Read Item - Appendageal Tumours With Eccrine Differentiation

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

Appendageal Tumours With Eccrine Differentiation

Introduction

Numerous benign appendageal tumours of eccrine differentiation have been described (table 13.15). Only those affecting the scalp will be described here. Most are rare and many are very rare.

Clinical Features and Investigation

Cylindromas are dome shaped nodules that mainly occur on the scalp and forehead of middle aged females. They arise from the intradermal coiled duct region of the eccrine gland. Cylindromas may be single or multiple (turban tumours), bald, smooth, firm, pink, pedunculated slow-growing tumours that reach up to 5 cm in size. Occasionally tumours are painful and in some families they occur as an autosomal dominant trait with variable penetrance. They can be inherited in conjunction with multiple trichoepitheliomas, parotid adenomas and eccrine spiradenomas. Long standing lesions may become locally aggressive or occasionally undergo malignant transformation, however metastasis is very rare.

On histology the low power appearance of these tumours is characteristic. Isolated islands of baseloid cells are surrounded by thick PAS positive hyaline basement membrane, and fit together like the pieces of a jigsaw puzzle. The islands contain palisading small dark cells at the periphery and centrally there are larger cells with pale vesicular nuclei. Small duct like structures may be present in the centre of the lobules. Malignant degeneration is characterised by loss of the PAS positive basement membrane and very large cellular islands almost completely composed of the larger pale cells.

Syringomas are adenomas of the intraepidermal eccrine ducts. They present as multiple asymptomatic, skin-coloured or pale yellowish-brown dermal papules, 1 to 5 mm in size on the lower eyelids and cheeks of young women. Occasionally syringomas are seen on the chest and abdomen, and linear forms and eruptive generalised forms, which can mimic eruptive granuloma annulare may occur. There is a rare form of syringoma that presents as plaques of cicitricial alopecia with a papular surface. A palmer-planter keratoderma may be associated. In addition occult syringomas may produce a diffuse non-scarring alopecia.

Histology shows a dense dermal stroma containing small comma-shaped (or tadpole-like) islands of baseloid cells, which contain a tubular lumen. These islands are eccrine ducts with a two layer cell lining and contain amorphous debris in the central lumina. Within tumour, there may also be solid strands of baseloid cells independent of the ducts, and milia-like keratin cysts near the skin surface.

Chondroid syringomas are solitary, slow growing firm intradermal or subcutaneous nodules, between 0.5 and 3 cm in diameter. They occur most commonly on the head and neck of the elderly, but can also be seen on the trunk.

Histologically two types of chondroid syringoma are recognised: firstly chondroid syringoma with small tubular lumina that shows a circumscribed dermal tumour containing numerous small islands of epithelial cells and duct-like structures in a myxoid, chondroid and fibrous stroma; and secondly the more common chondroid syringoma with tubular branching lumina. This contains larger tubular structures as well as more solid islands of epithelial cells within the dense stroma.

Eccrine hidrocystoma is a cystic dilatations of the eccrine duct, into which eccrine sweat empties. They usually present as a solitary cystic, pale blue dome shaped papules around the

cheeks and eyelids of a young women. Occasionally lesions are multiple, with up to 200 hidrocystomas can be seen on the one patient. The lesions may wax and wane, becoming more prominent in the summer when sweat production is increased. The cysts in the dermis are unilocular and lined by two layers of cuboidal cells resembling an eccrine duct.

Eccrine spiradenoma presents as a solitary grey pink nodule on the head and neck, trunk or less commonly the extremities. They occur in adults and are often painful. Giant forms, linear lesions and multiple lesions have been described, as has their occurrence in association with cylindroma. Histology shows one or more well demarcated basophilic nodules in the dermis and subcutis surrounded by a thin fibrous capsule. The lobules are composed of two cell types admixed together and arranged in cords, and some contain duct like structures. Large pale cells are predominantly located in the centre of the lobules and outnumber the small baseloid cells found towards the periphery.

