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Pilomatrixoma and pilomatrix carcinoma deceptively similar but distinct entities

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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 "Clacifying Epithelioma o Malherbe" named thus in recognition of Malherbe\'s description of the entity in I 880, even though erroneously ascribed the tissue of origin to be the sebaceous gland'. 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 lesion [2]. These are rare benign skin neoplasms with an incidence rate that varies from 1 in 924 dermatopathologic specimens to I 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 al [3] and more recently Kaddu et al[4] 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 calcification[5] Histologically they are composed of nests of basaloid cells which undergo abrupt keratinization forming ghost or shadow cells in a background of inflammatory ce Is and calcification[6]. The recommended treatment is excision which is usually a simple matter as the lesion is well circumscribed. Recurrence is rare [2] but has been documented. Though the term 'giant calcifying epithel ioma" had been coined[7] to denote a tumour that behaved aggressively and recurred, it was not until 1980 when Lopansri and M ihrn flrst used the term "pi lomatrix 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 1994[4].

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)

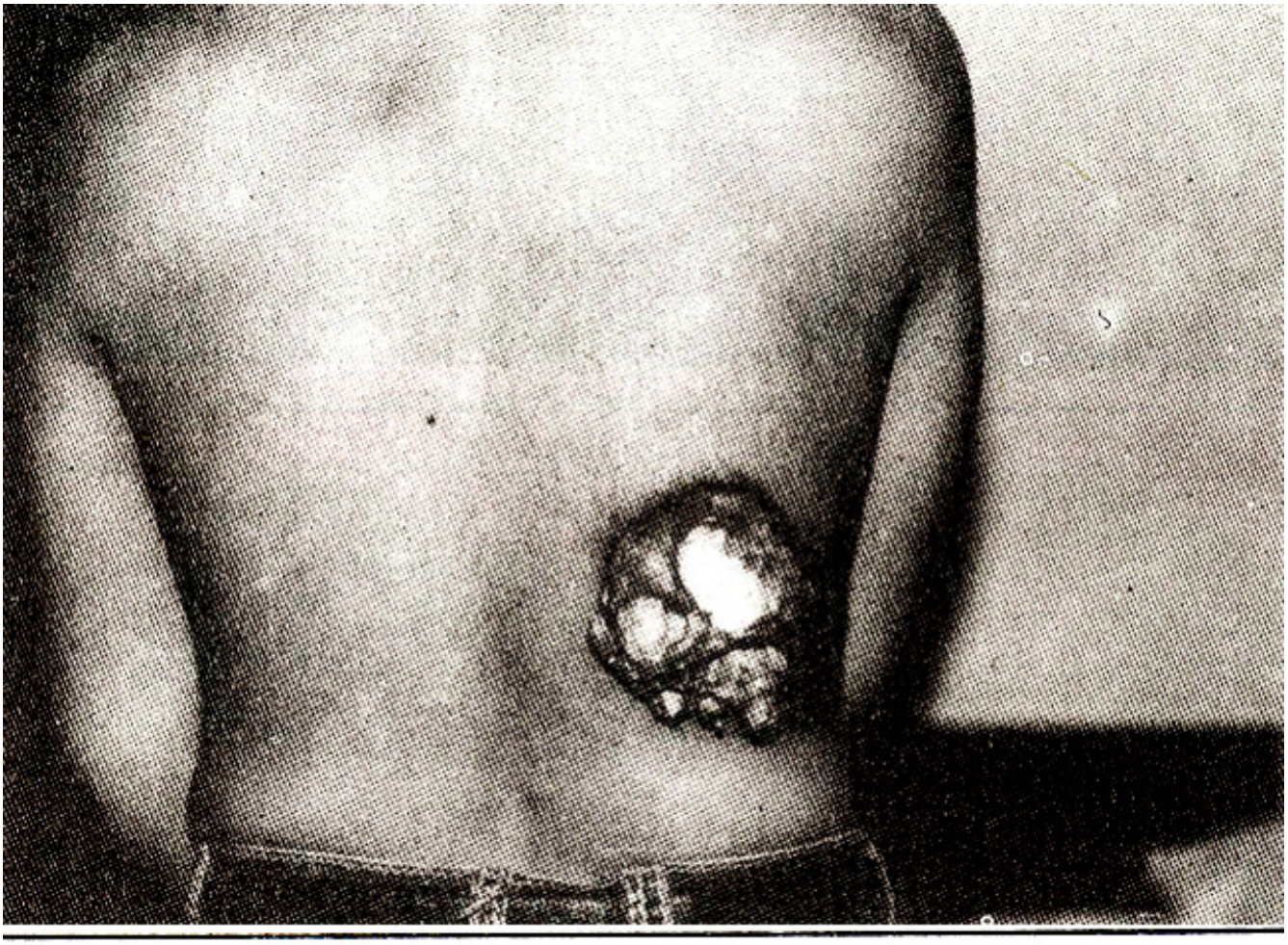

Figure 1 Pre-operative view of the lesion

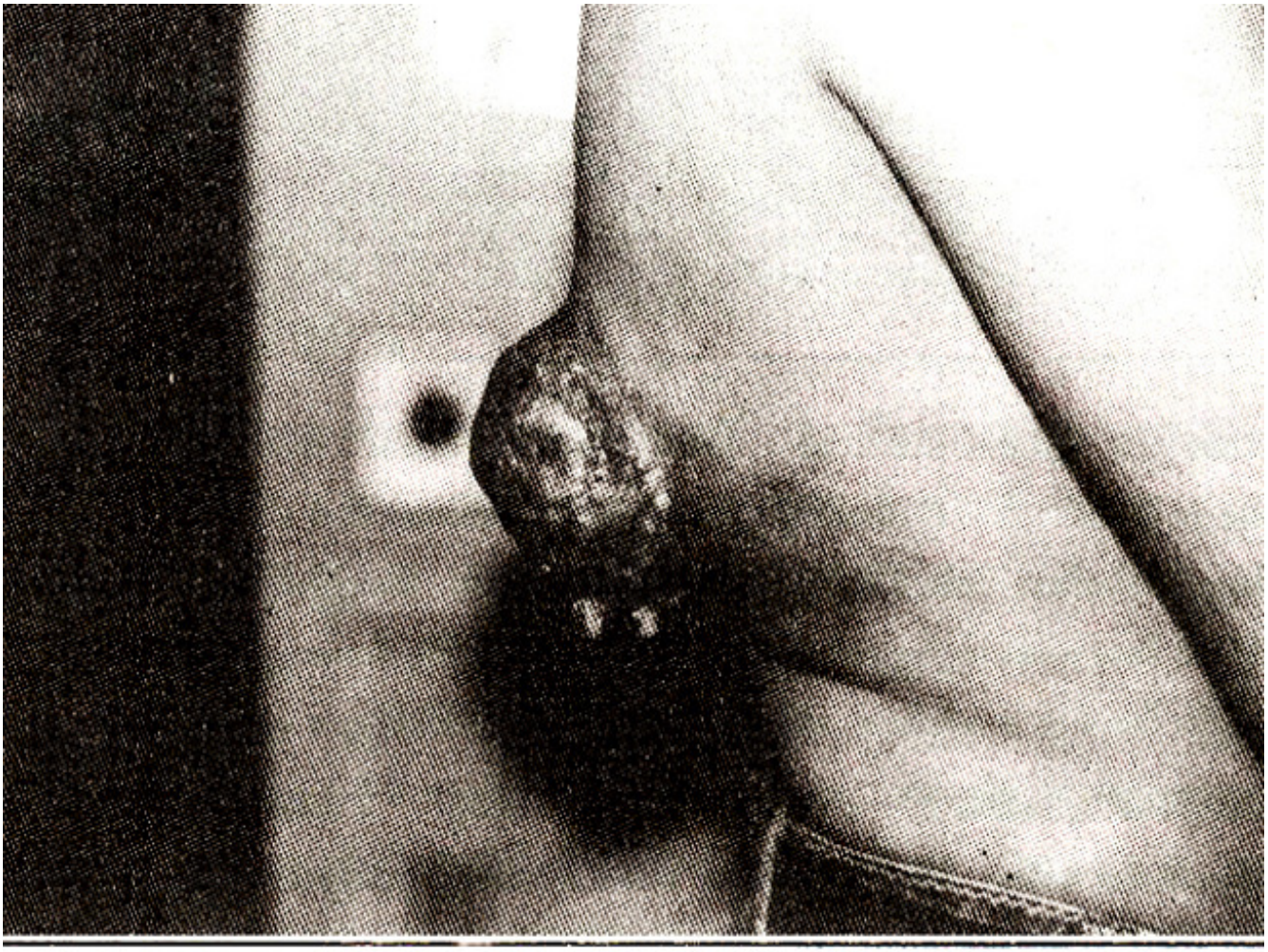

Figure 2. Pre-operative view of the lesion.

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 lvrnphadenopathy 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 l9 was desigiied (Figure 3)

