

Pilomatrixoma and Pilomatrix Carcinoma deceptively Similar but Distinct Entities

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Introduction

Pilomatrix carcinoma is a skin tumour about which there is a relative paucity of material in the plastic surgical literature. In the course of their practice most plastic surgeons will have come across the benign skin lesion designated "Calcifying Epithelioma of Malherbe" named thus in recognition of Malherbe's description of the entity in 1880, even though erroneously ascribed the tissue of origin to be the sebaceous gland¹. Forbis and Helwig in 1961 reviewed 228 such tumours, demonstrated the tissue of origin to be the hair matrix and proposed the term "pilomatrixoma" as more representative of the lesion². These are rare benign skin neoplasms with an incidence rate that varies from 1 in 924 dermatopathologic specimens to 1 in 2200 surgical pathologic specimens. They are slow growing, typically stony hard dermal nodules, usually less than 3 cm. in size and are most commonly found on the head and neck. They tend to occur in children and teenagers, with a slight female preponderance Taffe et al³ and more recently Kaddu et al⁴ have however documented a second peak of onset in the middle aged and elderly. Radiological examination classically shows a solitary, sharply demarcated subcutaneous tumour with extensive sandlike or dense calcification⁵ Histologically they are composed of nests of basaloid cells which undergo abrupt keratinization forming ghost or shadow cells in a background of inflammatory cells and calcification⁶. The recommended treatment is excision which is usually a simple matter as the lesion is well circumscribed. Recurrence is rare² but has been documented. Though the term 'giant calcifying epithelioma' had been coined⁷ to denote a tumour that behaved aggressively and recurred, it was not until 1980 when Lopansri and Mihren first used the term "pilomatrix carcinoma" to describe a recurrent lesion with certain distinctive histological features⁸. Since then there have been occasional reports in the literature but it remains a rare tumour with less than 30 cases reported till 1994⁴.

Case Report

A 32 year old male presented to the out-patients with a two and a half year history of a lesion on the right side of the back (Figures 1 and 2)

