PERIORBITAL KERATOACANTHOMATA*†

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ALTHOUGH usually coming under the purview of the dermatologist, keratoacanthomata occur sufficiently frequently near the eye to engage the attention of the ophthalmologist. His advice may be sought, not only in the matter of diagnosis, but also when there is interference with vision or when the tumour encroaches uncomfortably near to the eyelid margins. It will be for him to decide between radical and expectant treatment.

Clinically, the lesions have quite distinctive and uniform characteristics and they run a predictable course. In the early proliferative stage, the tumour rises sharply from the normal skin and has a covering of skin which appears stretched. It is hemispherical and is surmounted by a firmly adherent, central, keratinous plug. Fine telangiectases sometimes traverse its smooth surface. In size, it may vary between 0-3 and 2-0 cm. Enlargement continues evenly for from 2 to 8 weeks. The maximal size may be maintained for several weeks before the stage of regression ensues.

Unless radical measures of treatment are adopted, the tumour may take up to a further 12 weeks to recede completely. In the process, there is contraction of the borders, elongation of the central cavity, extrusion of the corneous plug and flattening of the whole lesion. Finally a scar, which is normally not obtrusive, remains.

The first description of the lesion is generally ascribed to Jonathan Hutchinson (1888). In the ensuing years, descriptions of the tumours have appeared under a large variety of titles, the term molluscum sebaceum given by MacCormac and Scarff (1936) having attained considerable popularity. Subsequently the label keratoacanthoma has emerged and now meets with general acceptance.

That keratoacanthomata are by no means rare is shown by Baer and Kopf (1962) in a comparison of the incidence of squamous cell carcinoma and keratoacanthomata. A summation of the numbers from seventeen published series showed that the relative figures of the two types of tumours were 3,866 squamous cell carcinoma to 883 keratoacanthoma, i.e. 18 per cent.

From the same source comes the analysis of the distribution of the lesions. Of 592 tumours, 420 affected the face and of those 33 (5·6 per cent.) appeared on the eyelids. At the same time it was reported that 161 affected the cheeks, 100 the nose, 23 the forehead, two the eyebrows, and thirteen the temples. It is thus apparent that periorbital lesions represent a not inconsiderable proportion of all tumours and the majority of facial tumours.

In another analysis, Baptista (1966) found that, of 1,270 observations, 60·25 per cent. were made up of squamous cell carcinoma, 24·09 by basal cell carcinoma, 7·87 per cent. by keratoacanthomata, and 7·9 per cent. by Bowen’s epitheliomata.

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So distinctive are the clinical features of keratoacanthomata that there is rarely difficulty in diagnosis. It is, of course, important always to exclude cystic rodent ulcers adjacent to the eye with some superficial resemblance to a keratoacanthoma (Fig. 1). The longer history, the unremitting progress, the periodic crust detachment, the translucency, and the softer consistency are pointers to a basal cell carcinoma.

FIG. 1.—Hemispherical basal cell carcinoma having superficial resemblances to a keratoacanthoma.

Opinion is divided regarding the malignant metaplastic potentiality of keratoacanthomata. That these normally benign lesions can occasionally become malignant was claimed by Belisario (1965) who quoted four cases of his own and listed the following published references: Dupont (1954), Ferguson Smith (1948), Zimmerman (1956), Beerman (1957), Belisario (1959, 1962, 1964), Lapière (1962), Glazunov (1961), Nicolau (1964), Maragnani (1965).

On the other hand, Champion and Rook (1963) challenged the claim that malignancy has ever been conclusively proved. They referred to the difficulty of histological proof and the onus of deciding between the pseudo-epitheliomatous proliferation encountered in keratoacanthomata and the alleged metaplasia. Julian Huxley would expect any cancerous change to be demonstrable biologically with cancerous invasions of tissues, and metastases.

Baptista (1966) described six such borderline cases and noted deep trabecular elongations around the nerves, an observation described only in carcinoma, but still concluded only that the possibility of malignancy remained problematical. Inasmuch as all keratoacanthomata suspected of becoming malignant would be promptly excised, it seems to us that this problem may well prove insoluble.

