ORIGINAL RESEARCH PAPER

General Surgery

SYRINGOCYSTADENOMA PAPILLIFERUM – A RARE CASE REPORT

KEY WORDS: Adnexal tumor, Apocrine glands, Naevus Sebaceous, Basal Cell Carcinoma

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ABSTRACT

Syringocystadenoma papilliferum is a rare benign adnexal tumor of apocrine or eccrine glands. It is a childhood tumor with a relatively higher incidence at birth. The common sites of occurrence are the face and scalp. The propensity of the nodular lesion is towards the trunk, but here in our case, we present a case of nodular syringocystadenoma papilliferum of the scalp. The tumor can arise de novo or from pre-existing naevus sebaceous. The tumor rarely has a malignant course, most often basal cell carcinoma. Herein, we present an adolescent with the nodular type of syringocystadenoma papilliferum of the scalp, treated with surgical excision.

INTRODUCTION :

Syringocystadenoma papilliferum is a very rare benign adnexal tumor of the apocrine and eccrine sweat glands. Predominantly it occurs in childhood, with a vast majority of incidence at birth. It may develop during puberty in about 15% to 30% (Karg et al., 2008). The lesions may arise de novo or from pre-existing naevus sebaceous in a few cases (Helwig, 1955). Rarely, it may transform into a malignant lesion. It is usually asymptomatic slow-growing reddish papule or plaque. It is rarely pigmented. The most common site of incidence is the scalp and face. The other rare areas that are reported occurrence are nipple, breast, external genitalia, and over the extremities (Kasashima et al., 2016). The mutations associated with sporadic forms were BRAF V600E and less often KRAS, while the lesions arising from naevus sebaceous had underlying HRAS mutations (Fontecilla et al., 2018).

Case history :

An eleven-year-old female with no previous medical history presented a painless, intermittently pruritic papule over the scalp. The lesion has grown over the past year. On examination, a reddish pigmented patch of size 4 x 3 cm was noted over the scalp over the right occipital region with clearly defined margins with surface nodularity. The lesion was not associated with any discharge or bleed. No regional lymph nodes were palpable. A skull radiograph ruled out any underlying bony involvement. A biopsy of the lesion showed epidermis with papillomatous extensions and duct-like projections. It was lined by columnar cells in the dermis in the presence of dilated ducts connecting to the epidermis. The stroma has dense infiltration by plasma cells. A histological diagnosis of syringocystadenoma papilliferum was made. Complete surgical excision of the tumor with primary closure of the scalp was done. At an interval of six months, follow-up for two years showed no recurrence.