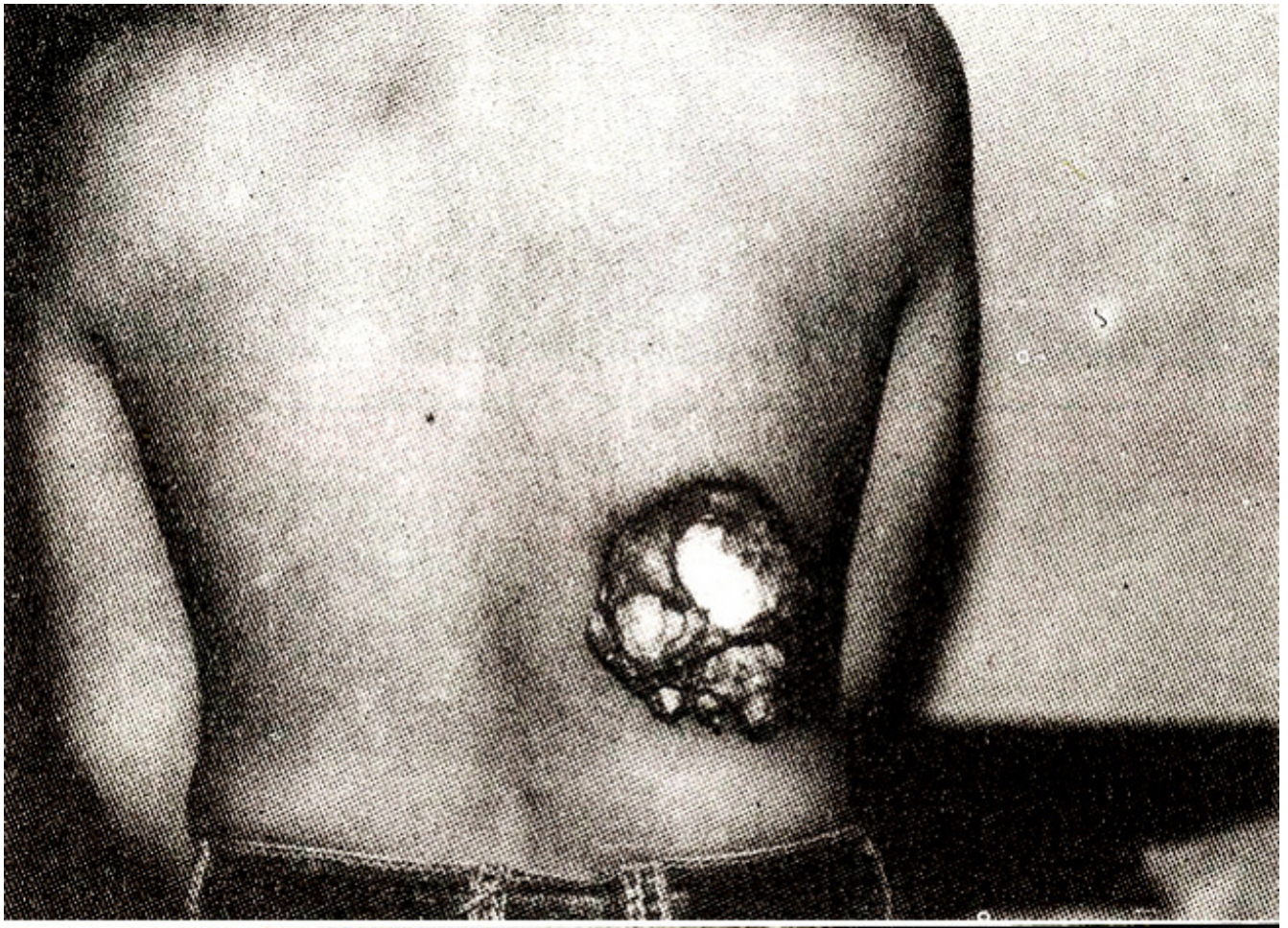


Figure 1 Pre-operative view of the lesion

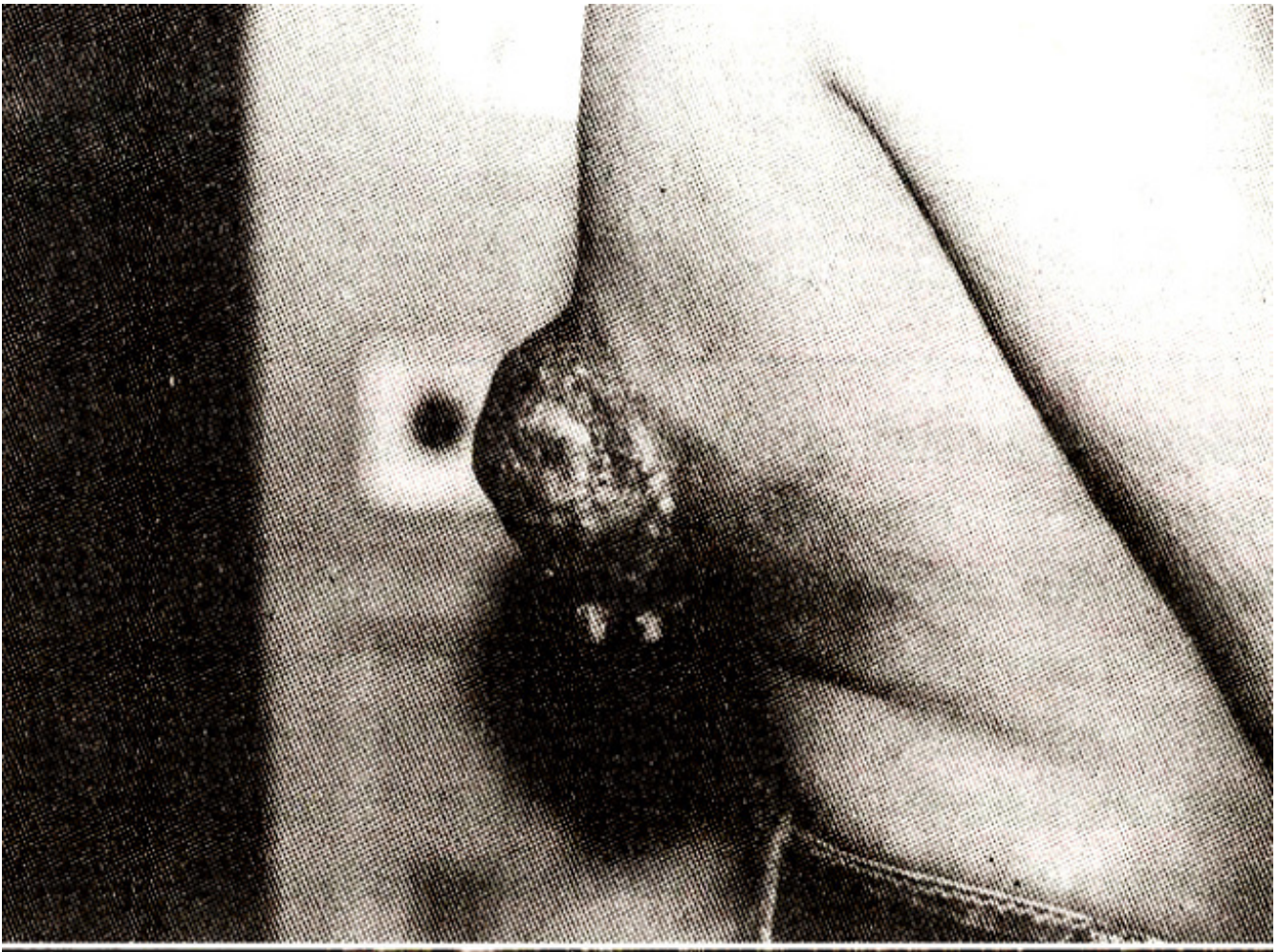


Figure 2. Pre-operative view of the lesion.

which had been increasing steadily in size, more rapidly over the last three months prior to presentation. The patient had initially sought homeopathic treatment to no obvious benefit. On examination the lesion was a 10 cm. fungating, ulcerated mass present below the right scapula and mobile over the underlying chest wall. There was no palpable lymphadenopathy and the chest radiograph was unremarkable. The clinical differential diagnosis included a squamous or basal cell carcinoma or a soft tissue sarcoma. As the incisional biopsy was inconclusive a decision to proceed to definitive surgery was taken. Under prophylactic antibiotic cover the lesion was excised with a 2 cm. margin at a level deep to the latissimus dorsi muscle. All margins appeared well clear of tumour macroscopically which was confirmed on microscopic examination of frozen sections of the resected specimen. The resultant 12 cm. roughly circular, excision site constituted a substantial defect as the patient was of small height and build. A Slide-Swing skin flap Type I9 was designed (Figure 3)

