Proliferating Tricholemmal Cysts

A Case Report

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SUMMARY

Attention is focused on an uncommon, but well-defined clinical and histological entity, which is still often misdiagnosed as a squamous carcinoma. The case described in this article supports the concept that the origin of the cyst is from the outer hair sheath. The importance of some form of irritation which stimulates proliferation of the epithelium is also mentioned.

S. Afr. med. J., **54**, 833 (1978).

Attention was first focused on proliferating tricholemmal cysts by Jones[1] who described a series of 9 cysts under the name of proliferating epidermoid cysts. According to this author epidermoid cysts included keratinous and sebaceous cysts. Until that time these tumours had been known by various other names, and some published cases of a transformation from a benign to a malignant skin cyst were undoubtedly lesions of this kind.[1,2]

In the same year, Reed and Lomar[3] described 14 cases. They stressed the features indicating a pilosebaceous origin. They also drew attention to histological criteria suggesting that these lesions could have a malignant potential despite a benign clinical course. They named these tumours invasive pilomatrixomas. In 1971, Dabska[2] described 12 cases of this uncommon tumour with locally invasive, but otherwise benign characteristics, and called them giant hair matrix tumours.

More recently, in a review of tumours of the hair follicle, Headington[4] mentioned that these tumours show various histological features which are, however, distinctive enough to confirm that the lesions are proliferating tricholemmal cysts on clinical and histological grounds.

We wish to describe an additional case and give support to the concept that trauma may be instrumental in stimulating proliferation of the epithelium.

CASE REPORT

A 66-year-old Black woman presented with a tumour of 30 years' duration in the left occipital region. The tumour had gradually increased in size. Two years previously, a diagnosis of squamous carcinoma had been made from a biopsy specimen. At that stage, the patient had refused further treatment.

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Date received: 4 July 1978.

On examination the tumour was 4,5 cm in diameter, slightly lobulated, and situated subcutaneously with an ulcer 1,5 cm in diameter in the overlying skin. Under the previous biopsy site was a nodule, 1,75 cm in diameter, attached to the original tumour. There were no enlarged lymph glands and the patient appeared perfectly healthy. The tumour was surgically removed with a small rim of surrounding skin and subcutaneous fat (Fig. 1).

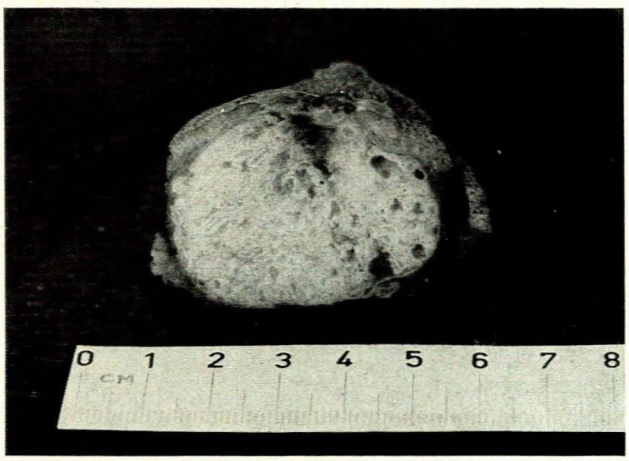

Fig. 1. Cut surface of tumour. Note convoluted appearance.

Pathological Findings

Macroscopically the tumour was well circumscribed and appeared partially encapsulated. The cut surface showed numerous thin membranous whorls. On one side was a nodule, 1,5 cm in diameter, which appeared more solid, with no visible convolutions.

On histological examination the tumour consisted of an intertwining network of stratified squamous epithelium supported by fibrous connective tissue. The periphery of the tumour was sharply demarcated from the surrounding tissue by a layer of stratified squamous epithelium broken in areas by fibrous tissue invaginations (Fig. 2). This peripheral layer was in continuity with the overlying skin. The epithelium itself showed basaloid cells with a tendency towards palisading on a thick hyaline basement membrane. The basaloid cells gradually matured towards mature squamous cells with well-defined intercellular bridges. Occasional cells had atypical nuclei and there were sparse mitotic figures. A small number of cells showed individual keratinization and there were numerous poorly-formed keratin pearls. Well-formed squamous eddies were not a feature. Towards the surface, the epithelium showed formation of abundant non-layered keratinous material

without an underlying granular layer. There was also extensive necrosis of the epithelial layer resulting in the formation of cyst-like spaces. Some of the epithelial cells had clear, foamy cytoplasm which stained strongly with the PAS method. Prior digestion with diastase gave negative staining.

