

Metastasising pilar tumour of scalp

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SUMMARY A case of pilar tumour of the scalp, treated by local excision and radiotherapy, later metastasised to the neck. The variable histological growth patterns of the primary tumour and its metastases are described. It is concluded that the pilar tumour is a genuine neoplasm of the hair follicle that is occasionally capable of malignant behaviour.

Mann *et al* described two cases of pilar tumours showing histological evidence of invasion.¹ Both these cases showed a non-invasive and an invasive component, but neither had metastasised at the time of reporting. Follow up on one of the patients, case 3 in the report of Mann *et al*, is now available¹: Metastases of the primary scalp tumour were excised from the posterior triangle of the neck. Paradoxically, the metastases displayed a growth pattern similar to that interpreted as a non-invasive component in the primary tumour.

Controversy reigns as to whether the pilar tumour represents a hyperplastic or a neoplastic process; this report shows that the lesion is capable of metastasising, albeit only to local sites. With the possible exception of the metastasising tumour of the scalp recorded in 1916,² visceral metastases of pilar tumour have not been observed.

Case history

The patient, an English woman born in 1906, had had a cyst underlying the skin of the occipital scalp for many years. For two years before its excision the lesion had slowly increased in size and had become inflamed. When the lesion was removed, together with surrounding skin and subcutaneous tissue in November 1977 it measured 1.8 × 1.3 cm and had ulcerated the overlying epidermis. No regional lymphadenopathy was detected at that time. Local surgery was followed by a course of radiotherapy to the area.

In July 1981 a nodule, which had been present for three months and which measured 1.5 cm in diameter, was excised from the subcutis of the left posterior triangle of the neck, at least 12 cm from the site of the original scalp lesion. In January 1984 three further recurrent nodules, the largest 2 cm in diameter, were excised from the same area of the neck. The patient

remained well and free of recurrence two years after this third operation.

Pathology

The lesion excised from the occipital scalp in 1977 extended into subcutaneous tissue and showed three distinct histological patterns:

1 A small proportion of the tumour resembled a typical benign pilar tumour and was composed of interlacing nodules of smaller peripheral cells that were palisaded and matured into larger central cells with abrupt central keratinisation (Fig. 1). There was slight nuclear atypia and a mitotic rate of 1/10 high power fields.

2 Most of the tumour was formed by small cells with a high nuclear:cytoplasmic ratio arranged in confluent nodules, many undergoing central keratinisation. There was loss of polarity and maturation of the cells and no peripheral palisading (Figs. 1 and 2). The nuclei were hyperchromatic and pleomorphic, and many multinucleated tumour giant cells were present. Numerous apoptotic cells were identified, and mitoses, including abnormal forms, ranged up to 35/10 high power fields. The cytoplasm of many tumour cells contained abundant glycogen.

3 Superficial and ulcerated areas of the tumour were composed of sheets and strands of cells exhibiting severe nuclear pleomorphism with the occasional multinucleated tumour giant cell (Fig. 3). Areas of tumour necrosis were present, but keratinisation was not identified. Mitoses, including abnormal forms, numbered up to 20/10 high power fields.

Occasional areas of transition between the first and second growth patterns, as well as between the second and third patterns were apparent (Fig. 2). The stroma of the tumour was formed by fibrous and hyalinised connective tissue with a lymphocytic infiltrate and areas of cholesterol clefts.

Both metastases to the neck, excised four and seven



Fig. 1 Nodule of malignant pilar tumour at centre of field is surrounded by interlacing strands of typical benign pilar tumour. (Haematoxylin and eosin.) $\times 60$.