Eccrine Poroma is a tumour of the intraepidermal portion of the eccrine duct. I presents as a juicy dome shaped nodule that is slightly scaly, usually on the palms or soles, but may occur anywhere. The histology shows marked epidermal acanthosis, consisting of small regular cuboidal baseloid cells. These lesions are sometimes mistaken histologically for a superficial BCC. **Dermal duct tumour** belongs in the eccrine poroma group. It is a benign proliferation of the eccrine dermal duct in the papillary dermis. It is composed of small, cuboidal, baseloid cells that are regular in size and shape. It resembles a poroma, but is located in the dermis and without any epidermal connection. They present as a firm nodule on the head and neck region or the lower limbs, occasionally with overlying verrucous change.

Microcystic adnexal carcinoma is a particularly aggressive tumour that generally occurs on the upper lip and face, but has been described on the scalp. It affects adults of all ages. Clinically the lesions may be a relatively inconspicuous elevated plaque that may be painful due to neural invasion. Histology shows cords of small adnexal-type keratinocytes with ductal differentiation in a very sclerotic desmoplastic stroma. Horn cysts and abortive hair follicles are seen in some areas, while other areas may resemble syringoma. Microcystic adnexal carcinomas are locally invasive and Mohs surgery is generally recommended.

Eccrine carcinoma presents as a slow growing infiltrating plaque on the scalp. Lesions behave like morphoeic BCC and local recurrence following excision is common, however metastasis is rare. Many consider these carcinomas to simply be BCC with eccrine differentiation. On biopsy it is composed of branching tubular structures lined by atypical baseloid cells and embedded in a light dermal stroma. Many of the ducts are comma shaped and resemble those seen in syringoma. Neurotropic spread is common and is associated with pain in the lesion and with a worse prognosis.

Primary cutaneous adenocystic carcinoma is a very rare tumour with a predilection for the scalp and chest. Both local recurrence and metastasis are common. Histologically it closely resembles adenoid cystic carcinoma of the salivary gland. The tumour is composed of islands and cords of baseloid cells that contain tubular structures within. There is abundant sialomucin in the tubular structures and diffusely in the lobules. These tumours should be differentiated from metastasis either from a salivary adenoid cystic carcinoma or a large intestinal mucinous adenocarcinoma.

Mucinous eccrine carcinoma is a slow growing tumour that usually occurs on the face, scalp, axilla or trunk of an older individual. The morphology is of a painless, red-brown nodule, 0.5 to 1 cm in diameter that tends to recur locally and metastasises early. Histology shows the tumour is divided into a number of compartments. In each compartment islands of epithelial tumour cells float in a sea of mucin and are connected by fine fibrovascular septae.

Management

Surgical excision provides a diagnosis and definitive treatment for solitary eccrine tumours. A punch biopsy of a single small lesion also provides a diagnosis when there are multiple lesions. Local destructive therapies, such as cautery, trichloroacetic acid and cryosurgery can be used to produce a favourable cosmetic outcome for unsightly multiple lesions.

Eccrine carcinomas tend to be aggressive and complete surgical extirpation is recommended,

if achievable.

Key Points

A number of eccrine tumours can occur on the head and neck regions. Syringomas, cylindromas, chondroid syringomas, eccrine spiradenomas and eccrine hidrocystomas have a characteristic morphology and can often be diagnosed clinically. The other eccrine tumours may require histology for a clinical diagnosis.

Table 13.15 Classification of Eccrine Tumours

Benign	Malignant
eccrine hamartomas	microcystic adnexal carcinoma
eccrine hidrocystoma	eccrine carcinoma
papillary eccrine adenoma	adenoid cystic carcinoma
aggressive digital papillary adenoma	mucinous eccrine carcinoma
chondroid syringoma	malignant chondroid syringoma
syringoma	malignant cylindroma

cylindroma

eccrine spiradenoma

eccrine poroma

dermal duct tumour

hidroacanthoma simplex

syringoacanthoma

syringofibroadenoma

hidradenoma (acrospiroma)

malignant eccrine spiradenoma

malignant eccrine poroma

Hidradenocarcinoma

eccrine ductal adenocarcinoma

other sweat duct carcinomas

(after Weedon)