Case Report

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Syringocystadenoma Papilliferum of Scalp: A Rare Case Report

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Abstract

Syringocystadenoma papilliferum also known as naevus syringocystadenomatosus papilliferus is an exuberant proliferating lesion, commonly seen on the scalp in association with an organoid naevus, and showing predominantly apocrine differentiation. It is a benign adnexal skin tumour of the apocrine or the eccrine type. It is relatively a rare neoplasm. These lesions may be present at birth or in childhood, but the majority are seen on the face and scalp of young adults. There is frequently a history of papillomatous expansion of a small pre-existing lesion at or around puberty and lesions often occur in a pre-existing organoid naevus. The lesion is composed of multiple warty papules, some of which are translucent and pigmented. The microscopic appearance is characteristic and shows ducts connecting to the surface, containing papillary processes and lined by two epithelial cell layers. Treatment of this tumor includes local excision in most of the cases. We report a case of syringocystadenoma papilliferum of the scalp in a 12-year-old female, which was clinically diagnosed at first as squamous cell carcinoma of the scalp but was later histologically confirmed as syringocystadenoma papilliferum.

Key words: Child, Scalp, Syringocystadenoma papilliferum

INTRODUCTION

Syringocystadenoma papilliferum is a rare benign hamartomatous adnexal tumour which originates from the apocrine or the eccrine sweat glands. It is relatively a rare neoplasm, predominantly a childhood tumour. In about 50% of those who are affected, it is present at birth, and in a further 15-30%, the tumour develops before puberty.[1] Syringocystadenoma papilliferum occurs with equal frequency in both sexes.[2]

Syringocystadenoma papilliferum occurs most commonly on the scalp or the face. Presentation with multiple lesions is rare; those arising outside the head and neck region are even more uncommon. The lesion of syringocystadenoma papilliferum usually measures between 1 and 3 cm and <4 cm in diameter.[3] The tumour has varied clinical presentations.

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We present a case of syringocystadenoma papilliferum in a 12-year-old female child presented with an ulcer on scalp.

CASE REPORT

A 12-year-old female child presented with the complaint of an ulcerative lesion with alopecic patch over scalp associated with itching and bleeding. Initially, the patient had a nodule of around 1 cm over the right side of the parietal region of the scalp, which did not increase in size. There was no growth of hair over the swelling. The nodule turned into a 1.5 cm ulcer in the past 1 year, which gradually increased in size. There was no regional lymphadenopathy. No other skin lesions were noted elsewhere. Skin sensations were normal. A presumptive clinical diagnosis of squamous cell carcinoma was made. The patient was worked up. Her routine investigation were normal.

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Patient underwent excision reconstruction under general anaesthesia. The lesion was excised completely with a normal margin of around 1 cm and with a depth up to the subcutaneous plane. Gross sample was sent to our department for histopathological examination.

Pathologic Finding

Gross

Single gray white soft tissue mass measuring 5.5 cm × 2 cm × 1.5 cm along with skin attach on one side. Skin is irregular. On cut surface gray white verruciform growth measuring - 2.5 cm × 1 cm present.

Microscopic

Section shows epidermis and dermis. At one place, ulcerative area is present in the epidermis. The epidermis shows varying degrees of papillomatosis. Cystic invaginations extend downwards from the epidermis, with numerous villous projections extending into the lumen of the cyst (Figure 1).

The upper portion of the invaginations and large segments of the cystic invaginations are lined by squamous, keratinizing cells similar to those of the surface epidermis (Figure 2). In the lower portion of the cystic invaginations, numerous papillary projections extend into the lumina of the invaginations. The papillary projections and the lower portion of the invaginations are lined by glandular epithelium consisting of two rows of cells (Figure 3). The luminal row of cells consists of high columnar cells with oval nuclei and eosinophilic cytoplasm. The outer row of cells consists of small cuboidal cells with round nuclei and scanty cytoplasm. Dense plasma cell infiltrate is present in the stroma of the papillary projections (Figures 3 and 4).

Beneath the cystic invaginations, deep in the dermis, groups of tubular glands with large lumina are present. The cells lining the large lumina are flattened showing evidence of active decapitation secretion (Figures 1 and 3).