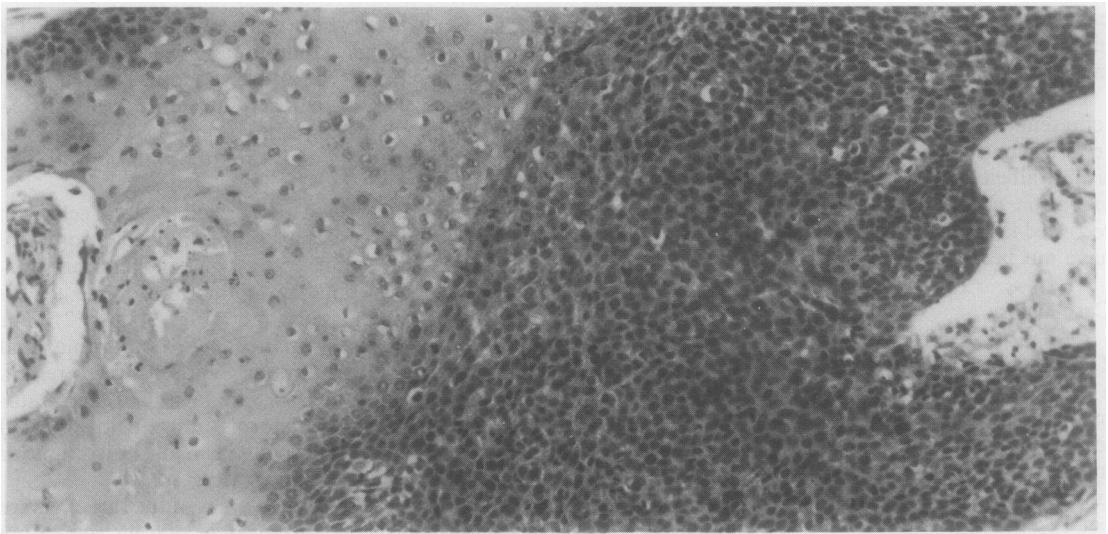


Fig. 2 Transitional zone between benign pilar tumour with squamous differentiation (left) and malignant pilar tumour with nuclear hyperchromasia and crowding (right). (Haematoxylin and eosin.) $\times 160$.

years after the primary tumour, displayed a growth pattern very similar in appearance to pattern 2 of the primary lesion (Fig. 4). The nodules of tumour contained central keratin and the malignant cells contained diffuse cytoplasmic glycogen. There was no evidence that either metastasis was present in a lymph node; both were situated in fibromuscular tissue. Tumour was identified around nerve trunks in the more recent metastasis.

Discussion

Keratinous cysts of the skin are divided into two groups—pilar and epidermoid—on the basis of their different modes of keratinisation.³⁻⁵ Pilar cysts, derived from the external root sheath of the hair follicle and occurring most commonly on the scalp, are characterised by trichilemmal keratinisation. This occurs in the external root sheath wherever it is no

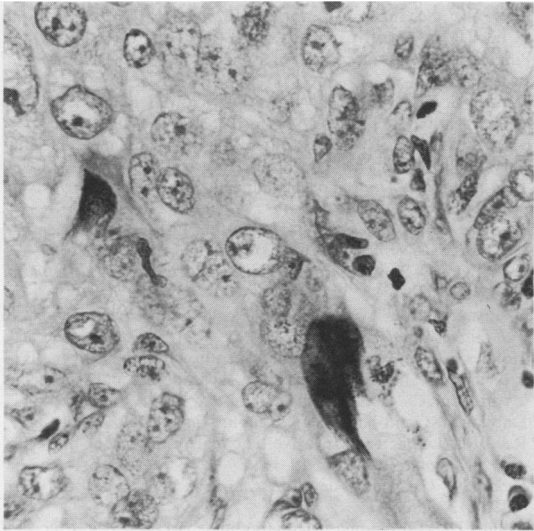


Fig. 3 Undifferentiated pilar tumour showing marked nuclear pleomorphism. (Haematoxylin and eosin) $\times 400$.