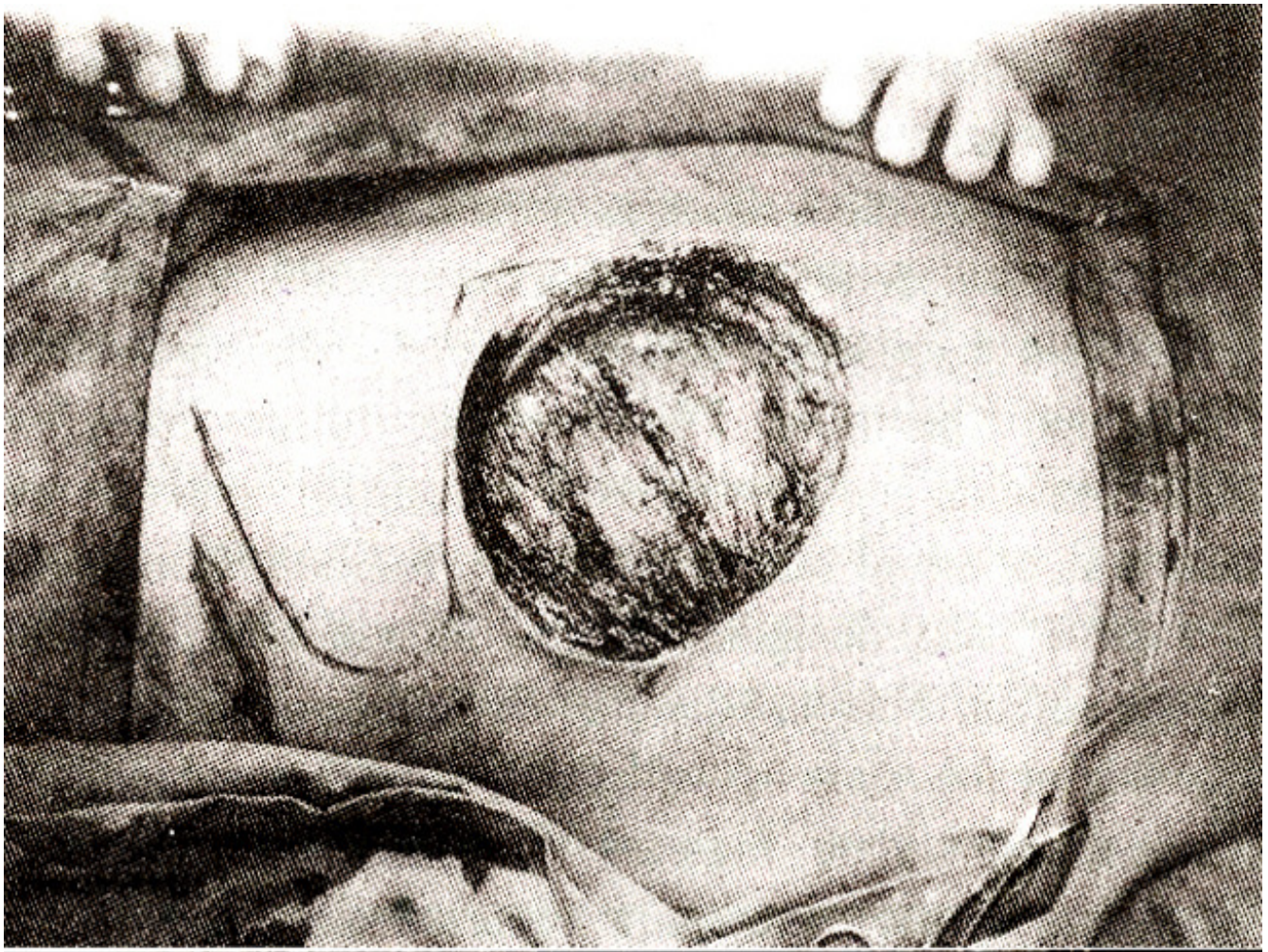


Figure 3. The defect following excision and the slide-swing flap type I marked out, and transposed to close the defect. This had the dual advantage of a reduction in the size of the scars as well as closure of the secondary defect at the same time (Figure 4).

