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SYRINGOCYSTADENOMA PAPILLIFERUM IN NEVUS SEBACEOUS AT UNUSUAL SITE

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ABSTRACT

Nevus sebaceous is a benign, congenital hamartoma of the skin, with a predilection for the scalp and less commonly occurs on the face, around the ears, neck or on the trunk. In late childhood and early adulthood, there is a significant risk of developing secondary tumors, the most common of which are syringocystadenoma papilliferum and basal cell carcinoma. A case of syringocystadenoma papilliferum associated with nevus sebaceous in a 26 year old female patient in the nipple areolar region. A rare site of occurance of syringocystadenoma papilliferum arising in nevus sebaceous is described. To the best of our knowledge, this appears to be the first case reported in the nipple areolar complex.

Keywords: Nevus sebaceous, Syringocystadenoma papilliferum, nipple areola

INTRODUCTION

Nevus sebaceous is a congenital skin lesion commonly seen by pediatricians, dermatologists, and plastic surgeons.[1] It has a predilection for the scalp and less commonly occurs on the face, around the ears, neck, or on the trunk. The most common benign and malignant neoplasms arising in this disorder include trichilemmoma, syringocystadenoma papilliferum, keratoacanthoma, apocrine cystadenoma and basal cell carcinoma.

Syringocystadenoma papilliferum is a benign adnexal tumor of the skin,[2] which can occur either de novo or on some organoid nevus such as nevus sebaceous. This tumor presents generally over the scalp and forehead. The clinical presentation of nevus sebaceous and syringocystadenoma papilleferum varies widely but the histological appearance is uniform and characteristic, forming the basis of diagnosis.[3]

CASE REPORT

A 26 year old female patient presented with history of pigmented papules in bilateral nipple and areola region, since birth. Lesion formed an irregular nodule during puberty and progressed to a verrucous plaque, on right side, since one month.

On examination, her general condition was good. Local examination showed a pigmented, non tender, verrucous nodule in right nipple areolar complex, measuring 3.5x2.5x2cm. (Fig. 1)

Excision biopsy of the lesion was done on the right side and sent for histopathological examination. Grossly, the specimen was a single, irregular, warty mass measuring

3.5x2.5x2cm. Cut surface was grey white with tiny cysts.

Microscopy showed dilated, keratin filled infundibulum surrounded by numerous mature and immature sebaceous glands. (Fig.2) There were cystic invaginations from the surface, lined by stratified sqaumous epithelium transforming into a double layered epithelium. (Fig.3) Outer layer of epithelium was lined by cuboidal cells and the inner layer by columnar cells, a few of which showed decapitation secretion. Papillary projections into the cystic invagination were evident. Dense mononuclear inflammatory infiltrate, predominantly plasma cells were seen in the papillary core and around the cyst wall. (Fig.4) A good number of apocrine cysts were also seen. These classical histological features favored a diagnosis of Syringocystadenoma Papilliferum associated with Nevus Sebaceous.

DISCUSSION

Nevus sebaceous is a congenital hamartoma of the skin, first described by Jadassohn. It is defined by hyperplasia of the epidermis, hair follicles, sebaceous and apocrine glands. Nevus sebaceous often presents at birth, is quiescent in childhood and grows during puberty, probably due to increased levels of serum androgens present in neonatal period and at puberty.[4] It is sporadic and occurs with equal frequency in males and females of all races.[1] In a study by Rosen et al,[1] out of 631 cases, most common locations of nevus sebaceous were scalp, face or neck [Scalp (62.8%), Forehead (10.3%), Cheek (9.8%), Neck (3.7%), Eyebrow (1.7%), Back (1.5%), Eyelid (0.8%), Chest (0.5%), Abdomen (0.2%) and Breast (0.2%)]. In late childhood and early adulthood, there is a significant risk of developing secondary tumors, mainly syringocystadenoma papilliferum and basal cell carcinoma.[4]