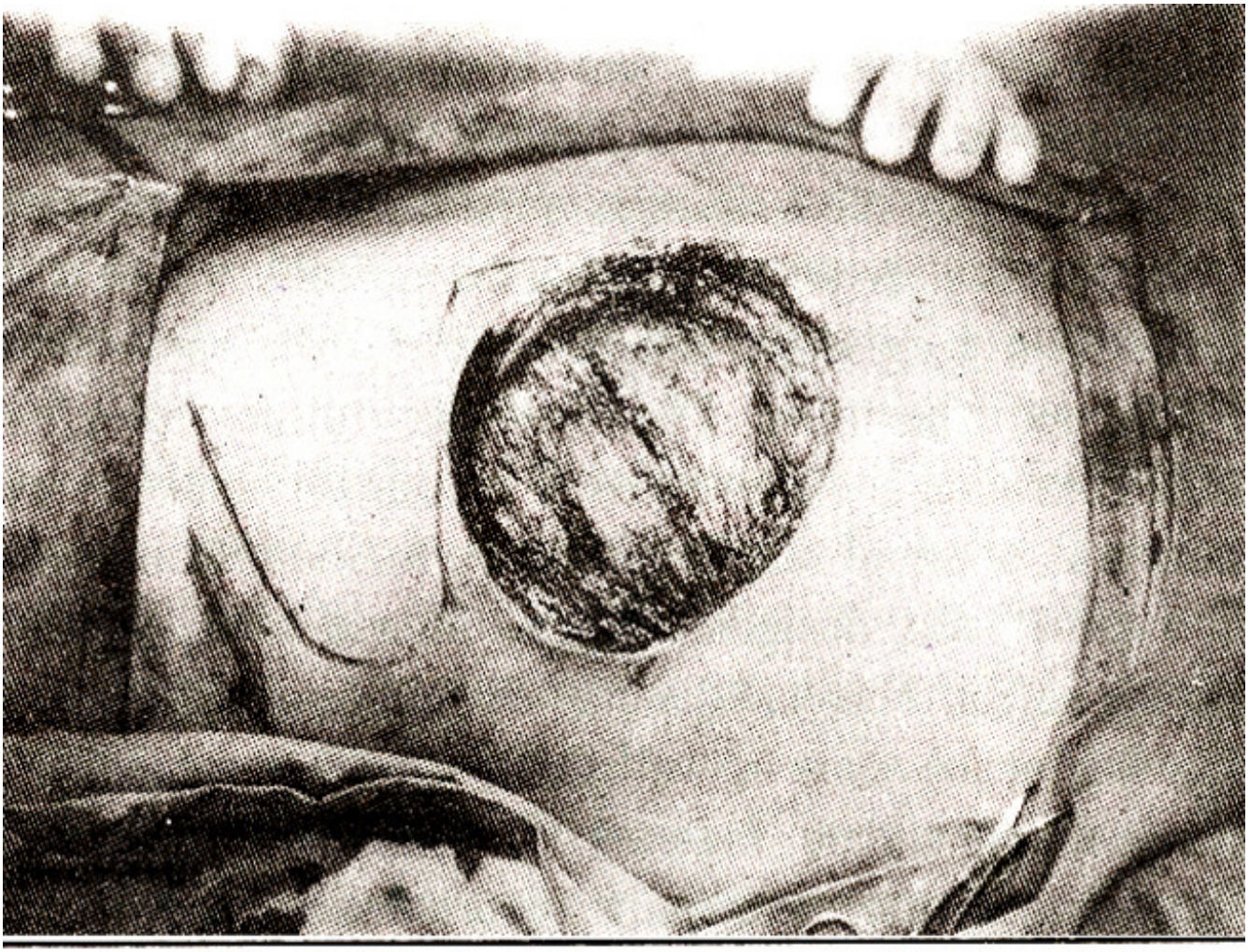

Figure 3. The defect following excision and the slide-swing flap type I marked out.

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).