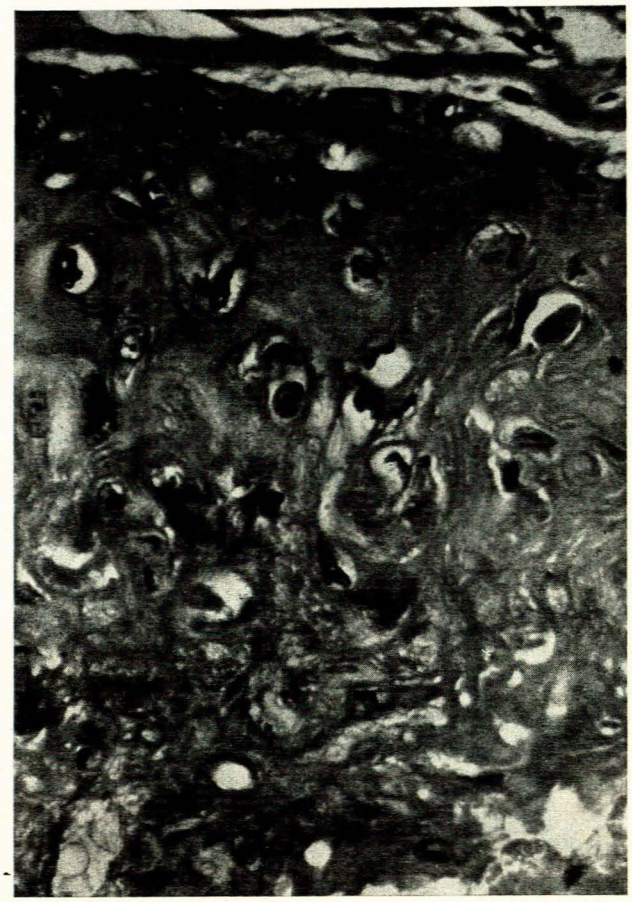

Fig. 2. Stratified squamous epithelium with basaloid cells resting on a prominent basement membrane. Intercellular bridges and nuclear atypia are evident (H and E × 400).

The fibrous stroma surrounded all the strands of epithelium and was well vascularized. Numerous foci of multinucleated giant cells could be seen removing the keratin material. In some areas the stroma was oedematous and in others it had a glassy hyaline appearance. One small island of ghost cells and a few small calcifications could be seen.

The nodule of tumour tissue underlying the biopsy scar had a different appearance. It seemed to protrude from the margin of the main tumour and to grow with a pushing edge. There was a gradual transition in morphology from the original tumour to discrete islands of medium-sized squamous cells with intercellular bridges and individual cell keratinization. The cell nuclei were more atypical and mitoses were more frequent, at an average of 1 per high-power field. There was no layer of basaloid cells and no keratin accumulations. A number of epithelial islands showed central necrosis. The periphery was well-defined, but there was no limiting layer of epithelium. Many of the cells contained diastase-labile PAS-positive material.

In summary, this part of the tumour appeared more active, less mature, and the tumour cells more closely resembled cells from the transitional area of the upper outer hair sheath.

DISCUSSION

Proliferating tricholemmal cysts are an established but rare entity. Their importance lies in the frequency with which they are misdiagnosed both clinically and histologically as squamous carcinomas.[1-3] Typically they are found in the occipital area of elderly Black women. However, they can also occur in males, non-Negroid races and on other regions of the body. The long duration and slow growth of these tumours are also characteristic. The tumour may reach a large size and the overlying skin is likely to become ulcerated. Histologically, cellular atypia and occasional mitoses may cause concern, but current knowledge points to a benign course with rare local recurrences following excision. Only one case of metastasis to a local lymph gland has been described.[4]

There are a number of histological features which indicate a probable origin from the outer hair sheath, for example the prominent thick basement membrane, the diastase-labile strongly PAS-positive vacuolated cells similar to those described in the infundibular portion of the sheath[5] and the peripheral palisading of basaloid cells. Furthermore, the transitional area of the outer hair sheath normally has cuboidal cells with intercellular bridges and desmosomes and shows partial keratinization.[6] The cells of the new tumour nodule reflect some similarity to these cells.

It is generally agreed that the proliferation of the epithelium of a pre-existing hair sheath cyst is caused by some form of irritation, e.g. trauma, infection of chemical irritation by the contents of the cyst. This concept is strengthened by the finding in our patient of the less mature and more active appearing portion of the tumour under the site of the surgical incision done 2 years previously.

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