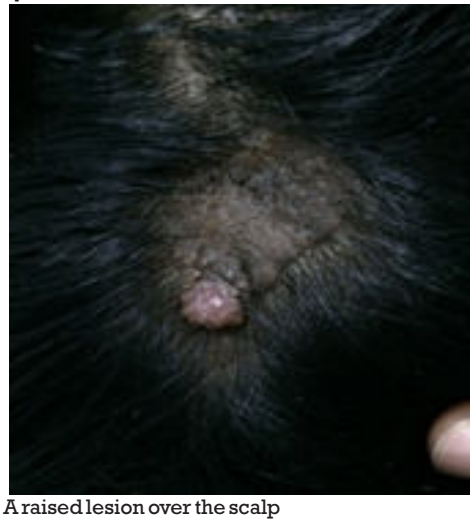

Fig 1: A raised lesion over the scalp

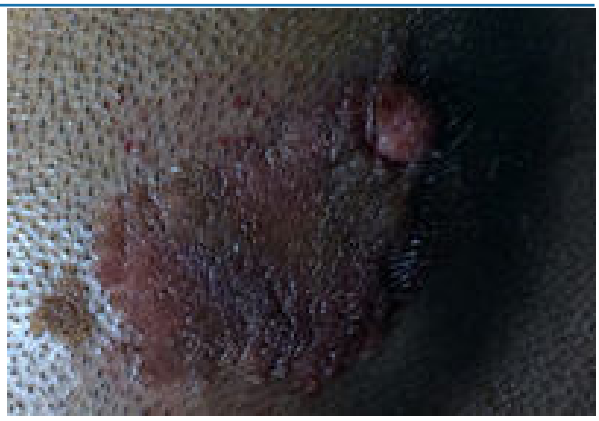

Fig 2: A reddish patch over the scalp

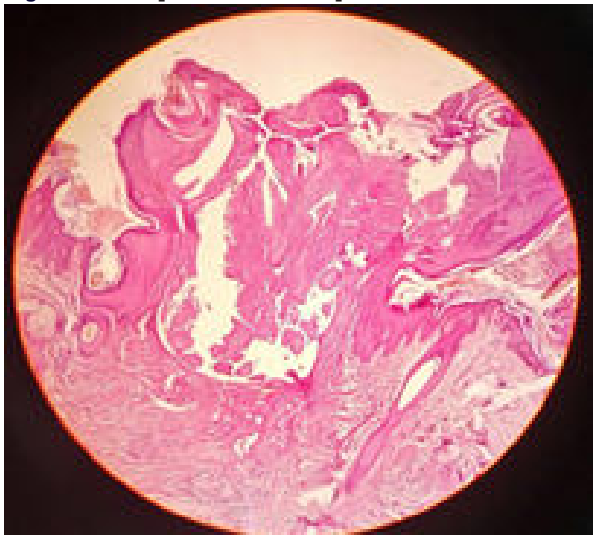

Fig 3: Microscopic picture of Syringocystadenoma papilliferum

DISCUSSION :

Syringocystadenoma papilliferum (SCAP) is a rare benign adnexal neoplasm of either apocrine or eccrine sweat gland origin. Usually, it manifests in childbirth or early childhood. The nodular variety usually has a propensity to the trunk but is presented in the scalp. Multiple tumors of adnexal origin have been reported to arise from naevus sebaceous, and syringocystadenoma papilliferum may be included in them. Rarely, it may turn into malignancy with a higher propensity towards basal cell carcinoma rather than squamous cell carcinoma (Helwig, 1955). Ulceration or rapid enlargement is indicative of malignant transformation. SCAP develops from pluripotential appendageal cells or hybrid type apoeccrine glands. The activation of MAPK and PI3k-Akt pathways may

predispose to SCAP (Parekh et al., 2016).

Naevus sebaceous led the differential diagnosis in this patient. The histological differential diagnosis includes other apocrine and eccrine adenomas such as hidradenoma papilliferum. Though the chance of malignant transformation is negligible, the treatment of choice is surgical excision and biopsy. But recurrence is common, though there was no recurrence in our case in a two-year follow-up. Carbon dioxide laser excision and Mohs micrographic surgery also have been performed (Goyal et al., 2015). This case was particularly interesting as a clinical diagnosis of naevus sebaceous turned out to be SCAP.

CONCLUSION :

Syringocystadenoma papilliferum is a rare benign adnexal neoplasm. The propensity of a nodular lesion is towards the trunk; it may occur in the other areas such as the scalp. Hence, a differential diagnosis of syringocystadenoma papilliferum should be in place of patchy or nodular scalp lesions, and surgical excision is the treatment of choice.

REFERENCES :

1. Fontecilla, N. M., Kent, R. A., & Marathe, K. S. (2018). An 8-year-old girl with a papule on her cheek. *Pediatric Dermatology*, *35*(4), 511–512. https://doi.org/10.1111/pde.13469
2. Goyal, N., Aggarwal, I., & Bhargava, S. (2015). Cytological diagnosis of syringocystadenoma papilliferum a rare adnexal tumor. *Journal of Cytology*, *32*(2), 130. https://doi.org/10.4103/0970-9371.160571
3. Helwig, E. B. (1955). Syringadenoma Papilliferum: Lesions With and Without Naevus Sebaceous and Basal Cell Carcinoma. *A.M.A. Archives of Dermatology*, *71*(3), 361. https://doi.org/10.1001/archderm.1955.01540270073011
4. Karg, E., Korom, I., Varga, E., Ban, G., & Turi, S. (2008). Congenital Syringocystadenoma Papilliferum. *Pediatric Dermatology*, *25*(1), 132–133. https://doi.org/10.1111/j.1525-1470.2007.00607.x
5. Kasashima, S., Kawashima, A., & Fujii, T. (2016). Syringocystadenoma papilliferum of the male nipple: Syringocystadenoma papilliferum of the male nipple. *Journal of Cutaneous Pathology*, *43*(8), 679–683. https://doi.org/10.1111/cup.12593
6. Parekh, V., Guerrero, C. E., Knapp, C. F., Elmets, C. A., & McKay, K. M. (2016). A Histological Snapshot of Hypothetical Multistep Progression From Nevus Sebaceus to Invasive Syringocystadenocarcinoma Papilliferum. *The American Journal of Dermatopathology*, *38*(1), 56–62. https://doi.org/10.1097/DAD.0000000000000370