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Case Report

Linear syringocystadenoma papilliferum of neck: a rare variant adnexal tumour

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ABSTRACT

Syringocystadenoma papilliferum is a rare benign adnexal tumour presenting as exuberant proliferating lesion showing differentiation in apocrine pattern. The three clinical variants are linear, plaque and solitary of which linear variant is the rarest. We report a case of linear variant of syringocystadenoma papilliferum, which had typical histopathological features. As there is risk of malignancy in long standing cases, early diagnosis and treatment is of utmost importance.

Keywords: Benign adnexal tumour, Apocrine pattern, Linear, Rare variant, Malignancy

INTRODUCTION

Syringocystadenoma papilliferum is a rare benign adnexal tumour presenting as exuberant proliferating lesion showing differentiation in apocrine pattern.[1] It presents at birth in 50% of cases with equal frequency among both genders.[1] Most common site is head and neck region.[2] The three variants are linear, plaque and solitary of which linear variant is the rarest.[3] We report a case of linear variant of syringocystadenoma papilliferum over left side of neck since birth.

CASE REPORT

A 42 year old male presented with history of three asymptomatic linearly arranged nodules, over left side of neck since birth, which increased in size gradually with occasional itching and blood stained discharge on and off for past few months. There was no systemic complaints. On cutaneous examination, there were three nodules of size 1.5×1.5 cm arranged linearly, covered with hyperpigmented crust over left side of neck (Figure 1). There was no associated regional lymphadenopathy. Excisional wedge biospsy was done and specimen was sent for histopathological examination.

Histopathological section studied showed numerous papillary projection of epidermis forming cystic invaginations into dermis (Figure 2). Papillary projections lined by 2 rows of cells composed of inner columnar cells and outer cuboidal cells with apocrine decapitation (Figure 3). Stroma showed inflammatory cells composed of numerous plasma cells (Figure 4). All these microscopic features showed characteristic features of syringocystadenoma papilliferum.

Based on both clinical and histopathological features, final diagnosis of syringocystadenoma papilliferum was made. Patient was referred to plastic surgeon for complete surgical excision. He was followed up for a period of 12 months and there was no evidence of recurrence.

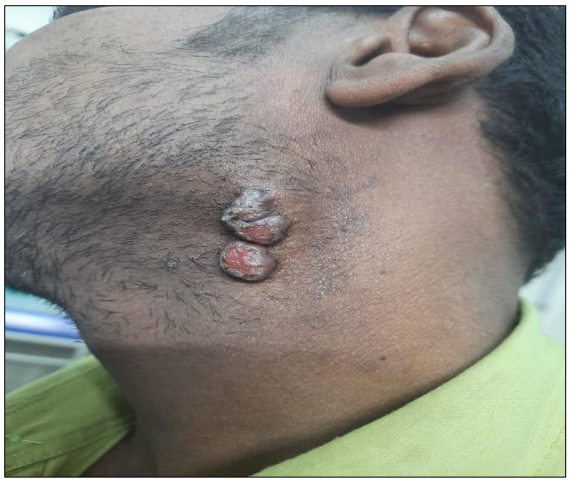

Figure 1: Three linearly arranged erythematous Nodules covered with crusting present over lateral aspect of left side of neck.

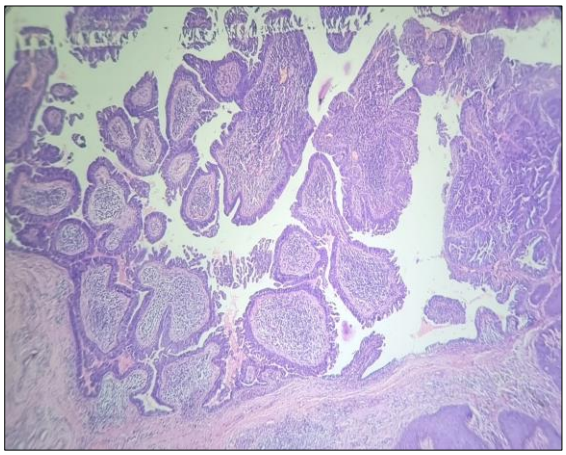

Figure 2: Numerous papillary projections of epidermis forming cystic invaginations into the dermis. (H and E stain, 10x magnification).