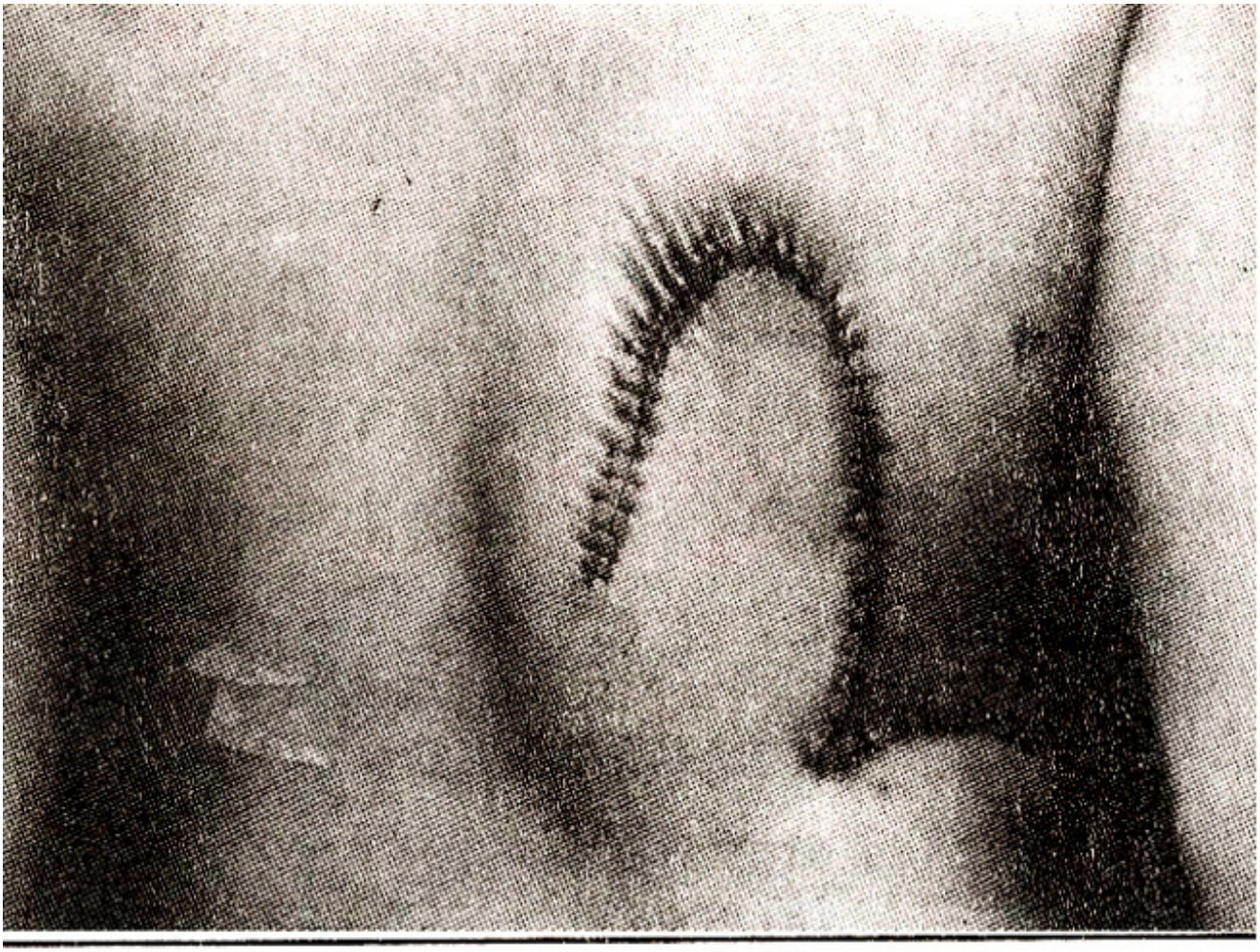

Figure 4. Closure of the defect

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 tinie 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).