Recurrences of keratoacanthoma are uncommon but well authenticated. Baer and Kopf (1962) referred to recurrences after only partial surgical removal and also after repeated attempts at eradication; Belisario (1959–1965) to atypical forms appearing after treatment; and Champion and Rook (1963) to recurrences in cases after treatment. It seems probable that certain measures of treatment actively predispose to recurrences.

In a consideration of treatment, the policy of "masterly inactivity" has much to recommend it, especially when the clinical appearances leave no doubt as to the diagnosis. There is, however, a body of opinion in favour of early excision and amongst the reasons put forward are the better cosmetic appearance of the resultant scar, the removal of doubt as between a squamous cell carcinoma and a keratoacanthoma, immediate improved cosmetic appearance, and the avoidance of malignant change or recurrence.

Barry (1962) mentioned a compromise as a third possibility, especially where the clinical diagnosis can be made with confidence and symptoms render some form of radical treatment necessary. In these cases partial removal by paring down all protruding tissue
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followed by curettage, cauterization, freezing, or electrocoagulation can be adopted. The advantage of this procedure is that it allows a limited histological examination. However, we consider that the risk of recurrence is greater after measures which include curettage.

The similarity of histological features in a keratoacanthoma and a squamous cell carcinoma are sometimes such as to render the pathologist reluctant to make a diagnosis on a histological basis alone. He may claim that the rapid growth of the tumour results in a variety of histological pictures and that the changes of low grade squamous cell carcinoma approximate to the pseudo-epitheliomatous proliferation of a keratoacanthoma and are not sufficiently precise for absolute differentiation.

Barry (1962) described keratoacanthoma in ocular pathology, referring specifically to six specimens received in the laboratory. Stress was laid on the difficulty in distinguishing the tumour from epidermoid carcinoma because of pseudo-invasion, atypical cytology, and the presence of mitotic figures. Often, too, the specimens received from ocular biopsies are small or sectioned tangentially and so increase the likelihood of a wrong diagnosis.

However, in dealing only with the eruptive stage of a classical lesion, certain gross features may be regarded as characteristic. Bisection of the tumour results in the exudation of a cloudy fluid and the cut surface has a yellow-brown colour with some peripheral streaking from keratinization. The keratinous crater, too, will be apparent. Under the low power, the peripheral epithelium is seen to be stretched and thinned towards the summit where it suddenly folds back on itself at the corneous plug to form an epithelial “overhang”. The massive acanthosis results in the pseudo-epitheliomatous proliferation which projects through the subjacent tissues as downgrowing epithelial pegs.

The initial changes are localized in the upper half of the hair follicle and in the immediately adjacent epidermis. They consist of a thickening of the external epithelial sheath followed by dilatation of the lumen into a craterlike excavation. Proliferation of the cells of the malpighian layer laterally and in depth produces the bulk of the tumour. The basement membrane largely remains, although it may be ruptured in places. There is marked cellular reaction in the dermis which shows some hypertrophic prolongation between the epithelial downgrowth. The inflammatory infiltrate is constant and consists mainly of lymphocytes and histiocytes but also of polymorphs, eosinophils, and plasma cells.

The pseudo-epitheliomatous cytological abnormalities include nuclear polychromasia, abnormal mitoses, loss of cellular polarity, variations in size, a tendency to form keratin pearls, and cellular spindles penetrating deeply into the dermis.

Case History

A man aged 66 years presented at the Out-patients Department on April 18, 1967, complaining of a painless nodule on the nose just medial to the left eye. It had commenced some 6 weeks before and was enlarging fairly quickly. The lesion was hemispherical, sessile, and smooth and had a prominent keratotic plug. It measured 1.3 cm. across.

He was observed during the ensuing weeks and the increase in size continued. By mid-May (Fig. 2), it appeared to have reached its maximum size of 1.9 cm. and there was slight ulceration at one margin. Most of the surface was taken up with the dark-brown, almost black, plug. By June 2 recession had begun and the lesion consisted mainly of a large horn on a base which was becoming constricted (Fig. 3). Recession continued during the next 4 weeks and on July 14 the horn “dropped off”.