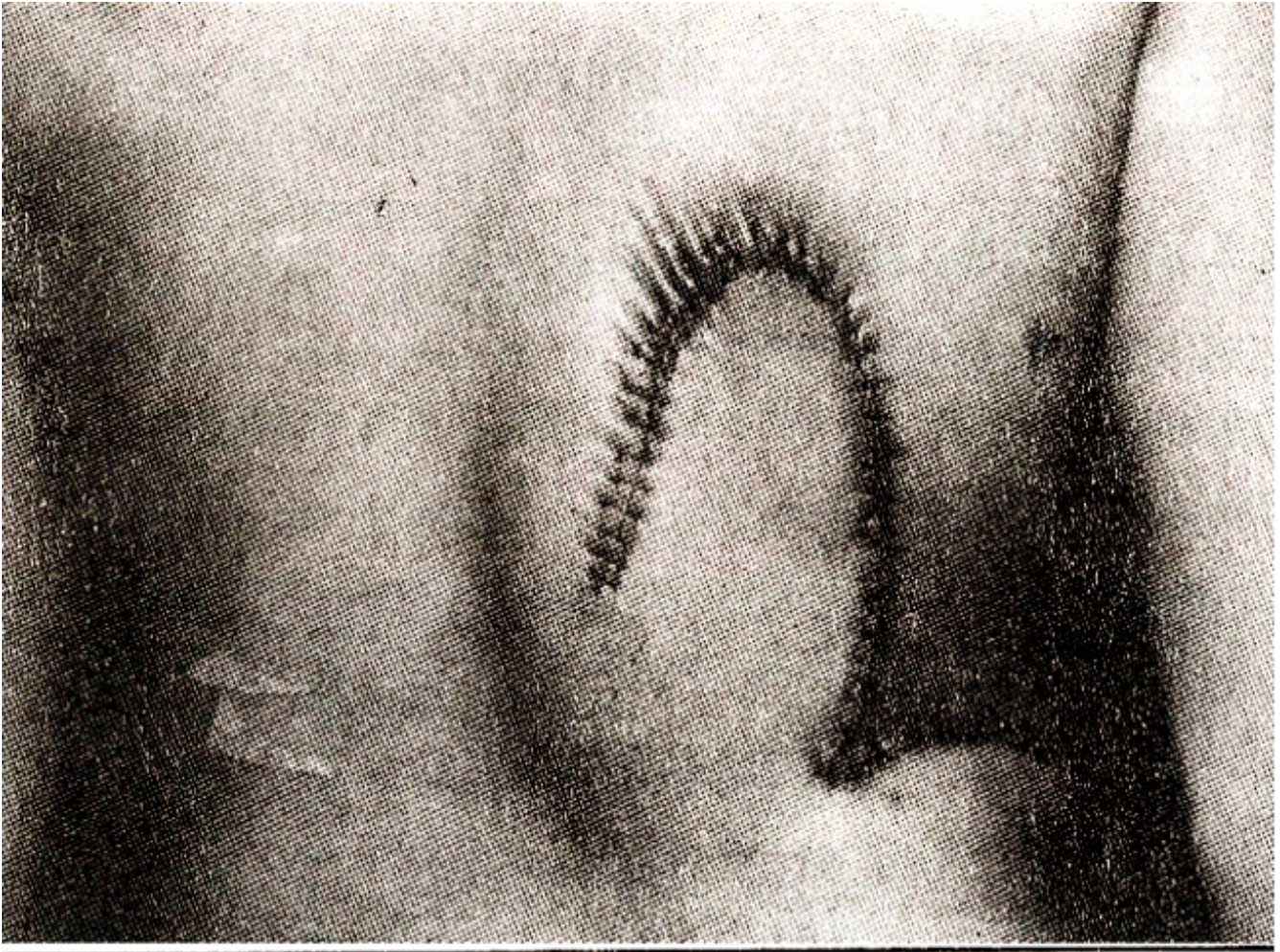


Figure 4. Closure of the defect.

Histological examination of paraffin sections originally reported the lesion as a pilomatrixoma. This was later revised to a pilomatrix carcinoma due to the identification of an invasive neoplastic component of hyperchromatic basophilic cells. As the margin of clearance was 2 cm no further treatment was considered necessary at this time aside from a three monthly out-patient clinic follow-up. The wound initially developed a slight serous discharge at one point, which responded to local treatment and continued antibiotics. Eighteen months post surgery the patient remains well with the wound healed and no evidence of any recurrence (Figure 5).

