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Paediatric cutaneous adnexal tumours: a study of 559 cases

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Summary

Cutaneous adnexal tumours encompass a wide group of lesions with apocrine, eccrine, follicular, sebaceous and mixed differentiation. The large majority are benign and represent sporadic lesions, though malignant forms are occasionally encountered and some cases develop in the setting of inherited tumour syndromes. Accurate histological classification can be difficult as there are numerous histological appearances, many of which are individually uncommon, and complex, overlapping and historically variable nomenclature is typical. The aim of this study was to review and classify the spectrum of cutaneous adnexal tumours seen in patients 18 years of age and under in two major tertiary centres over a 20 year period. A total of 559 cases were included, with 60% occurring in female patients. The large majority (87%) occurred in the head and neck region and were benign. Only one (0.2%) was malignant. The original diagnosis was supported by histological review in 99% of cases of pilomatricoma reviewed, but in only 71% of non-pilomatricoma cases reviewed. The most common lineage was follicular (97%), with pilomatricoma accounting for the large majority of lesions. Predominant glandular/ductal differentiation was seen in 3% of cases, with no tumours showing predominant sebaceous differentiation.

Key words: Skin; adnexal; tumour; cutaneous; appendageal; children; histopathology.

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INTRODUCTION

Cutaneous adnexal tumours are a group of tumours that exhibit morphological features of differentiation towards one or more of the types of appendageal structures present in normal skin. The classification of these tumours is complex, with numerous lesions described, many of which are individually rare. Different neoplasms have been described under a single name (e.g. acrospiroma), and various names have been employed to describe identical lesions (e.g., conventional trichoepithelioma and cribriform trichoblastoma) or different parts of the morphological spectrum of a single lesion (e.g., fibrofolliculoma and trichodiscoma). Additionally, there are complex historical variations in attributing apocrine or eccrine differentiation to many lesions showing predominant glandular/ductal differentiation.[1] Although adnexal tumours as a group are not rare, many of the individual entities are only uncommonly encountered in routine anatomical pathology practice, either in the adult or paediatric population, and there is well recognised interobserver difficulty in the classification of many lesions.[2] While most cutaneous adnexal tumours are benign and complete excision provides a cure, they may be the initial sign of syndromes which confer an increased risk of future tumours and internal malignancies.[3,4] In such a situation, accurate classification is of critical importance.

In the present study we reviewed the entire spectrum of cutaneous appendageal tumours in children received over a 20 year period at two major tertiary referral centres.

MATERIALS AND METHODS

This study was approved by the ethics committee of Princess Margaret Hospital, Perth, WA.

The computerised records of PathWest Laboratory Medicine, QEII Medical Centre, and PathWest Laboratory Medicine, Princess Margaret Hospital, from January 1995 to December 2015 were searched. Histopathology reports of patients 18 years old and under with a diagnosis of a cutaneous adnexal tumour or basal cell carcinoma were retrieved. Cases of naevus sebaceus of Jadassohn (without cutaneous adnexal tumour or basal cell carcinoma) were not retrieved.[1] A total of 582 cases were identified, including 560 cases reported as cutaneous adnexal tumour and 22 cases reported as basal cell carcinoma. The clinical data, including age, location and the original diagnosis were extracted. As pilomatricoma is common in this age group, histological diagnosis was considered likely to be routine and consistent. To validate this, a subset of 140 cases, representing those with slides available at QEII Medical Centre accessioned between August 2010 and October 2015, and those with slides available at Princess Margaret Hospital accessioned between October 2009 and October 2015, were reviewed by a single dermatopathologist. The haematoxylin and eosin stained sections of 72 lesions with a diagnosis other than pilomatricoma were reviewed at a multiheader microscope by two dermatopathologists and a paediatric pathologist, blinded to the demographic details and original diagnosis. Four cases were excluded due to unavailability of material and/or poor preservation, notably including one case diagnosed as sebaceous adenoma for which slides were not available which was included in a previous study.[2] The other three cases excluded were reported as 'trichofolliculoma' (two cases) and 'trichoepithelioma'. An additional four cases were considered on histological review to represent lesions other than cutaneous adnexal tumour: one case representing accessory tragus; one case representing intradermal melanocytic naevus; one case showing normal sebaceous glands; and one case representing primitive neuroectodermal

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tumour (PNET). The last of these had originally been diagnosed as hidradenocarcinoma, but a revised diagnosis of PNET was confirmed by subsequent immunohistochemical and electron microscopic examination. All 22 lesions reported as basal cell carcinoma were confirmed as such on histological review and these cases were excluded from further analysis. This left a final histological study number of 42 non-pilomatricoma adnexal tumours, which were classified using the nomenclature employed in a recent comprehensive monograph on cutaneous adnexal tumours.[5]

RESULTS

A total of 559 lesions met the inclusion criteria for the study. The age at presentation ranged from 2 months to 18 years, with a median age of 9 years; 223 (39.8%) occurred in males and 336 (60.1%) in females (Table 1). The anatomical distribution of the tumours is detailed in Fig. 1 and 2: 63% were on the face, 3% on the ears, 7% on the scalp, 14% on the neck, 9% on the torso and 4% on the legs. The most common (predominant) histological differentiation (97% of cases) was follicular, followed by apocrine/eccrine, with no lesions showing predominant sebaceous differentiation. Typical examples of some of the lesions are illustrated in Fig. 3.

