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REVIEW ARTICLE

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Cutaneous Tumours of Pilar Origin with Associated Syndromes

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Abstract:
The normal skin is lined by epidermis , dermis & subcutis other appendages.Skin appendages develops from ectoderm during early embryonic life.These specialized structures are located deep within the dermis & in subcutis.Includes pilo-sebaceous units(hair follicles & sebaceous glands), eccrine & apocrine glands.Hair follicleshow invagination from epidermis.Hair bulb is highly vascular part and is seen in reticular dermis.The inner mitotically active cells lines the dermal papillae undergo keratinization- hair shaft and inner root sheath.Each hair shaft consists of an innermost medulla, surrounded by highly keratinized cortical layer, and an outermost thin layer - cuticle.The outer 2 layers of hair bulb form the outer root sheath, containing glycogen-rich (clear) cells and is separated from the dermal connective tissue by a thick glassy membrane. Adnexal tumor is a benign or malignant lesion with the differentiations .The lesions may be single or multiple.Some tumors may be associated with syndromes (familial or sporadic).Malignant lesions are rare, locally aggressive ,shows nodal involvement & distant metastasis. The tumours are divided based on the follicular differentiation into benign, malignant and hyperplastic lesions. The benign neoplasms are Trichoblastoma, Pilomatricoma, Trichilemmoma, Trichofolliculoma, Fibrofolliculoma. The malignant lesions are Pilomatriceal carcinoma and proliferating trichillemaltumours. The various types of the differentiationsincludes, hair germ, infundibular, outer root sheet, matricel, follicular and mesenchymal. There are several syndromes associated with the pilar origin tumours with the gene abnormalities.

WHO histological classification of appendageal tumours

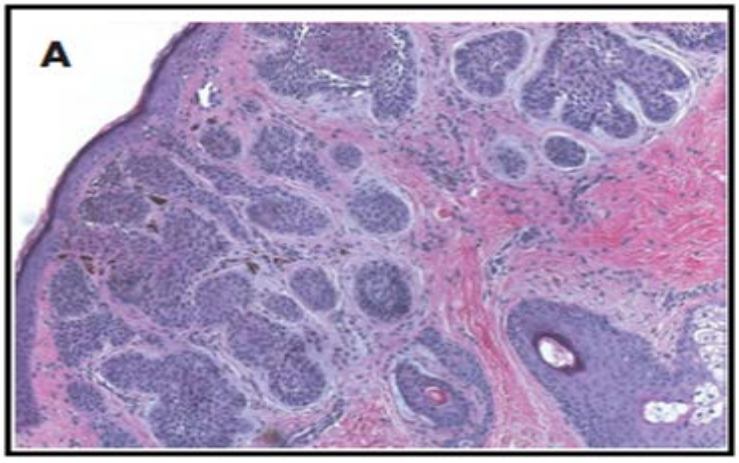
Tumours with apocrine and eccrine differentiation		Tubular adenoma	8211/0
Malignant tumours		Tubular papillary adenoma	8263/0
Tubular carcinoma	8211/3	Syringocystadenoma papilliferum	8406/0
Microcystic adnexal carcinoma	8407/3	Hidradenoma papilliferum	8405/0
Porocarcinoma	8409/3	Mixed tumour (chondroid syringoma)	8940/0
Spiradenocarcinoma	8403/3		
Malignant mixed tumour	8940/3	**Tumours with follicular differentiation**	
Hidradenocarcinoma	8400/3	***Malignant tumours***	
Mucinous carcinoma	8480/3	Pilomatrical carcinoma	8110/3
Digital papillary carcinoma	8408/3	Proliferating tricholemmal tumour	8103/1
Adenoid cystic carcinoma	8200/3	***Benign tumours***	
Apocrine carcinoma	8401/3	Trichoblastoma	8100/0
Paget disease of breast	8540/3	Pilomatricoma	8110/0
Extramammary Paget disease	8542/3	Tricholemmoma	8102/0
		Multiple tricholemmomas	8102/0
Benign tumours		Trichofolliculoma	8101/0
Hidrocystoma	8404/0	Fibrofolliculoma / trichodiscoma	8391/0
Syringoma	8407/0		
Poroma	8409/0	**Tumours with sebaceous differentiation**	
Syringofibroadenoma	8392/0	Sebaceous carcinoma	8410/3
Hidradenoma	8402/0	Sebaceous adenoma	8410/0
Spiradenoma	8403/0	Sebaceoma	8410/0
Cylindroma	8200/0	Cystic sebaceous tumour	8410/0

TUMOURS OF PILAR ORIGIN

Basaloid Follicular Hamartoma:-

It is a unique benign lesion and associated with systemic and cutaneous disorders. Inherited (AD) or acquired. The lesions are solitary in acquired cases. They involve middle aged people and the the most common site is scalpand face. The systemic diseases associated are myasthenia gravis, BCC, SLE and alopecia. The mode of inheritance is seen in adulthood. Clinically the lesion will present as multiple, widely spread, small flesh-coloured or pigmented papules. The other clinical features associated are alopecia and cystic fibrosis.

