomatosis. Underlying dermis shows large lobules of sebaceous glands, hair follicles and many undifferentiated solid structures. The deeper dermis revealed presence of many cystically dilated ectopic apocrine glands lined by double layered epithelium and containing amphophilic secretions. The overlying epidermis revealed cystic invaginations with multiple tumor islands lined by tall columnar luminal epithelial cells showing decapitation type of secretions and outer low cuboidal cells. The core of the papillae revealed dense mononuclear infiltrate of lymphocytes and plasma cells. No cytological atypia was noted.

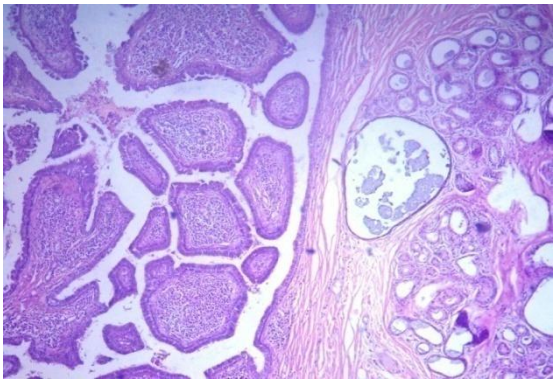

Figure2: Tumor islands and underneath cystically dilated apocrine glands. (H&E, 04X)

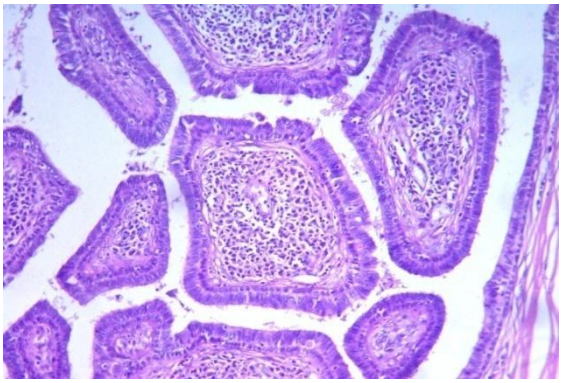

Figure3: Tumor islands lined by double epithelium with fibrocollagenous cores. (H&E stain, 10X)

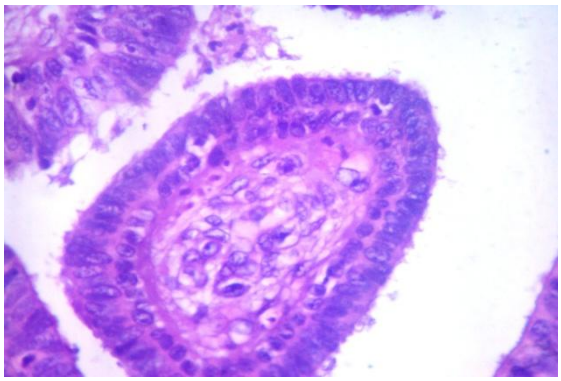

Figure4: Tumor islands lined by tall columnar luminal epithelial cells and outer low cuboidal cells. The core of the papillae revealed dense mononuclear infiltrate of lymphocytes and plasma cells (H&E stain, 40X)

The final histopathological diagnosis was Syringocystadenoma papilliferum arising in a congenital Nevus Sebaceous of Jadassohn.

IV. Discussion

Syringocystadenoma papilliferum is an exceedingly rare hamartomatous proliferative malformation derived from apocrine sweat glands of skin. About 50% are present at birth or appear during infancy and tend to proliferate around puberty. [1]

3 clinical types have been described [2] [3]

A) **Plaque type**: presenting as a alopecic patch on scalp and my enlarge during puberty to become nodular, verrucous or crusted.

Plaques commonly tend to be associated with a Naevus Sebaceous of Jadassohn in one third of the cases. [4]

B) **Linear type**: consists of multiple reddish pink firm papules or umbilicated nodules 1-10 mm in size commonly occurring over face and neck.

C) **Solitary nodular type**: they are domed pedunculated nodules 5-10mm in size with a predilection for the trunk shoulder and axillae.

Syringocystadenoma Papilliferum is commonly associated with hamartomas of follicular or sebaceous gland origin,and in one third of cases Syringocystadenoma papilliferum is associated with Naevus Sebaceous of Jadassohn. [3] [4]

Naevus Sebaceous of Jadassohn is a congenital organoid naevus appearing as a yellowish patch at birth and tend to become raised, papillomatous or verrucous at puberty under the influence of androgens. [5] [6]

It evolves over the years going through [5]:

A) **Infantile stage**: appears as a alopecic orange yellow plaque.
B) **Adolescent stage**: under the influence of androgens the plaque thickens with verrucous hyperkeratosis, hyper pigmentation and sebaceous gland proliferation.
C) **Adult stage**: characterized by presence of large malformed sebaceous glands, ectopic apocrine glands and prominent epidermal hyperplasia. It is during this period that many benign and malignant neoplasms develop.

Malignant change is heralded by rapid increase in size, appearance of new lesions, bleeding and appearance of metastatic lymph nodes.

Shapiro et al. [7] in his extensive review has outlined the various benign and malignant neoplasm arising in the Naevus Sebaceous of Jadassohn. The common benign tumors include Syringocystadenoma papilliferum, trichoblastoma, trichelemmoma, sebaceous adenoma, apocrine adenoma, spiradenoma, hidradenoma, syringoma, osteoma. And the malignant tumors include Squamous cell carcinoma, basal cell carcinoma, sebaceous cell carcinoma, apocrine carcinoma, verrucous carcinoma, mucoepidermoid carcinoma.

Of the gamut of lesions known to arise in this congenital naevus, Syringocystadenoma papilliferum and trichoblastoma are the commonest benign neoplasm and otherwise and Basal cell carcinoma is the commonest malignancy.[7] [8]

Kaddu et al.[9] in his study of 316 cases neoplasm arising in Naevus sebaceous of Jadassohn found 7.6% of benign and 2 cases of malignant cases all occurring in adulthood.

Munoz-Perez et al. [10] in their series of 226 cases found 18% of benign tumors in subjects all above the age of 14 years.

There are very few case reports or references of Syringocystadenoma papilliferum in Indian literature; courtesy the extreme rarity, under reporting and ignorance.

Kumar V. et al. [11] in 1991 reported a single case of Syringocystadenoma papilliferum in a 10 year old girl.

Golvalkar R.M. et al. [12] in 1994 reported a single case of Syringocystadenoma papilliferum arising in backdrop of Naevus sebaceous of Jadassohn in a 13 year old girl.

Sood A. et al. [13] in 2000 reported 2 cases of Syringocystadenoma papilliferum in a 5 year old boy and 20 year old man with lesions in the axilla and neck respectively.

Because of the apparently benign appearance of this neoplasm the clinician is put into an ambivalence either to stay and play with the tumor or to scoop and hoot.

Tumor–Or-No Tumor; a watchful alertness is the key as by the adulthood malignant tumors can occur in 40% of Naevus Sebaceous of Jadassohn. [6]

Radiotherapy and other destructive procedures are ineffective and best avoided. [2]

Surgical excision with reconstruction is the treatment of choice. [2]

V. Conclusion

Syringocystadenoma papilliferum is an extremely rare neoplasm commonly arising in association with congenital Naevus sebaceous of Jadassohn, because of its propensity to undergo malignant change, it is better to tread a cautious line and take preemptive steps early in the better interest if the patient, lest the prognosis be marred by catastrophic development of malignancy.

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