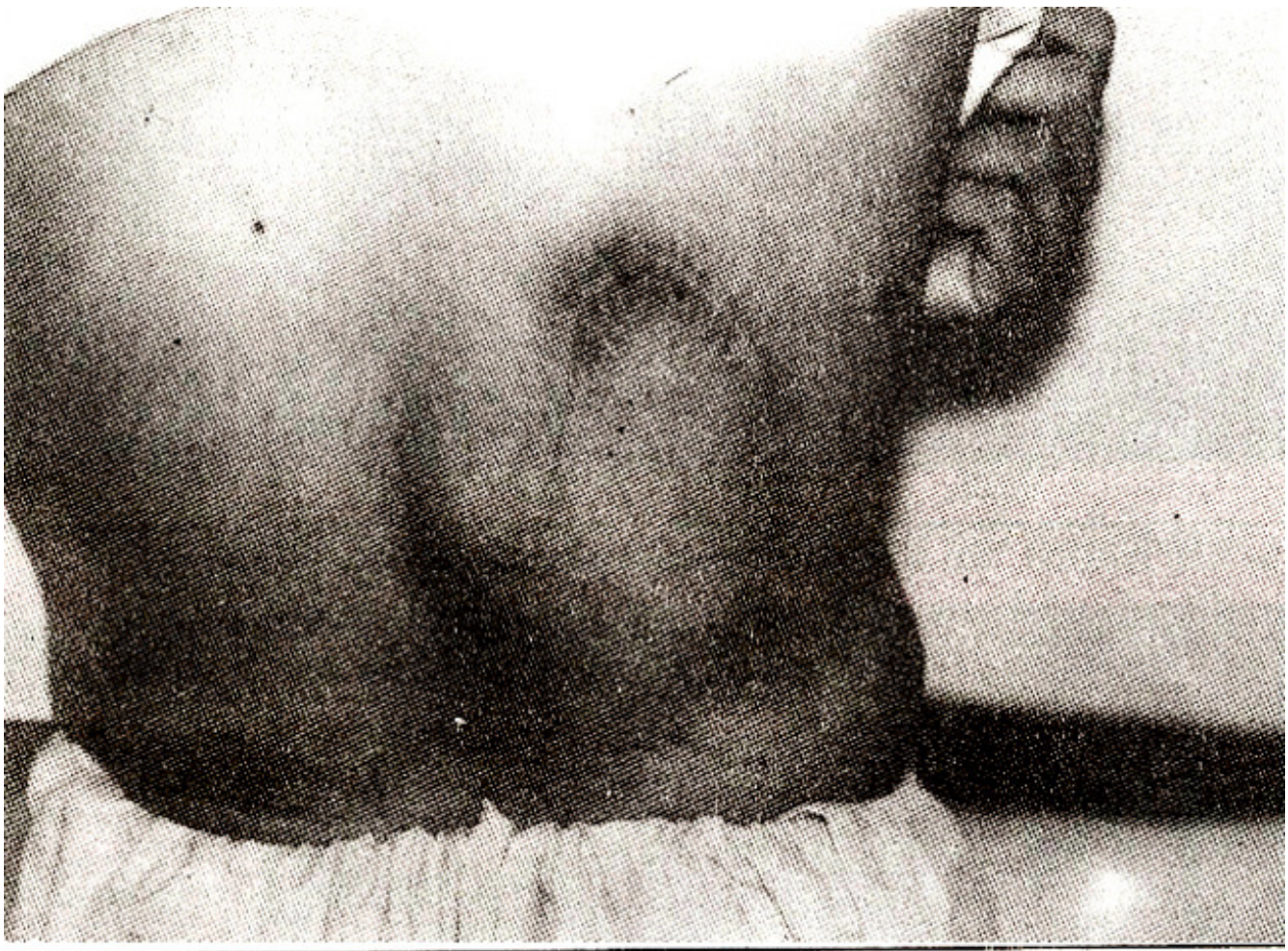


Figure 5. The resultant scar.

Discussion

In keeping with the rarity of the carcinomatous lesion reports in the literature are few. In the largest review of pilomatrix carcinoma to date totaling 20 such cases, Sau et al. reported that the tumours varied in size from 1—10 cm. in size and occurred more often in middle-aged men with a predilection for the posterior neck and back¹⁰. Follow-up of 17 patients revealed a local recurrence in 10 (59%) and multiple recurrences in 3 patients. As early as 1927 Gromiko reported a case of calcifying epithelioma with three recurrences which ultimately necessitated amputation of the right arm¹¹. Though the biologic potential of pilomatrix carcinoma is similar to basal cell carcinoma, as it is a locally invasive lesion that does not metastasize, yet there have been reports of pulmonary metastases¹² and multiple visceral metastases leading to a fatal outcome¹³.

