

PhotoDermDiagnosis

Linear growth over the thigh in a young girl

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A 16-year-old female presented with multiple asymptomatic nodular swellings over back of left thigh since birth which increased in size over the last 4 years associated with pain and bloody discharge from the same. There were no complaints referable to other systems and no family history of similar lesions. Examination revealed multiple verrucous nodular lesions with papillomatous exuberant friable surface arranged in a linear pattern over the posterior aspect of left thigh which ranged in size from 1 cm to 5 cm in diameter (**Figure 1**). There was no significant lymphadenopathy. Rest of the cutaneous examination was normal. Systemic examination did not reveal any abnormality.

A diagnostic biopsy was taken from one of the nodular lesions. Sections were stained with hematoxylin and eosin. Histopathological



Figure 1 Multiple nodular verrucous lesions with a papillomatous surface arranged in a linear fashion.

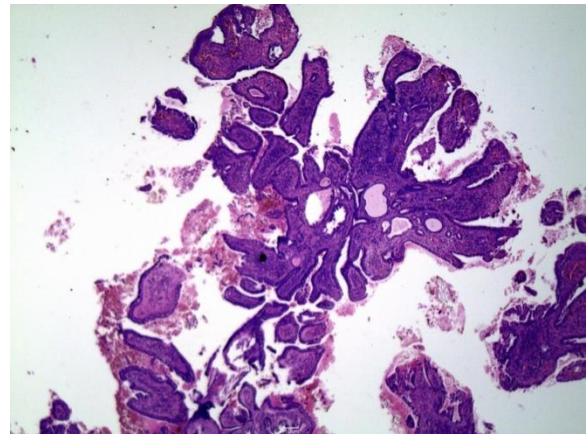


Figure 2 Invaginations in the epidermis with multiple papillary projections (Hematoxylin and eosin, x100).

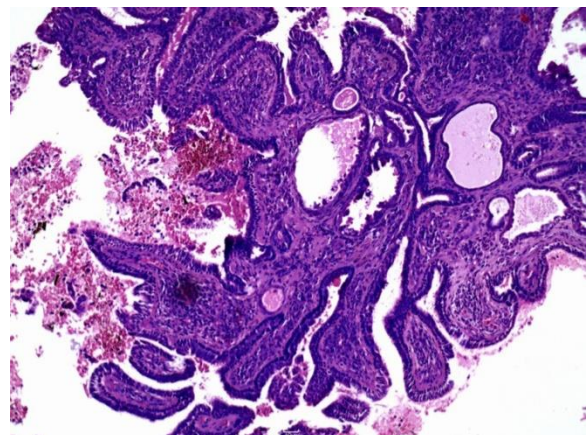


Figure 3 Papillae in the epidermis lined by bilayered epithelium with mixed infiltrate of lymphocytes, plasma cells and neutrophils in the dermis (Hematoxylin and eosin, x400)

features are shown in **Figure 2** and **3**. What is your diagnosis?

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Diagnosis

Syringocystadenoma papilliferum

Histopathology showed a single crateriform invagination connected with surface epidermis that was lined by double layer of epithelium. The epithelium showed apocrine secretion towards the lumen whereas towards the base the cells were cuboidal. The floor of this invagination was thrown into the folds and papillary projections at places formed solid areas. Within these solid areas were seen elongated as well as rounded ductal structures. The surrounding dermis showed a mixed infiltrate of lymphocytes, plasma cells and neutrophils

Discussion

A diagnosis of syringocystadenoma papilliferum (SCAP) was made based on the clinical presentation and the histology. SCAP is a benign hamartomatous adnexal tumor with unknown etiology. It is thought that most of the lesions are apocrine and some are eccrine in differentiation.¹ Tumor may occur *de novo* (75% of cases) or within a nevus sebaceous.² It usually presents as a papular lesion or a smooth hairless plaque on the scalp and forehead. Most of the lesions appear at birth or appear in infancy and may become elevated and nodular, verrucous or crusted at puberty. Less common presentation include lesions on unusual anatomic sites e.g. trunk, vulva, thighs, breast, eyelids or axilla.³ A mature lesion usually comprises of a single, papillomatous, verrucous, sometimes erosive plaque or nodule measuring 1-3 cm in diameter, sometimes multiple or linearly arranged cutaneous nodules are also seen.² Ulceration or rapid enlargement may indicate malignant transformation, usually to basal cell carcinoma (10% of the cases), rarely adenocarcinoma.³

Syringocystadenocarcinoma papilliferum is the malignant counterpart characterized by solid areas and cytologically malignant cell. The differential diagnosis includes tuberculosis verrucosa cutis, linear epidermal nevus, verrucous carcinoma, eccrine nevus, pyogenic granuloma.

Excisional biopsy is the gold standard in diagnosis. Microscopic examination is characteristic and shows papillary projections and epidermal invaginations. The papillary projections and lower portion of invaginations are lined by glandular epithelium often consisting of two rows of cells. The luminal row of cells consists of high columnar cells with oval nuclei and faintly eosinophilic cytoplasm. The outer row of cells consists of small cuboidal cells with round nuclei and scanty cytoplasm.⁴ Another diagnostic feature of this neoplasm is the presence of mononuclear inflammatory infiltrate consisting of mainly plasma cells in the fibrous tissue of the papillary projections.⁴ Positive staining in the luminal cells for alcian blue, colloidal iron, and periodic acid-Schiff (PAS) which is diastase resistant favor the apocrine differentiation of this tumor.⁴ The treatment of choice is surgical excision.² Other treatment options available are CO₂ laser and Mohs micrographic surgery.^{2,5} SCAP is a benign tumor but the excisional biopsy must be done to rule out the malignant transformation.

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