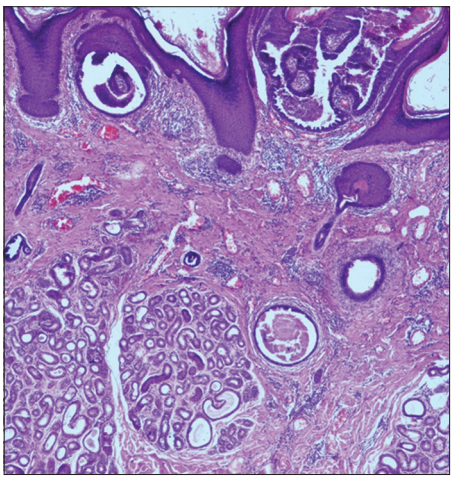

Figure 1: Epidermis with ulceration, papillomatosis with cystic invaginations downward from epidermis, with numerous villous projections extending into the lumen of the cyst

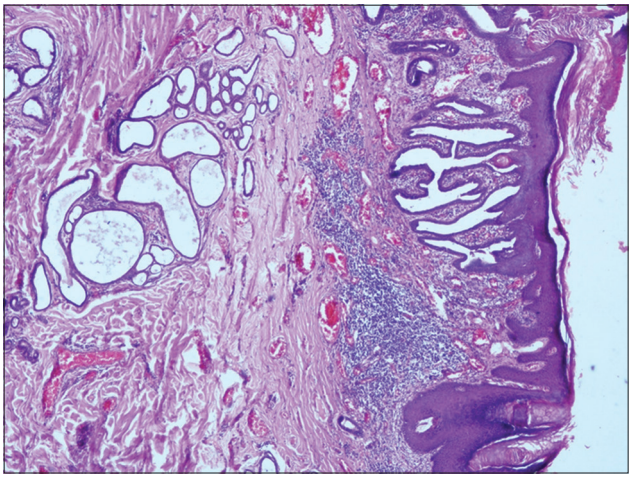

Figure 2: Cystic invaginations lined by squamous, keratinizing cells

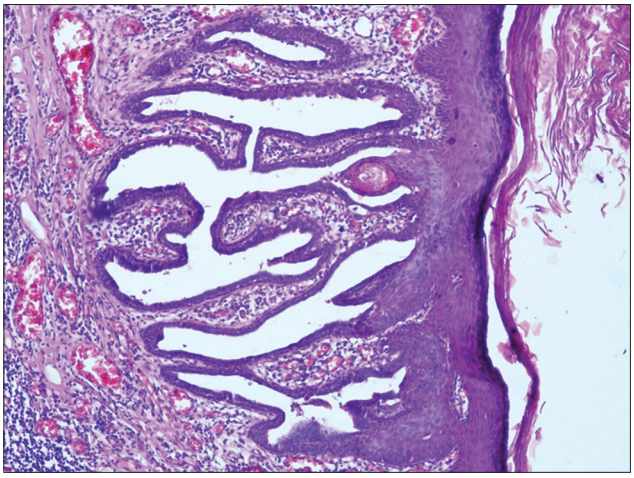

Figure 3: Papillary projections lined by glandular epithelium consisting of two rows of cells inner columnar and outer cuboidal cells. Stroma shows dense plasmacytic infiltrate

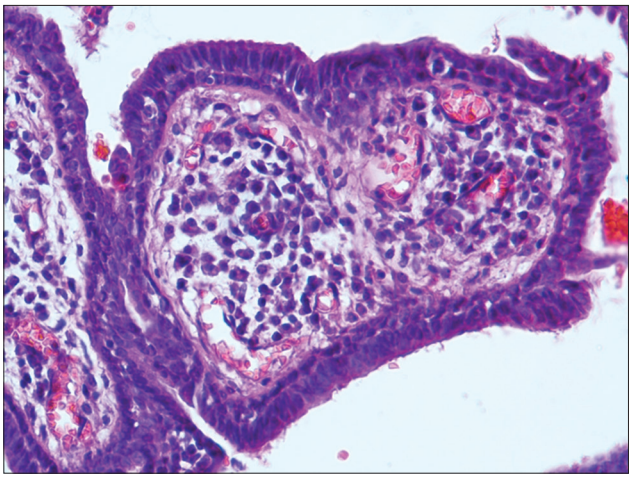

Figure 4: Plasma cell infiltration and tubular glands in the dermis with secretion

On the basis of gross and histopathological examination, a diagnosis of syringocystadenoma papilliferumwas offered.