The importance of the above is to highlight that not all tumours of hair matrix origin follow an innocent course. Add to it is the fact that it can at times be difficult to clinically distinguish a pilomatrix lesion in the adult from a variety of skin lesions like a sebaceous cyst, fibrohistiocytic proliferations, basal cell carcinoma, keratoacanthoma and cutaneous metastases⁴. Histologically the distinction between pilomatrixoma and pilomatrix carcinoma can be difficult and an erroneous diagnosis of the much common pilomatrixoma can be made. Flow cytometric DNA content analysis reveals no significant difference between pilomatrixoma and pilomatrix carcinoma¹⁴. After extensive review of the microscopic slides of their own and other cases which exhibited aggressive behaviour, Lopnasri and

Mihm⁸ advised caution in interpreting tumours with extensive basaloid proliferation and mitoses. **They put forward two microscopic features which, in their opinion, should denote pilomatrix carcinoma:**

i) Active proliferating hyperchromatic vesicular basaloid cells with numerous mitoses.

ii) Infiltration into fat lobules and/or into underlying structures.

In summary awareness of the malignant potential of certain skin tumours of hair matrix origin is important in preventing the untoward sequelae of inadequately managed pilomatrix carcinoma. Where there is the slightest doubt the lesion should be subjected to early wide excision and close monitoring thereafter.

References

1. Mallierhe A, Chenantais J. Note sur l'epitheliome calcifie des glandes sebacees. Prop. Med., 1880;8:826-28.
2. Forbis R Jr., Helwig EB. Pilomatrixoma (calcifying epithelioma). Arch. Dermatol 1961 ;83:606-18.
3. Taffe A, Wyatt EH, Bury HPR. Pilomatricoma (Mallierhe) A clinical and histopathological survey of 78 cases. Int J. Dermatol., 1988;27:477-8ü.
4. Kaddu S, Soyer HP, Cerroni L, et al. Clinical and histopathological spectrum of pilomatricomas in adults. Int J. Dermatol., 1994;33:705-8.
5. Aslan G, Erdogan B, Aköz I, et al. Multiple occurrence of Pilomatrixoma A case report. Plast. Reconstr. Surg., 1996;98:510-13.
6. Singh B, Tolette-Velcek F, Alexis R. Pathological case of the month. Arch. Pediatr. Adolesc. Med., 1995;149:551-52.
7. Sasaki CT, Yue A, Enriques R. Giant calcifying epithelioma. Arch. Otolaryngol . 1976;102:753-55.
8. Lopansri S, Mihm MC. Pilomatrix carcinoma or calcifying epitheliocarcinoma of Mallierhe. Cancer, 1980;15:2368-73.
9. Sehrudde J, Petrovici V. The use of slide-swing plasty in closing skin defects: A clinical study based on 1308 cases. Plast. Reconstr. Surg., 1981;67:467-68.
10. Sau P, Lupton GP, Graham JH. Pilomatrix carcinoma. Cancer, 1993;71:2491 —98.
11. Grotniko N Zur Kenntnis der bosartigen umwandlung des kalkigen hautepitheliomas Arch. Pathol. Anat., 1927;265: 103-16.
12. Gould E, Kozzon R, Kowalczyk AP, et al. Pilomatrix carcinomas with pulmonary metastasis. Cancer, 1984;54:370-72.
13. Niedermeyer HP, Pens K, Imler H. Pilomatrix carcinoma with multiple visceral metastases report of a case. Cancer, 1996;77:1311-14.
14. Rabkin MS, Wittwer CT, Soong VY. Flow cytometric analysis of a case of pilomatrix carcinoma showing multiple recurrences and invasion of the cranial vault. J. Am. Acad. Dermatol 1990;23:104-8.