Syringocystadenoma Papilliferum Arising Within A Nevus Sebaceous: A Case Report*

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ABSTRACT

Syringocystadenoma papilliferum is a rare benign adnexal skin tumor of apocrine or eccrine differentiation. It usually appears at puberty wherein a third of cases arise within a nevus sebaceous. We report a 14 year-old male with an erythematous fleshy plaque on the scalp of 3 years duration that developed from a pre-existing hairless plaque since birth. Histopathology confirmed the above diagnosis.

Keywords: Syringocystadenoma papilliferum, nevus sebaceous, adnexal tumor

INTRODUCTION

Syringocystadenoma papilliferum is a rare benign tumor derived from apocrine or eccrine glands. First described by Werther in 1913 as an unusual tumor, it was termed as naevus syringadenomatosus papilliferus. It commonly appears at puberty wherein one-third of cases arise from a nevus sebaceous. Syringocystadenoma papilliferum usually presents as a solitary brown to red papillomatous or verrucous, moist plaque on the head and neck that often becomes indurated and crusted due to stromal fibrosis and accumulation of dried blood or exudates from associated apocrine gland secretions. [1,2,3]

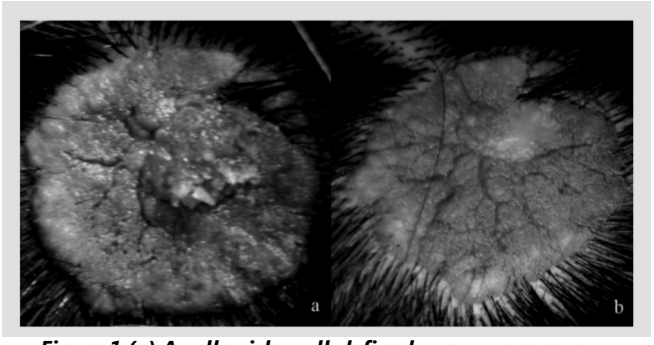

Figure 1 (a) A yellowish, well-defined, waxy, verrucous plaque with an erythematous fleshy plaque on the center, (b) Patient 7 months post excision and Electrocautery of syringocystadenoma papilliferum

CASE HISTORY

An otherwise healthy 14 year-old male presented at our clinic with a slowly growing plaque on his scalp of three years duration. The lesion was noted to develop from a pre-existing hairless plaque since birth. Except for the presence of blood tinged discharge and occasional pruritus, there were no associated signs and symptoms. Clinical examination revealed a solitary, erythematous, well defined, round fleshy plaque measuring 1x1.5cm on top of a yellowish, well-defined, round, waxy, verrucous, hairless plaque measuring 4x4 cm on the scalp. (Figure 1a)

A 3mm punch biopsy was performed on both lesions. Histopathologically, the yellowish, waxy, verrucous plaque revealed hyperkeratosis, orthokeratosis and papillomatosis. Follicular infundibula were widely dilated and plugged by keratin. Enlarged and mature sebaceous glands in the superficial dermis and mature apocrine glands in the deeper dermis were present. (Figure 2a and 2b)

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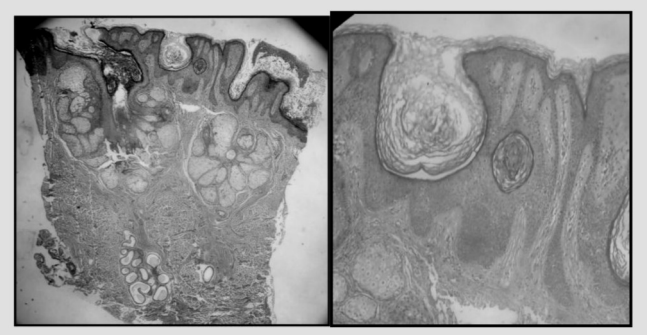