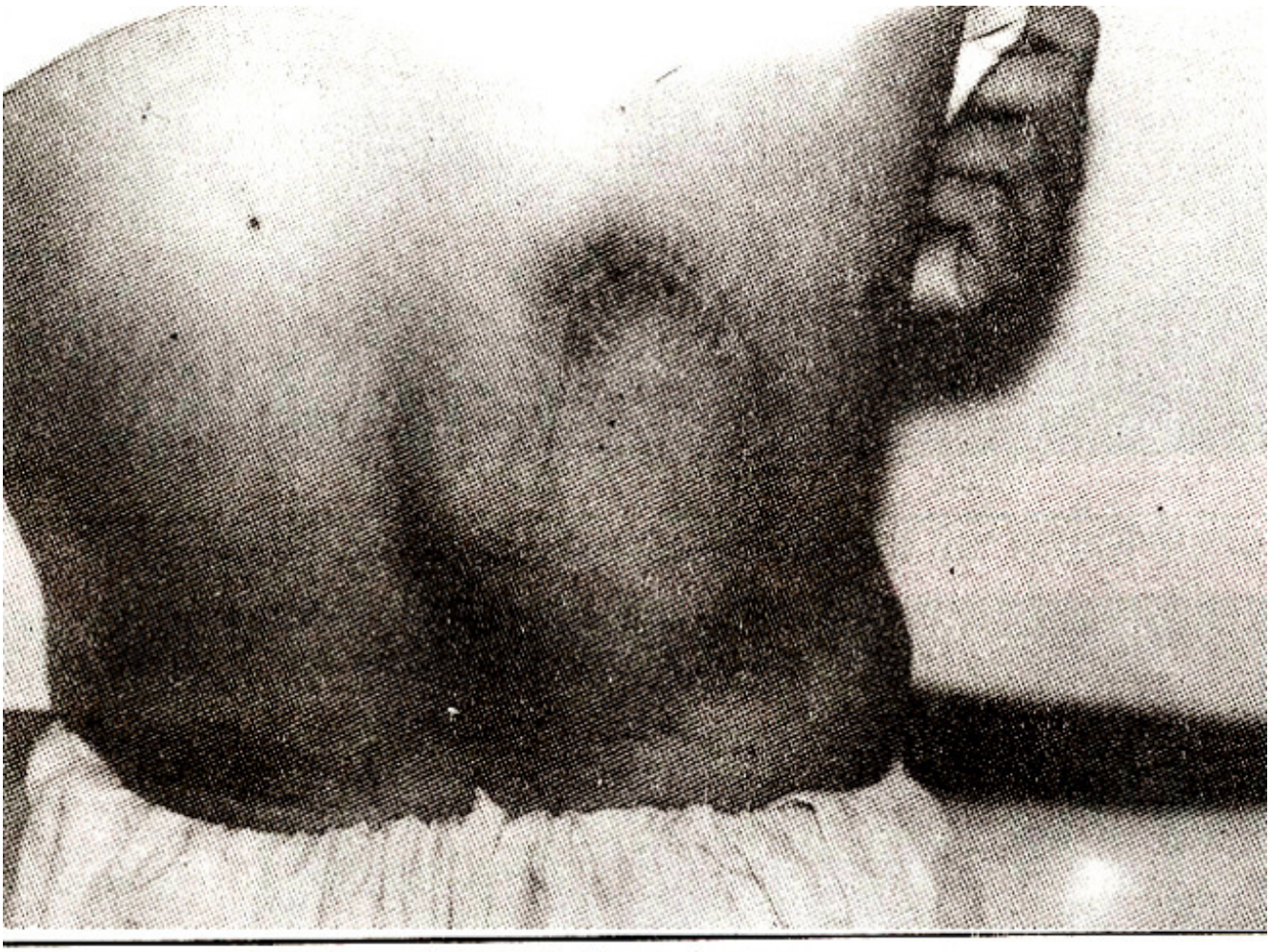

Figure 5. The resultant scar.

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 back[10]. Follow-up of 17 patients revealed a local recurrence in 10 (59%) and multiple recurrences in 3 patients. As early as 1927 Gromiko reproted a case of calcifying epithelioma with three recurrences which ultimately necessitated amputation of the right arm[11]. Though the biologic potcntial of pilomatrix carcinoma is similar to basal cell carcinoma, as it is a locally invasive lesion that does not metastasize, yet there have beeii reports of pulmonary metastases[12] and multiple visceral metastases leading to a fatal outcome[13].

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 metastases [4]. Histologically the distinction between pilomatrixoma and pilornatrix 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 carcinoma [14]. After extensive review of the microscopic slides of their own and other cases which exhibited aggressive behaviour, Lopnasri and

Mihm [8] advised caution in interpreting tumours with extensive basaloid proliferation and rnitoses.
They put forward two microscopic features which, in their opinion, should denote pilomatrix carcinoma:
1) Active proliferating hyperchromatic vesicular basaloid cells with numerous rnitoses.
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 he subjected to early wide excision and close monitoring thereafter.

References

