Read Item - Appendageal Tumours With Apocrine Differentiation

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Abstract: Doctor's resource on Appendageal Tumours With Apocrine Differentiation

Appendageal Tumours With Apocrine Differentiation

Introduction

Apocrine derived tumours are rare. Those that affect the scalp are syringocystadenoma papilliferum, apocrine hidrocystoma and tubular apocrine adenoma. Cylindromas are probable eccrine derived.

Clinical Features and Investigation

Apocrine hidrocystoma is a cystic dilatation of the apocrine secretory duct that presents as a small (less than 10 mm), solitary, well defined, dome shaped, translucent nodule with a blue hue. They occur most commonly on the outer canthus of the eye and the penis, but can also be found on the scalp, ear, chest or shoulders. Multi-lobulated lesions can also occur. Histology shows one or several large cystic spaces lined by a row of secretory epithelium that shows decapitation secretion . Papillary projections can delve into the cystic spaces.

Syringocystadenoma papilliferum is an exuberant proliferating lesion that is most commonly seen on the scalp and is accompanied by alopecia. It may appear at birth as a solitary translucent or pigmented papule with a blue hue, or as several papules in a linear arrangement following Blaschko's lines. Extensive verrucous plaques may develop at puberty when the congenital lesions enlarge and become papilomatous and crusted with androgen stimulation. About one third of syringocystadenoma papilliferum appear later in life in association with an organoid naevus.

Histology shows epidermal papillomatosis, and beneath this a cystic invagination extends downwards into the dermis. Numerous papillary projections extend into the lumen of the cystic invagination. The cyst is lined by a double layer of columnar cells showing decapitation secretion and the underlying stroma contains abundant plasma cells.

Tubular apocrine adenoma is a very rare tumour that occurs most commonly in the scalp or the axilla. It presents as a dermal nodule with gradually enlarges and may grow as big as 7 cm in diameter. Histology shows a poorly circumscribed dermal tumour connected to the epidermis. The tumour is composed of lobular masses that contain a central tubular structure. The tubular lumen is lined by a double layer of cells, the inner columnar cell layer shows

decapitation secretion, while the outer consists of myoepithelial cells. A malignant counterpart of this condition also exists.

Hidroadenoma papilliferum is a tumour of the anogenital region of women, presenting as a vulval or perianal tender nodule that sometimes bleeds. They do not occur on the scalp, but can occur on the nipple or eyelid. Histology shows a well circumscribed tumour in the dermis surrounded by a fibrous capsule, and with no connection to the overlying epidermis. The tumour consists of a large cystic space with numerous papillary folds projecting into the lumen and lined by apocrine epithelium.

Management

A diagnostic excisional biopsy is usually required.

Key Points

Apocrine cystadenoma, syringocystadenoma papilliferum and hidroadenoma papilliferum are all benign appendageal tumours of apocrine origin with distinctive histological features. Clinically a blue hue to a translucent nodule may suggest that it is of apocrine origin.