Figure 2 (a) Photomicrograph of a specimen taken from yellowish waxy, verrucous plaque on the scalp showing nevus sebaceous characterized by hyperkeratosis and papillomatosis; follicular infundibulum dilated and plugged by keratin, enlarged and mature sebaceous glands in the superficial dermis and mature apocrine glands in the deep dermis. (b) On higher magnification, Nevus sebaceous exhibiting hyperkeratosis and papillomatosis, dilated follicular infundibulum plugged with keratin

The second lesion consisting of the erythematous, fleshy plaque revealed papillomatosis in the epidermis. Several cystic invaginations extend downward from the epidermis and numerous papillary projections extend into the lumina of the invaginations. (Figure 3 a,b,c) Papillary projections were lined by two rows of cells: an inner layer of high columnar cells with oval nuclei and faintly eosinophilic cytoplasm showing hints of decapitation secretion and an outer layer of small cuboidal cells with round nuclei and scanty cytoplasm. (Figure 4) Also present were dense cellular infiltrates composed nearly entirely of plasma cells in the stroma, papillary projections with dilated and congested blood vessels, apocrine and tubular glands in the deep dermis. (Figure 5) Final clinicopathologic diagnosis was Nevus Sebaceous with Syringocystadenoma papilliferum.

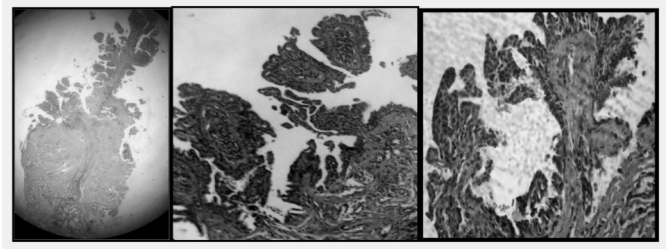

Figure 3 (a) Photomicrograph of a specimen taken from an erythematous fleshy plaque on the scalp shows SCAP characterized by cystic invaginations extending downward from the epidermis, numerous papillary projections, papillomatosis and mature apocrine glands in the lower dermis (b) On higher magnification are numerous papillary projections extending into the lumina of the invaginations (c) papillary folds projecting into the cystic spaces

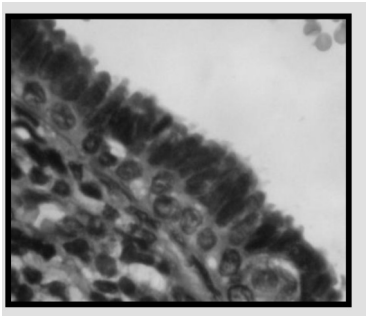

Figure 4 papillary projections lined by glandular epithelium consisting of 2 rows of cells: columnar cells with oval nuclei showing hints of decapitation secretion and cuboidal cells with round nuclei

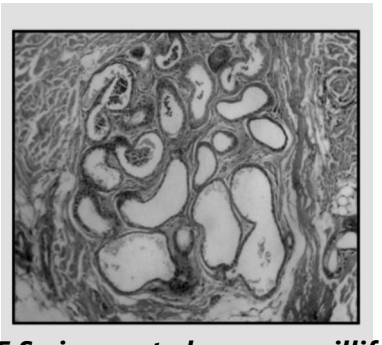

Figure 5 Syringocystadenoma papilliferum showing numerous apocrine glands in the deep dermis

Patient was referred to Pediatrics for general check-up and Ophthalmology for baseline eye exam. Physical examinations and routine laboratory work-ups revealed normal findings. Electrocautery with curettage of syringocystadenoma papilliferum was performed leaving a healed scar with no r ecurrence seven months post treatment. (Figure 1b) Patient was informed about possible occurrence of benign and malignant tumors that may complicate nevus sebaceous and was advised follow-up for any untoward clinical changes (bleeding or development of new lesions).

