PILOMATRIX CARCINOMA

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A rare case of aggressive pilomatricoma (Malignant pilomatricoma) in a 20 years old male is presented. Despite its propensity for recurrence it has a good prognosis since it is amenable for treatment with wide excision followed by radio therapy.

Key Words : Malignant Pilomatricoma, Pilomatrix carcinoma, Skin tumours

Introduction

Pilomatricoma is a benign tumour arising from the primitive epidermal germ cells differentiating toward hair matrix, hair cortex and inner root sheath.[1] Clinically the tumour produces a subcutaneous nodule 3 to 30 mm in diameter. Malignant change in a pilomatricoma is extremely rare. Some cases show invasive growth on recurrence and others are malignant from the begining.[2] Pulmonary metastases has been reported in 2 cases.[2] In the case reported by Mir et al[3] widespread metastases involving the heart, liver and kidney has been described.

Case Report

A 20-year-old male presented in March 1989 with a skin tumour in the left lumber region of 3 months duration. The tumour was excised.

Grossly it consisted of an irregular brown mass 5x4x2 cms with a cystic gray-brown tumour tissue on the cut surface. Microscopic examination showed a circumscribed pilomatricoma consisting of lobules of basaloid cells with abrupt keratinisation in the centre. Shadow cells, foreign body giant cell reaction and focal calcification were also present. Postoperative period was uneventful. In October 1989 the patient came back with a recurrent tumour at the site of previous operation. Excision of the tumour revealed features of infiltration of the skin above and the deeper tissues below by a nodular necrotic gray white tumour 4x3x3 cms in size (Fig.1). Histologically the tumour showed centrally necrotic highly cellular lobules of basaloid cells with vesicular nuclei and abnormal mitoses. Squamous differentiation was present (Fig.2). Infiltration of the underlying adipose tissue and skeletal muscle was evident (Fig.3). The features were those of pilomatrix carcinoma.

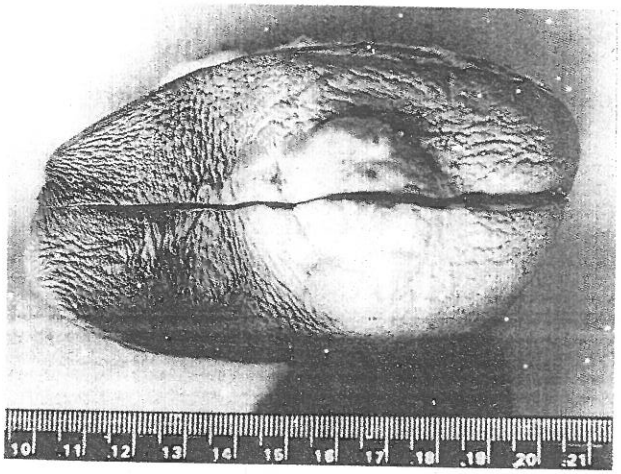

Fig. 1. Gross specimen of the recurrent tumour

In March 1990 the patient suddenly developed haemoptysis and dyspnoea and was admitted to a local hospital where he died. No postmortem examination was done.

Comments

The ultrastructure and histochemical characteristics of these tumour favour their origin from hair matrix cells.[1] The tumour usually appears as a slow growing deep dermal or subcutaneous nodule commonly located in

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the head, neck or upper extremity. Lopansri and Mihm[4] reviewed 5 previously reported cases and adding their own case, identified a group of pilomatricoma with microscopic

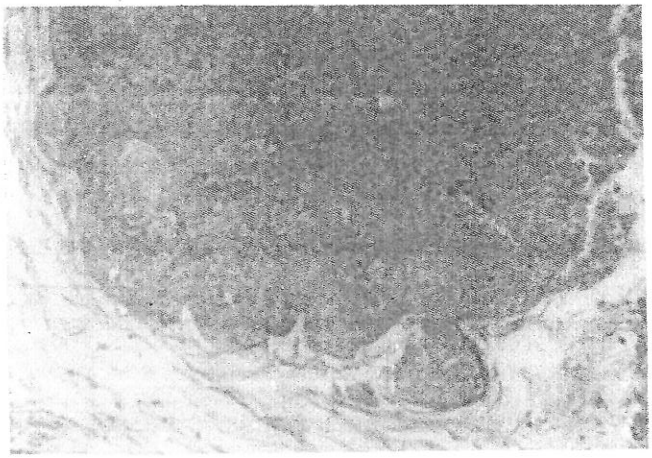

Fig. 2. Highly cellular lobule of basaloid cells with infiltrating margins, squamous differentiation seen (H & E x 200)

aggressive behaviour as indicated by active proliferation of basaloid cells with anaplasia and numerous mitoses and by infiltration of underlying structures. The term pilomatrix carcinoma or calcifying epitheliocarcinoma of

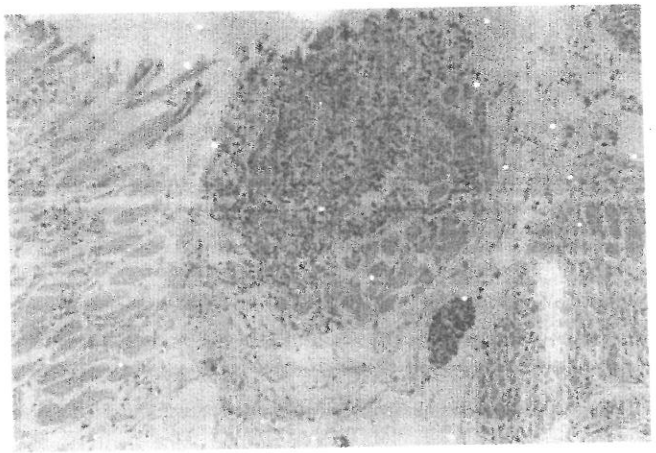

Fig. 3. Tumour infiltrating skeletal muscle (H & E x 100)

Malherbe was suggested by them. Large cystic centres containing necrotic debris is a feature described in pilomatrix carcinoma[2] and was seen in the present case. Transformation of basaloid cells into eosinophilic shadow cells similar to benign pilomatricoma and squamous differentiation have been observed in pilomatrix carcinoma.[1,2] Though size of the tumour is not related to aggressive behaviour, Sasaki et al[4] encountered a pilomatricoma of large size in the right preauricular area which showed aggressive infiltrating behaviour and recurrence.

Distant metastases was not reported although vascular invasion was noted in the cases reported by Gromiko and Prandetsky et al.[4] Further 5 cases have been recorded[1,2] with pulmonary metastases in 2 cases.[2,3] In the present case evidences supporting malignancy includes recurrence of the tumour and histological features of proliferation of basaloid cells, abnormal mitotic activity and invasion of underlying skeletal muscle. The patient died within 5 months of the recurrence with unexplained haemoptysis and dyspnoea (? pulmonary metastases).

The treatment of these patients includes wide excision followed by radiotherapy. The infiltrative pattern of the tumour, an indicator of possible recurrence, is to be carefully evaluated in every case.

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