DISCUSSION

Syringocystadenoma papilliferum is a rare non-malignant adnexal sweat gland neoplasm characterized by asymptomatic, skin-colored to pink papules or plaques with a highly variable appearance, most commonly in the head and neck area. Syringocystadenoma papilliferum occurs most commonly on the scalp or the face; however, tumours may be seen in other locations including the vulva,[4] external ear,[5] lower leg,[6] scrotum[7] and breast[8] in about one fourth of the cases. It is usually first noted at birth or in early childhood and presents as a papule or several papules in a linear arrangement or as a plaque. The lesion increases in size at puberty, becoming papillomatous and often crusted. On the scalp, syringocystadenoma papilliferum frequently arises around puberty within a nevus sebaceous that has been present since birth.

Three clinical types have been described:

a. Plaque type: Presenting as an alopecic patch on the scalp and may 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
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 5-10 mm in size with a predilection for the trunk shoulder and axillae

Approximately one-third of cases of syringocystadenoma papilliferum arise in organoid naevi which is a precursor lesion for it.[2]

Yamamoto *et al.*[9] postulated an origin in pluripotent cells on immunohistochemical and ultrastructural grounds. Böni *et al.*[10] showed mutations in PTCH or P16 tumour suppressor genes in syringocystadenoma papilliferum. Kazakov *et al.* noted an overlap with tubular adenoma.[11]

Histopathologically, the epidermis shows varying degrees of papillomatosis. One or several cystic invaginations extend downward from the epidermis.

Frequently, there are malformed sebaceous glands and hair structures in the lesions of syringocystadenoma papilliferum. In about one-third of the cases, syringocystadenoma papilliferum is associated with a nevus sebaceous. In about 10% of the cases, a basaloid epithelial proliferation resembling basal cell carcinoma develops, but this is noted only in lesions that also exhibit a nevus sebaceous.

A few instances of transition of a syringocystadenoma papilliferum into an adenocarcinoma with regional lymph node metastases have been reported.[12]

Additional studies syringocystadenoma papilliferum can exhibit both apocrine and of eccrine differentiation. For example, positive immunoreactivity for gross cystic disease fluid proteins 15 and 24 and zinc-2 glycoprotein demonstrates evidence of apocrine differentiation.[13] On the other hand, immunohistochemical analysis of cytokeratins in syringocystadenoma papilliferum demonstrates similarities to eccrine poromas and the ductal component of eccrine glands.[14] In addition, light and electron microscopic features of some lesions show evidence of eccrine differentiation. It is probable that syringocystadenoma papilliferumarises from undifferentiated cells with the potential to exhibit both apocrine and eccrine modes of epithelial secretion. Most lesions of syringocystadenoma papilliferum exhibit apocrine differentiation; however, some demonstrate eccrine features. Studies have demonstrated loss of heterozygosity for patched and p16, a negative regulator of the cell cycle, in syringocystadenoma papilliferum, suggesting that these molecules may play a role in the pathogenesis of these lesions.[10]

The only treatment for syringocystadenoma papilliferum is excision biopsy, which also confirms the diagnosis. CO_2 laser excision of syringocystadenoma papilliferum of the head and neck is a clinical treatment option in anatomic areas unfavorable to excision and grafting. Syringocystadeno carcinoma papilliferum has been successfully treated with Moh's micrographic surgery.

CONCLUSION

Syringocystadenoma papilliferum is a rare neoplasm, present most commonly on head and neck area. In the present case, it was clinically diagnosed at first as squamous cell carcinoma of the scalp, but later, it was histologically confirmed as syringocystadenoma papilliferum. Such a presentation of this tumour may include multiple differential diagnoses on clinical background so it must be sent for a histopathological examination to confirm the diagnosis. The excision of the tumor is sufficient in most of the cases.

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