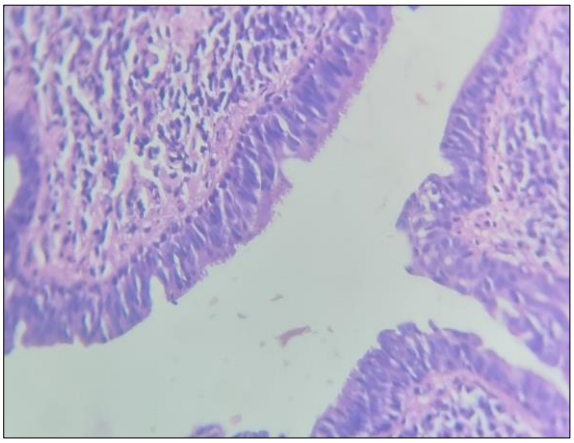

Figure 3: The papillary projections are lined by two rows of cells composed of inner columnar cells and outer cuboidal cells with apocrine decapitation (H and E stain, 40x magnification).

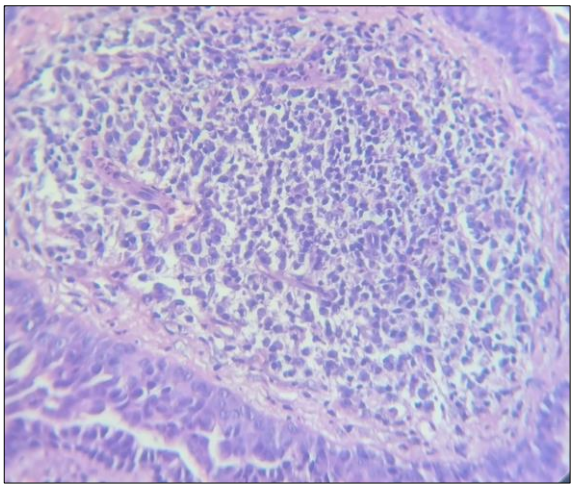

Figure 4: The stroma shows inflammatory cells composed of numerous plasma cells (H and E stain, 40x magnification).

DISCUSSION

Syringocystadenoma papilliferum was first described as Nevus syringocystadenomateous papilliferum by John Stokes in 1917.[3] It mostly present at birth and also at puberty, so called as childhood tumour.[4] Among the three clinical types, plaque, linear and solitary, linear type is the rarest.[3] our case, is a linear type of syringocystadenoma papillifereum since birth. Linear type usually presents as erythematous papule, plaque or nodule over head and neck region, which increases in size at puberty.[3] Its origin of development is from pluripotent cells and commonly exhibits apocrine differentiation.[4]

In our case, clinical differential diagnosis included cutaneous tuberculosis, nodular variant of Basal cell carcinoma and squamous cell carcinoma. Histological differential diagnosis included: apocrine hidrocystoma which shows large cystic spaces, lined by columnar cells and peripheral layer of myoepithelial cells with apocrine decapitation secretion.[5] The other histopathological differential diagnosis was tubular apocrine adenoma which shows numerous irregularly shaped tubular structures lined by luminal layer of columnar cells and peripheral layer of cuboidal cells.[5] In our case, histopathology shows characteristic features of Syringocystadenoma papilliferum. Immunohisto-chemistry helps to differentiate origin of tumour as apocrine or eccrine.[1] It was not done in our case due to feasibility issues.

Malignancy is reported to occur in 10% of cases, mostly with Basal cell carcinoma.[1] Syringocystadenocarcinoma papilliferum is the malignant counterpart of syringocystadenoma papilliferum.[6] It manifests as rapid increase in size, bleeding, ulceration and histological features shows asymmetry, poorly circumscribed lesion with increased mitotic activity.[6]

CONCLUSION

Linear syringocystadenoma papilliferum is a rare variant and hence we report this case. As there is risk of development of malignancy in long standing cases, early diagnosis and treatment is of utmost importance.

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