Mixed appendageal tumour: case report

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Summary—A case of a mixed dermal appendage tumour is described. The relevant literature is reviewed and problems with the histological diagnosis discussed.

The dermal cylindroma is an uncommon tumour of sweat gland origin with a characteristic histological appearance. A close association exists between this and another appendage tumour, the trichoepithelioma (Crain and Helwig, 1961). When this association manifests itself in patients with multiple lesions, those of the scalp are purely cylindromata whilst those elsewhere are partly cylindromata and partly trichoepitheliomata. Both tumours are associated with an autosomal pattern of dominant inheritance and it is possible that they are a single genetic entity (Welch et al., 1968).

This short paper describes a patient with multiple tumours who presented in an unusual manner. A problem in establishing the histological diagnosis is also illustrated.

A 74-year-old lady was admitted with profuse bleeding from an infected tumorous mass on her scalp. The bleeding had begun on the previous day. She gave a history of having a smaller bleed the year before which had settled spontaneously. She stated that the tumours had been present for many years, that there was no family history of similar
tumours and that she had not previously sought medical advice regarding the lesions. Examination revealed multiple raised tumours of the scalp some of which were infected and discharging purulent matter. Tumours were also present over both breasts, the axillae and the trunk. Some were pedunculated and one large lesion in the left flank was fungating.

Patients with multiple cylindromata usually present with one or more of the following symptoms or signs: pain, odour, hearing loss, cosmetic defects or symptoms of anaemia associated with chronic blood loss. Profuse haemorrhage has not been reported before in association with this condition in its benign form. A case of fatal haemorrhage has been reported in a cylindroma that had undergone malignant change (Zontschew, 1961). Malignant degeneration in a cylindroma is extremely rare. Seven cases have been reported, six in patients with multiple lesions (Lever and Schaumbarg-Lever, 1975) and one in a patient with a solitary lesion (Bourland et al., 1979).

The cylindroma has a typical histological appearance with irregularly shaped islands of epithelial cells surrounded by hyaline sheaths like cylinders. The islands contain two types of cells. Around the periphery are small cells with dark staining nuclei whilst centrally are larger cells with light staining nuclei. The trichoepithelioma does not have such a characteristic appearance. A histological spectrum exists with no sharp boundary between trichoepithelioma and trichofolliculoma on one hand and basal cell carcinoma on the other. It is also possible to find basal cell carcinomata with features of appendageal differentiation. This spectrum can make the histological diagnosis of a trichoepithelioma difficult. In view of this difficulty and the possible association of cylindroma and trichoepithelioma within the same tumour, care must be taken when interpreting histological sections from multiple skin tumours.

The patient was clinically in hypovolaemic shock and required a blood transfusion. She was treated with topical cleansing, pressure dressings, systemic antibiotic therapy and referred to the regional Plastic Surgery unit. A large portion of the tumour-bearing scalp was removed and replaced with a split skin graft. The fungating lesion in the left flank was excised. Histological examination of the scalp lesion was reported as a dermal cylindroma and the initial report of the lesion in the left flank was of an incompletely excised basal cell carcinoma. However, subsequent review of the histology suggested that despite the presence of ulceration and inflammation the flank lesion was benign, though not a typical cylindroma, having tumour foci with features suggestive of a trichoepithelioma.

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References


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