1. Mallierhe A, Chenantais J. Note stir I\'epitheliome calcifie des glandes sebacees. Prop. Mcd., I 880;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 (Malherbe) A cliuteul and histopathological survey of 78 cases. Int J. Derruatol., l988:27:477-8ü.
4. Kaddu 5, Soyer ftP, Cerroni L, et al. Clinical and histopatliological spectrititi of piloinairionias in adults. Int J. Dcrntatol., 1994;33:705-8.
5. AsIan G, Erdogan B, Akoz 1, et al. Multiple occurrence of Pilomatrixoma A case repoil. Plast. Reconstr. Surg., 1996;98:510-13.
6. Singh B, Tolete-Velcek F, Alexis R. Pathological ease of the otouth. Arch. Pediatr. Adolese. Med., 995;149:551-52.
7. Sasaki CT, Yue A, Enriques R. Giant calcifying cpithelioina. Arch. Otolarygol . 1976;102:753-55.
8. Lopansri 5, Mihm MC. Pilomatrix carcinoma or calcifying epitheliocarcinoma of Mallierbe. Cancer, 1980:15:2368-73.
9. Sehrudde J, Petrovici V. The use of slide-swing plasty in closing skin defects: A clinical study hated on 1308 eases. Plast. Reeonstr. Surg., 1981:67:467-68.
10. 5au P, Lupton GP, Graham JIl. Pilomatrix carcinoma. Cancer, I 993;7 1:2491 —98.
11. Grotniko N Zur Kenntnis der bosartigen umwandlung des kalkten hautcpitheliomas Arch. Pathol. Anat., 1927:265: 103-16.
12. Gould E, Knrzon R, Kowalczyk AP, et al. Piloinatrix eareinoms with pulmonary metastasis. Cancer, 1984:54:370-72.
13. Niedermever HP. Pens K, I loller 11. Pilomatrix carcinoma with multiple visceral metastases report of a ease. Cancer, 1996;77:1311-14.
14. Rabkin MS, Wtttwer CT, Soong VY. Flow cytometric analysis of a case of pilomatrix carcinoma showing multiple recurrences and invasion of the cranial vault. J. Ant. Acad. Dermatol 1990;23:104-8.