DISCUSSION

Nevus sebaceous is a benign lesion with epidermal, follicular, sebaceous and apocrine elements. Originally described by Jadassohn in 1895 as a hamartoma composed predominantly of sebaceous glands, it presents at birth as a solitary, yellowish, hairless patch mainly on the head and neck that becomes raised and papillomatous at puberty due to the effect of androgen on the glands.[4,5,] Mehregan and Pinkus described 3 clinico-pathologic

stages: 1) the early or infantile stage clinically appears as a smooth, yellowish-orange patch and is characterized by papillomatous epithelial hyperplasia and underdeveloped hairs, 2) the second, adolescent stage presents as thicker and verrucous lesions characterized by massive development of sebaceous glands, epidermal verrucous hyperplasia, and the maturation of apocrine glands, 3) the third or adult stage is characterized by the development of tumors of epidermal and adnexal origin such as syringocystadenoma papilliferum (5%), trichoblastoma (4.69%), trichillemomma (2%) and sebaceoma (2%). Other benign lesions reported include viral warts (2.4%), nevocellular nevus (0.83%), keratoacanthoma (0.67%), and seborrheic keratosis (0.50%).[4,6] The lifetime risk of malignant transformation of nevus sebaceous vary from 5 and 22% [7] Basal cell carcinoma, once thought to be the most common malignant tumor developed rarely (0.8%).[6] Thus, removal of nevus sebaceous for cancer prophylaxis in children is no longer recommended.[6] In the absence of signs for malignant transformation, the decision to undergo surgery would be based on safety, tolerability of anesthesia and the extent of excision required should there be increase growth of the lesion. Nevus sebaceous causes cosmetic deformity and alopecia.

Syringocystadenoma papilliferum is a hamartoma arising from pluripotent cells derived from apocrine or eccrine glands. Fifty percent are present at birth or infancy and commonly arises during puberty. [2,3] Seventy-five percent occurs on the head and neck although lesions on the trunk, axilla, genitalia and extremities have been reported.[8,9,10, 11] Three clinical types have been described: 1) the plaque type presenting as hairless area in the scalp that becomes nodular, verrucous or crusted during puberty and usually associated with nevus sebaceous, 2) the linear type consisting of groups of multiple, pink to red firm papules or nodules 1-10mm on the neck or face 3) the solitary nodular form appearing as domed, umbilicated or pedunculated lesions measuring up to 1cm on the trunk, shoulder, axilla and genital area. The two latter forms occur less often.[3] Diagnosis is by clinical suspicion and confirmed by histopathology, which shows epidermal papillomatosis, several cystic invaginations extending downward from epidermis and numerous papillary projections extending into the lumen of cystic invaginations. Invaginations and papillary projections are lined by two rows of cells: an outer layer of small cuboidal cells with basophilic nuclei and an inner layer of columnar cells exhibiting decapitation secretion. Apocrine glands are prominent in the dermis and inflammatory plasma cells are present in the stroma within the papillary projections.[3]

Syringocystadenoma papilliferum with nevus sebaceous has increased risk for malignant potential. The most common of which is basal cell carcinoma occurring in 10% of cases and is due to loss of allele at 9q22. Other tumors reported to develop include squamous cell carcinoma, verrucous carcinoma and ductal carcinoma. Long-standing syringocystadenoma papilliferum could evolve into a syringocystadenocarcinoma papilliferum. Complete surgical excision of syringocystadenoma papilliferum is recommended to eliminate risk of malignant degeneration. [1,3,12] Other treatment modalities include carbon dioxide lasers, Moh's micrographic surgery and electrocautery with curettage.

CONCLUSION

Syringocystadenoma papilliferum is a rare benign tumor wherein one-third of cases arise within a nevus sebaceous. There is a risk for malignant degeneration of syringocystadenoma papilliferum and complete surgical excision is the treatment of choice whenever possible. Close observation is recommended for nevus sebaceous. Surgical intervention may be required should there be signs of malignant degeneration.

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