

Syringocystadenoma Papilliferum - A Case Report

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Abstract: *Syringocystadenoma papilliferum* 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 Naves Sebaceous of Jadassohn. A strong clinical acumen, prompt excision and confirmation by histopathology underline treatment of this deceptively docile neoplasm. We report a case of 9 year old boy because of its rarity and tendency for vagrant behavior.

Keywords: Syringocystadenoma papilliferum, Naves sebaceous of Jadassohn, Apocrine gland origin, Plaque, Vagrant behavior

1. 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 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 to find mention even in medical literature.

2. Case Report

A 19 year old boy presented with a lesion over the scalp since birth. Lesion started to increase in size from last 4 years.

On examination: Single localised hyperpigmented plaque with overlying verrucous papule seen.



Figure 1: Received single soft to firm, whitish, skin covered, hair bearing, tissue mass measuring 5x3x0.6 cms

3. Histopathology

H&E stained section studied show epidermis and dermis. Epidermis is lined by Stratified squamous epithelium which shows features of varying degree of papillomatous hyperplasia. Underneath dermis shows increased number of sebaceous glands. Papillary projections in the dermis are lined by two rows of cells. Luminal row of cells consists of columnar cells and outer layer of cells consists of cuboidal cells. Core contains Lympho- plasmacytic inflammatory infiltrate and congested blood vessels. As shown in figures (2), (3), (4).

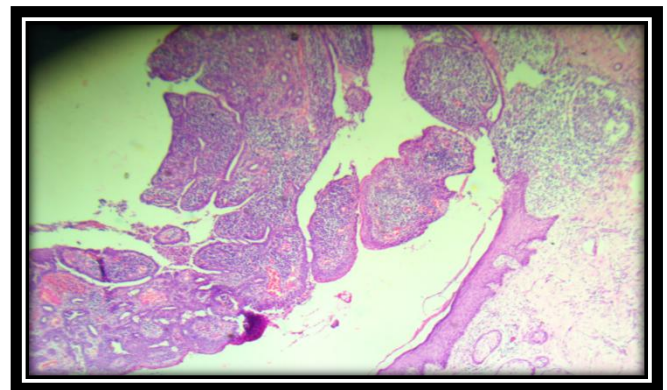


Figure 2: 10X

Epidermis with Papillary dermal projection lined by inner layer of columnar cells and outer cuboidal cells

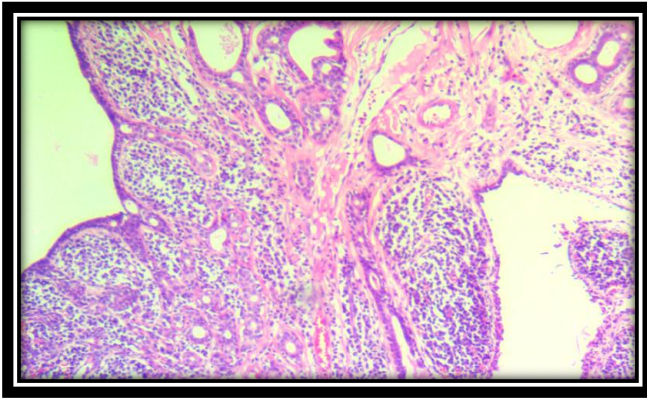


Figure 3: 10X

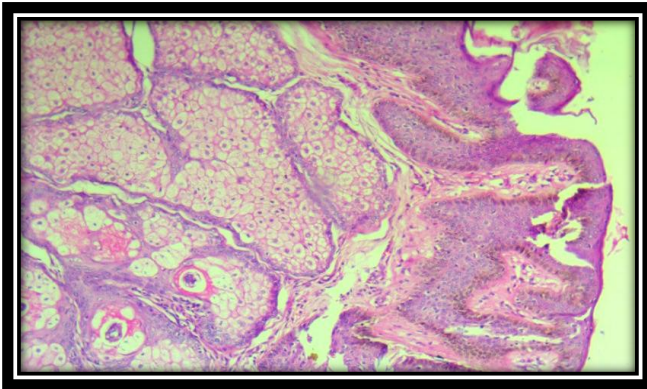


Figure 4: 40X - Varying degree of papillary hyperplasia and increase number of sebaceous glands

4. 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⁾

- 1) **Plaque type:** presenting as a alopecic patch on scalp and may enlarge during puberty to become nodular, verrucous or crusted plaques commonly tend to be associated with nevus Sebaceous of Jadassohn in one third of the cases⁽⁴⁾
- 2) **Linear type:** Consists of multiple reddish pink firm papules or umbilicated nodules 1-10 mm in size commonly occurring over face and neck.
- 3) **Solitary nodular type:** they are domed pedunculated nodules 5-10mm in size with predilection for trunk shoulder and axillae.

Syringocystadenoma papilliferum is commonly associated with hamartomas of follicular or sebaceous gland origin in one third of cases syringocystadenoma papilliferum is associated with Naves sebaceous of Jadassohn.⁽³⁾⁽⁴⁾

Nevus Sebaceous of Jadssohn is a congenital organoid nevus 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⁾

- 1) Infantile stage: appears as a alopecic orange yellow plaque
- 2) Adolescent stage: under the influence of androgens the plaque thickens with verrucous hyperkeratosis, hyper pigmentation and sebaceous gland proliferation.
- 3) Adult stage: characterized by presence large malformed sebaceous gland, 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 lesion, 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 Naveous sebaceous of Jadassohn. The common benign tumors include Syringocystadenoma papilliferum, trichoblastoma, trichelemmama, sebaceous 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 lesions known to arise in congenital nevus, syringocystadenoma papilliferum and trichoblastoma are the commonest benign neoplasm and Basal cell carcinoma is the commonest type of malignancy⁽⁷⁾⁽⁸⁾

Kaddu et el⁽⁹⁾ in his study of 316 cases neoplasm arising in nevus sebaceous of Jadassohn found 7.6% of benign and two cases of malignant cases all occurring in adulthood.

Govardhan R. M⁽¹⁰⁾ in 1994 reported a single case of syringocystadenoma papilliferum arising in backdrop of nevus sebaceous of Jadassohn in 13 year old girl. Surgical Excision with reconstruction is the treatment of choice.

5. Conclusion

Syringocystadenoma Papilliferum is an extremely rare neoplasm commonly arising in association with congenital nevus of Jadassohn because of its propensity to undergo malignant change, it is better to be cautious and presumptive steps adopted early in better interest of the patient.

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