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Fig. 2.—The tumour at its maximal stage of evolution.

Fig. 3.—Basal constriction of regressing keratoacanthoma just before its spontaneous detachment.

A perfectly healed crater remained (Fig. 4), the floor of which contained two separate small elevations. The patient was not seen until August 15, when a rapid recrudescence was noted and a tumour almost as large as the original one was found (Fig. 5). It was hemispherical but had an irregular surface and there was no keratotic plug. At the pole nearest to the inner canthus, there was a small ulcerated area.

Fig. 4.—Crater left after detachment of keratoacanthoma. Photomacrograph.

Fig. 5.—Recurrent tumour.

The fear of malignant change forced the decision to treat the nodule surgically. Radical excision was carried out using basal sedation and local anaesthesia.

Basal sedation with pethidine 50 mg., largactil 25 mg., and phenergan 25 mg. was given intramuscularly one hour before the operation. Local infiltration with lignocaine (Xylocaine) 2 per cent. and 1/10,000 adrenalin was used under and around the lesion and also in the mastoid region from which a full-thickness graft was taken.

The tumour was excised along with 2 mm. of surrounding healthy skin. There was no involvement of the underlying bone and, after the removal of the lesion, the medial palpebral ligament, angular vein, and fascia covering the lacrimal sac could be clearly seen in the dissected area.

Since the wound to be closed was about 2·5 cm. across and the skin of the inner canthal area was thicker than that of the lids and more firmly adherent to deeper structures, it was not possible to mobilize the skin into flaps. The danger that contraction in this area may produce unsightly
epicanthal folds and eversion of the puncta with resultant epiphora has been pointed out by Sorsby (1964).

It was therefore decided to cover the defect with a full-thickness skin graft which was taken from the retroauricular area just below the mastoid region. The graft was sutured to the recipient site with 6-0 silk sutures, the suture ends being left long. Because of the difficulty in maintaining pressure with the usual eye dressing in an area such as the inner canthus, an onlay graft in the form of a Stent mould was used after covering the graft with Vaseline gauze, the long suture ends being then tied over the mould. Firm, moderate pressure was thus assured over the graft area.

The first dressing was carried out after 7 days, the Stent mould being removed and the suture ends being cut short. The sutures were removed on the tenth day. Post-operative progress was uneventful and the graft took well. Fig. 6 shows the patient's appearance 4 months after the operation.

The possibility of malignancy was confirmed by the pathologist's report, which described a specimen 1.5 cm. in diameter, consisting of a squamous epithelium tumour the configuration of which would do well for a keratoacanthoma. However, there was a good deal of vigorous mitotic activity at the periphery of the growth and it was considered impossible histologically to dismiss malignancy (Fig. 7).
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Discussion

Because of the proximity of the tumour to the eye, conservative treatment was decided upon in the knowledge that spontaneous regression would ensue. It was also felt that an excision so near the inner canthus might conceivably result in ectropion. From the onset of the condition to its final detachment, some 19 weeks elapsed. This proved a trying time for the patient because its presence in his field of vision made him conscious of it at all times.

The healthy appearance of the scar after the pedicle had detached, gave rise to confidence that the problem had finally been resolved.

A rapid resurgence of the tumour almost to its original size was a startling phenomenon. The appearance was different from that of the original nodule and it was ulcerated over one segment. There appeared a very real probability that malignant metaplasia had occurred and no alternative to radical excision now seemed practicable, but there seemed some danger of constriction of the palpebral aperture if an extensive operation had to be undertaken. The histological appearances of the excised specimen tended to confirm that the new lesion was malignant as far as histological criteria can be accepted.

The final cosmetic result is, in fact, excellent with virtually no scarring (Fig. 6). After 6 months, there is no evidence of recurrence.

Summary

The nature, incidence, clinical findings, pathology, and treatment of periorbital keratoacanthomata are briefly described and the literature reviewed. Spontaneous retrogression, recrudescence, and possible malignant metaplasia are particularly considered, and these are illustrated by a case manifesting all these features.

In the management of keratoacanthomata near the eye, excision followed by skin grafting may be obligatory.

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