Syringocystadenoma papilliferum, formerly known as nevus syringocystadenomatous papilliferus, is a proliferating tumor with mostly apocrine differentiation but eccrine differentiation can also be seen. The histogenesis of this rare neoplasm is still unclear. It is theorized that syringocystadenoma papilliferum arises from the pluripotent cells of apocrine lineage.[5] In about 75% of cases, syringocystadenoma papilliferum arises within a preexistent naevus sebaceous of Jadassohn according to a large series of 100 cases by Helwig and Hackney.[6] In their series,[6] 55 lesions were on the scalp, 11 on the forehead and temple, 5 on the face and 3 on the upper lip, but none was reported on the nipple areolar complex. Less frequently reported sites are chest, abdomen, arm, eyelid and thigh.[2][7][8]

Clinically, the differential diagnoses of these lesions include linear epidermal nevus, nevus comedonicus, basaloid follicular hamartoma, cylindroma and eccrine nevus.[9] However, in our case, histopathological features were unmistakable and had no differential diagnosis.

The only treatment for syringocystadenoma papilliferum is excisional biopsy, which also confirms the diagnosis. CO2 laser excision of syringocystadenoma papilliferum of the head and neck is a clinical treatment option in anatomic areas unfavorable to excision and grafting.[10]

CONCLUSION

Syringocystadenoma papilliferum in nevus sebaceous is well described, but a relatively rare tumor, diagnosed only by histopathological examination. We report a case of syringocystadenoma papilliferum associated with nevus sebaceous in the right nipple areolar region, an exceptional site of presentation.

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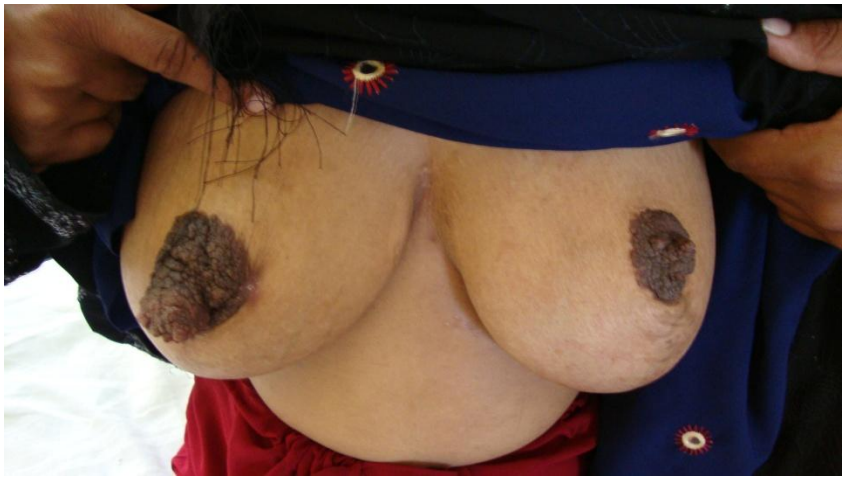

Fig.1: Clinical photograph showing bilateral verrucous lesion with nodule on right side

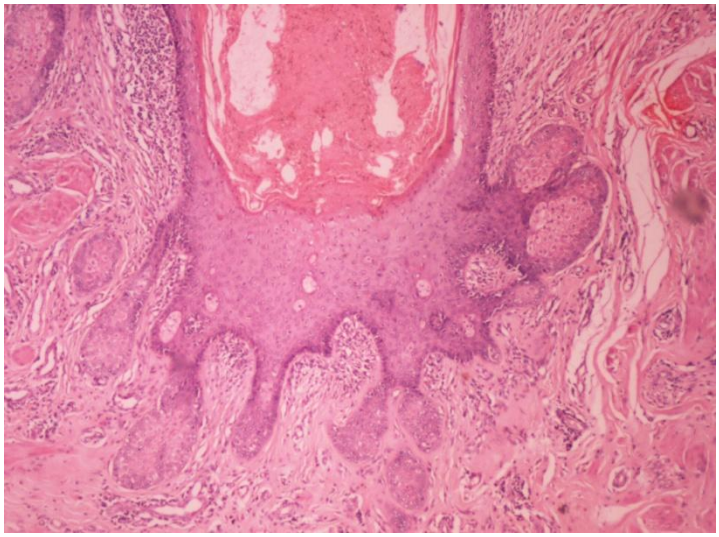

Fig.2: Photomicrograph of tissue section showing dilated keratin filled infundibulum with numerous mature and immature sebaceous glands. [HnE, 4x]