Microscopically, appears as multifocal proliferation of small basaloid cells in the cords, lobules, anastomosing strands &embeded in mucinous stroma. The peripheral palisading of the basaloid cells is characteristic features, Foci of keratinization & horn cyst & abortive hair follicles, makes BFH indistinguishable from trichoepithelioma. The differential diagnosis includes basal cell carcinoma, by the basaloid proliferation. The mitoses, decreased single-cell necrosis are the discriminating features from Basal cell carcinoma.



Basaloid differentiation

TRICHOFOLLICULOMA

It is a benign hamartomatous lesions of hair folllicles. The most preferred site is face. The lesion shows poor differentiation of pleuripotent stem cell towards hair follicles. They are small hair like follicles, radiates around one or more cyst like dilatations that opens to skin surface. The most common presentations are small, skin covered nodule having a small central ostium in which hair emerges. The lesion starts to develop at any age and diagnosed at second decades.

Microscopically, they are mainly composed of small squamous lined epithelial cyst with prominent granular layer centrally dilated and connected to epidermis and surrounded by cellular fibrous stroma.Numerous secondary & tertiary hair follicles radiates from the central cavity–Caput Medusae appearance. The sebaceous trichofolliculoma is a rare variantand its characterized by small lobules of well differentiated sebaceous glands lacking hair component. The main differential diagnosis includes follicular sebaceous cyst hamartoma, a rare hamartomatous lesion. Microscopy shows prominent epithelial component with folliculosebaceous proliferation like cysts and lack of epidermal connection & prominent stromal component.

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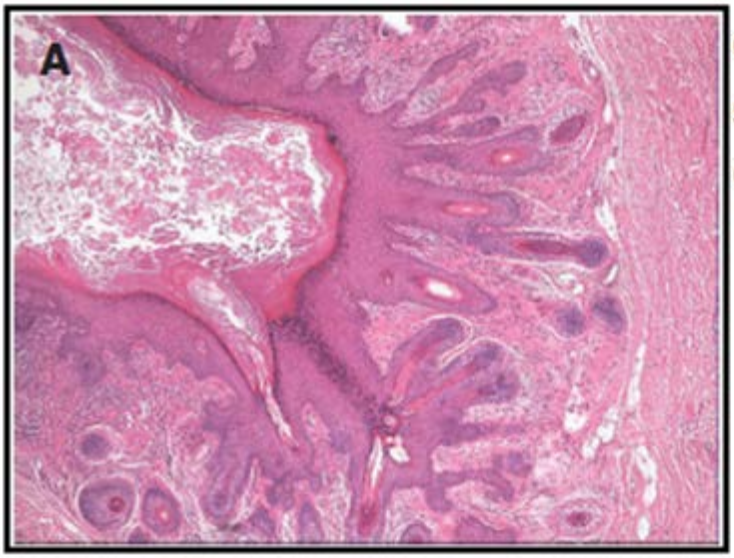
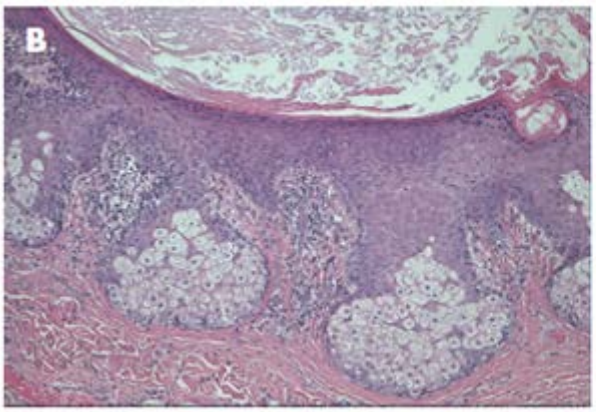


Cyst lined by squamous epithelium



Sebaceous trichofolliculoma

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