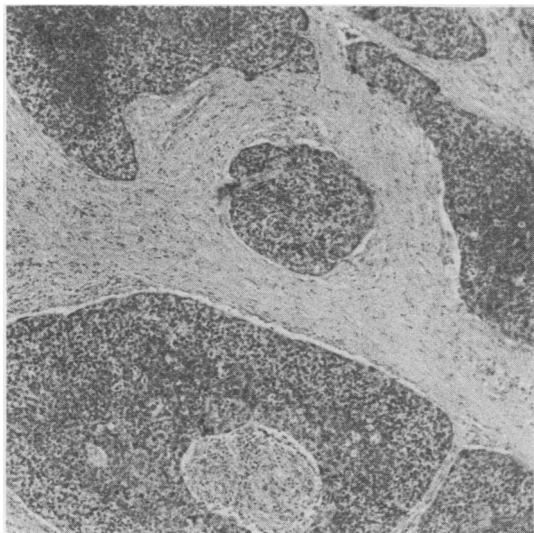


Fig. 4 Metastatic pilar tumour similar in appearance to malignant areas of pilar tumour in Fig. 1. (Haematoxylin and eosin) $\times 50$.

longer covered by the internal root sheath and differs from epidermal keratinisation by increase in size of the keratinocytes, with loss of their nuclei and abrupt transition into dense keratin without an intermediate keratohyaline layer.⁶⁻⁸

Small foci of intraluminal budding are observed

quite commonly in the wall of pilar cysts. Sometimes proliferating epithelium replaces most if not all of the pre-existing wall of the cysts.⁹ The stimulus responsible for this process and its biological nature have been disputed. Factors that may induce proliferation are thought to include trauma,¹⁰ infection, and irritation from the contents of the cyst.^{3 11 12} For this reason the pilar tumour is usually regarded as a pseudoepitheliomatous hyperplasia rather than as a neoplasm. Mann *et al*¹ argued that it represented a genuine neoplasm.

The histological features of a pilar tumour are distinctive. The lesion is composed of interlacing nodules of squamous epithelium that are sharply separated from the surrounding tissue and undergo abrupt trichilemmal keratinisation into amorphous and focally calcified keratin in their centre.¹ The usual lack of cytological atypia and stromal invasion distinguish the tumour from a squamous cell carcinoma⁹ for which it is often mistaken clinically.¹⁰ Many pilar tumours contain epithelial pearls and isolated dyskeratotic cells, findings that may arouse suspicion of an erroneous diagnosis of malignancy.⁶ Areas of epidermal keratinisation, forming laminated keratin with an intermediate keratohyaline layer, suggest differentiation of the tumour towards epithelium of the follicular infundibulum. Other areas of the tumour may resemble the lower part of the follicular outer root sheath, forming glycogenated clear cells, squamous eddies, a palisaded arrangement of basal cells at the periphery of nodules, and an investing thick hyalinised zone of collagen.⁹

It is accepted that pilar tumours, which show no cytological atypia, are capable of eroding bone and of recurring after inadequate excision.^{1 12-15} Examination of the earlier reports describing cases of squamous cell carcinoma arising in so called "sebaceous cysts" reported before Wilson-Jones's recognition of the proliferating pilar cyst as a separate entity,¹² shows a poorly documented assortment of cystic lesions.¹⁶ Many of these lesions may have been pilar tumours, as judged by their benign course, including rare occurrence of metastases.¹⁷⁻¹⁹

More recent descriptions of malignant change in pilar tumours conform to two patterns—namely, circumscribed nodules of cytologically malignant squamous epithelium, suggesting dedifferentiation within nodules of the pilar tumour (growth pattern 2 in our case), and diffuse spindle cell growth of malignant cells adjacent to tumour nodules and infiltrating surrounding stroma (growth pattern 3 in our case).^{1 13 14 16 20} Regional metastases from pilar tumours have been documented pathologically in three patients to date, including the present case.^{14 21} Metastases of spindle cell squamous carcinomas arising in other organs of the body may contain only the

squamoid component of the tumour.^{20 22} An analogy may be made between the behaviour of these tumours and malignant pilar tumours of the scalp, in that the reported metastases of the latter have all seemed to reproduce the histological appearance of the nodular primary lesion, while metastases resembling the apparently less differentiated infiltrative component have not so far been described.

The case reported here documents unequivocal regional metastases from a clearly malignant trichilemmal tumour. The possibility of such malignant behaviour has not met with general acceptance. Therefore, the much more common benign analogue should be regarded as a trichilemmoma, and there seems little point in retaining the term "proliferating trichilemmal cyst" for this entity. Distant metastasis or death attributable to these tumours have not yet been confirmed.

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