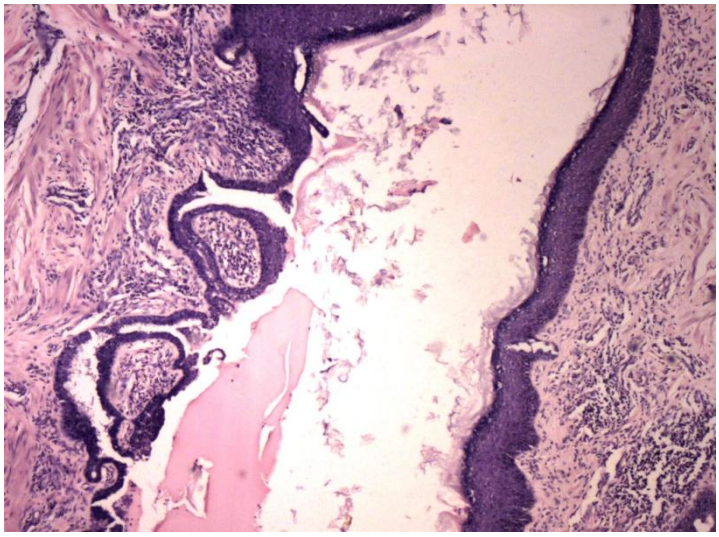

Fig.3. Photomicrograph of tissue section showing papillary projection with plasma cells [HnE,20x]

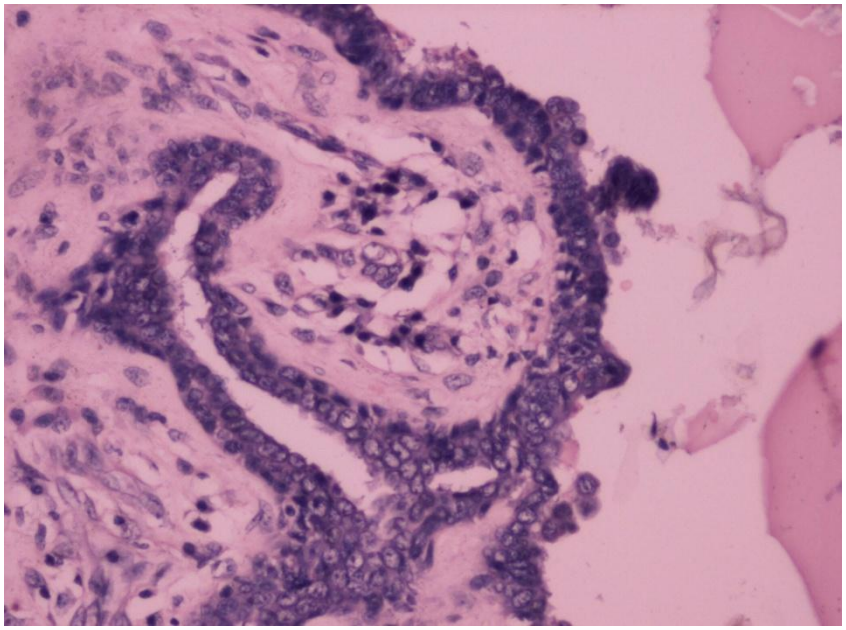

Fig.4: Photomicrograph of tissue section showing cystic invagination and transformation of stratified squamous lining to double layered epithelium.[HnE,10x]