Table 1 Summary of the histological diagnosis (after review), sex distribution and age range of the cutaneous adnexal tumours reviewed in this study

Histological entity	No.	Sex (M:F)	Age, range (median)
Follicular			
Pilomatricoma	517	212:305	5 mo–18 y (8 y)
Trichoblastoma	7	2:5	8–13 y (11 y)
Desmoplasic trichoepithelioma	7	1:6	11–17 y (14 y)
Trichofolliculoma	7	3:4	3–14 y (9 y)
Proliferating tricholemmal tumour	2	1:1	15–16 y
Congenital panfollicular naevus	2	0:2	2–7 mo (5 mo)
Apocrine/eccrine			
Syringocystadenoma papilliferum	6	2:4	10–17 y (13 y)
Syringoma	2	0:2	12–17 y (12 y)
Syringomatous carcinoma	1	0:1	7 y
Hidradenoma	5	2:3	5–17 y (11 y)
Poroma	2	0:2	7–16 y (12 y)
Apocrine hidrocystoma	1	0:1	15 y
Total	559	223:336	2 mo–18 y (9 y)

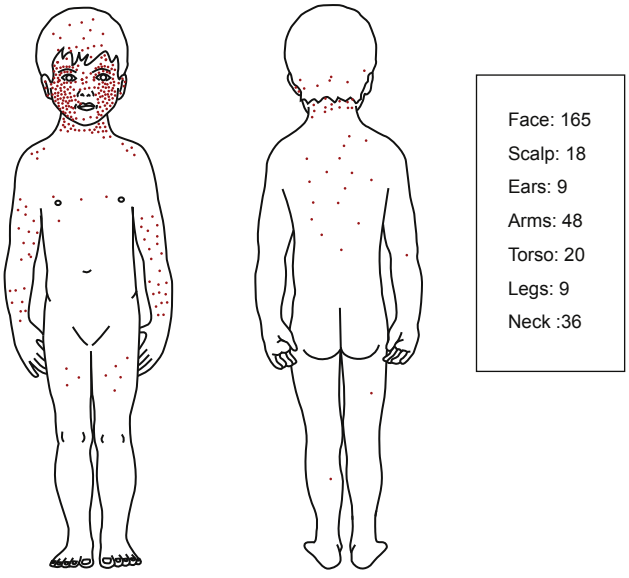

Fig. 1 Pilomatricoma site distribution (one centre only, $n = 286$).

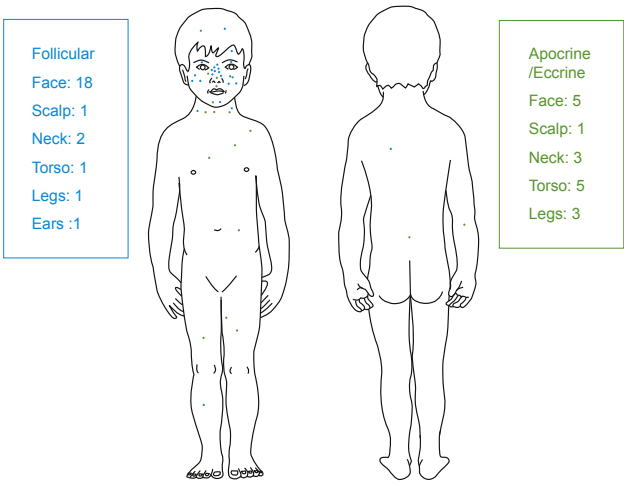

Fig. 2 Site distribution of all confirmed non-pilomatricoma cutaneous adnexal tumours ($n = 42$).

Of the cases of pilomatricoma, the original diagnosis was confirmed in 139 of 140 cases (99.3%) for which slides were available. One case was considered on review to represent an example of hybrid cyst with focal pilomatrical differentiation (Fig. 4).

Of the 42 cases confirmed as cutaneous adnexal tumour other than pilomatricoma on histological review, the review diagnosis was concordant with the original diagnosis in 30 cases (71%). Two cases were originally reported as malignant, both classified as syringomatous carcinoma. On review one of these was favoured to represent a desmoplastic trichoepithelioma with perineural involvement.[6] This lesion occurred on the nose of an 11-year-old boy. The sections showed a poorly defined dermal proliferation of epithelial strands in a desmoplastic stroma, with horn cyst formation and focal calcification (Fig. 5A). Perineural invasion of small nerve twigs was present (Fig. 5B). The original diagnostic report included a discussion of the possibility of desmoplastic trichoepithelioma, but the presence of perineural invasion was felt to speak against that possibility. The occurrence of perineural invasion in desmoplastic trichoepithelioma, now a recognised phenomenon,[6] had not been described in the literature at the time of the initial report. No residual tumour was present on re-excision of this site and the patient was well, without evidence of recurrence, at 7 years follow up. One case reported initially as a syringomatous carcinoma on the cheek in a 7-year-old girl was confirmed on histological review. This lesion consisted of irregular infiltrating ductal structures within a fibroblastic stroma in the dermis, with invasion into the subcutaneous fat (Fig. 6). The primary excision was complete and there was no evidence of recurrence 8 years post-excision.

In 11 cases a different benign cutaneous adnexal tumour classification was favoured on review. This included two cases reported as spiradenoma and one case reported as acrospiroma which were reclassified as cribriform trichoblastoma; two cases of syringoma which were reclassified as desmoplastic trichoepithelioma; two cases reported as