DERMAL CYLINDROMA AND TRICHO-EPITHELIOMA

A Report of Two Cases

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SUMMARY: A case of multiple Dermal Cylindromata of the scalp (turban tumours) is described. The relationship of this inherited tumour to Tricho-epitheliomata is discussed and a case of the latter is also presented.

Introduction

Ancell in 1842 first described the inherited scalp tumours which have subsequently been termed dermal cylindroma by virtue of their histological appearance of cylinders of cells surrounded by a hyaline stroma, situated within the dermis. The condition is transmitted as an autosomal dominant gene and the majority of recorded cases have shown a female preponderance, although this is not the case in all pedigrees. There is also considerable evidence that dermal cylindromata and tricho-epithelioma which have long been noted as occurring together, are manifestations of a single autosomal dominant gene with a variable phenotype. Welch, Wells and Kerr (1968) report two large pedigrees in both England and the United States of America in which tumours typical of both conditions were present.

Tricho-epithelioma was first described by Brooke in 1892 under the title of epithelioma adenoides cysticum. The clinical appearance is distinctive; numerous small, slightly translucent, papulo-nodules develop on the centre face, particularly in the naso-labial folds and peri-orbital regions.

In those cases in which both conditions co-exist, the pattern has usually been of scalp cylindromata, facial tricho-epithelioma and the rarer lesions on trunk or limbs showing either histology. However, individual cases in such dual pedigrees may manifest solely cylindromata or solely tricho-epitheliomata.

Dermal cylindromata appear as firm, flesh coloured, pink or red, hairless tumours usually painless, situated most commonly on the scalp or forehead. They vary in size from a pea to a hen's egg and in the extreme form may be so numerous as to completely replace the scalp as if by a turban—hence the synonym turban tumours. Multiple dermal cylindromata may be less common than solitary ones. Crain and Helwig (1961) report fifty-six patients with the condition, twenty-seven had solitary lesions, fifteen multiple lesions and fourteen did not state whether they had more than one tumour. However, as their series indicates that the lesions may develop very slowly, are as small as two millimetres diameter and present as early as four years or as late as sixty-nine years—it is possible that further lesions may develop and be overlooked or are not reported, Tricho-epithelioma can also occur as a solitary lesion and like solitary cylindroma is stated to be non-hereditary (Lever 1967).

Case Reports

Case 1—Spr L.E. aged 31*

First developed a tumour of the scalp 19 years ago when aged twelve. Similar lesions have continued to erupt on the scalp at intervals since then and all have slowly enlarged.

Tumours on trunk and limbs first noted fifteen years ago when aged sixteen. These also have increased in size and number over the years. All lesions are firm papulo nodules; those on the trunk and limbs are softer than the scalp lesions. They are symptomless and non-tender, purple-pink or flesh coloured; some are covered with telangiectases. The front of the scalp is now virtually replaced by tumour giving a turban appearance.

The scalp tumours vary from 0.5 to two centimetres diameter, and the trunk tumours from 0.2 to one centimetre diameter.

The family history, consistent with an autosomal dominant inheritance, is shown in Fig. 1.

Fig. 1. Family history. Case 1.

(Figs 1 and 5 produced by The Department of Medical Illustration Manchester Royal Infirmary)

The transmission is through the probands mother and maternal grand-father. Two of the probands elder aunts and the eldest uncle were affected but the age at which the tumours first appeared is not known. Only one of the probands seven sibs is known to be affected, the next senior brother having developed scalp tumours within the last year.

Histology

The condition was first diagnosed and confirmed histologically as multiple dermal cylindromata in 1962. Successive biopsy excisions of lesions from scalp (5), abdomen

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Fig. 2. Case 1. Dermal Cylindroma showing scalp and facial tumours.

Fig. 3. Case 1. After excision and graft.

Fig. 4. Case 1. Cylinders of cells within the dermis surrounding and surrounded by a hyaline stroma.

(2), leg (2), forehead (1), and neck (1), have been carried out at various Military hospitals from 1962-1969. All the histological sections irrespective of site have shown a similar histology. The report of 1969 of lesions from the left forehead, left thigh, abdomen and back of neck reads:—

“All four specimens contain sub-epidermal nodules with a similar microscopic appearance. The nodules are composed of sharply circumscribed nests of small uniform

cells some of which surround small hyaline droplets. The cell nests show a peripheral palisading surrounded by a thick hyaline membrane. There is no inflammation and the cellular morphology is uniform throughout. Diagnosis: Dermal Cylindromata."

Treatment

The patient was referred to the Department of Plastic Surgery, Cambridge Military Hospital. The larger portion of the scalp was excised and a split skin graft applied to the defect. At a later date individual lesions without the grafted area were excised and a toupee supplied to cover the bald area of scalp.

Figures 2 and 3 show dermal cylindroma scalp and facial tumours before treatment and after excision and graft. Figure 4 shows cylinders of cells within the dermis surrounding and surrounded by a hyaline stroma.

Case 2. Pte B aged 24

Small tumours in the region of the inner canthi of both eyes were first noted at four years of age and have remained virtually unchanged and symptomless. One year ago similar lesions appeared in both naso-labial folds and have gradually increased in size and number. All the lesions are slightly translucent, papulo-nodules varying in size from a pin head to three millimetres diameter.

The family history, consistent with an autosomal dominant inheritance, is shown in Fig. 5.

Fig. 5. Family history. Case 2.

The transmission is through the probands father and possibly paternal grandfather. A paternal aunt is affected, also her daughter, the probands' cousin. Neither of his sibs are affected.

Histology

The lesion is a circumscribed nodule overlaid by normal epidermis. Numerous small islands of basaloid cells lie within the mid and upper dermis. These islands surround cystic spaces, the majority of the cysts contain keratin and their morphology frequently resemble that of hair papillae. Diagnosis: Tricho-epithelioma.

Treatment

The treatment of this condition is not entirely satisfactory, although the tiny lesions can be destroyed with the electrocautery, recurrences are common. If larger lesions are

tumours differentiating toward epidermal sweat gland appendages—but whether towards eccrine or apocrine glands is still in dispute. Histochemical enzymatic studies have given conflicting or inconclusive results, but electron microscopy appears to favour an apocrine origin (Lever and Hashimoto 1966). The possible common genetic origin of the two tumours is in favour of an apocrine derivation since embryologically hair and apocrine glands are derived from the common primary epithelial germ whereas eccrine glands are derived from a separate sweat gland germ.

Malignant transformation of both tumours is very rare though there is no clear histological distinction between tricho-epithelioma and basal cell epithelioma of keratotic type and occasionally tricho-epithelioma may progress to a more ulcerative and destructive type of basal cell epithelioma (Ziprkowski 1966).

Malignant change in cylindroma of dermal origin has rarely been recorded (Lyon and Rouillard 1961), however in this context the prefix “dermal” is essential to avoid confusion with a group of tumours of the head and neck also designated as “cylindromas” which are adeno-cystic carcinomas arising from mucous and salivary glands of the upper respiratory and digestive tracts (Baxley and Farmer 1972). These tumours, by contrast, are highly malignant and slowly but inexorably fatal.

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REFERENCES

- ANCELL, H. (1842). *Med-chir. Trans.* **25**, 227.
 BAXLEY, W. W. and FARMER, J. C. (1972). *S. Afr. med. J.* **65**, 685.
 BROOKE, H. G. (1892). *Brit. J. Derm.* **4**, 269.
 CRAIN, R. C. and HELWIG, E. B. (1961). *Amer. J. clin. Path.* **35**, 504.
 LEVER, W. F. (1967). In: *Histopathology of the Skin*. 4th ed. Pp. 554 and 563. Pitman. London.
 LEVER, W. F. and HASHIMOTO, K. (1966). *Hautarzt* **17**, 161.
 LYON, J. B. and ROUILLARD, L. M. (1961). *Trans. St. John's Hosp. derm. Soc. (Lond.)*, **46**, 74.
 WELCH, J. P., WELLS, R. S. and KERR, C. B. (1968). *J. med. Genet.* **5**, 29.
 ZIPROWSKI, L. and SCHWEWACK-MILLET. (1966). *M. Dermatologica* **132**, 248.