PILOMATRIXOMA (CALCIFYING EPITHELIOMA OF MALHERBE AND CHENANTAIS)

(A case report)

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Summary

A case of pilomatrixoma (calcifying epithelioma) occuring in lumbar region of a male patient, was described and discussed in relation to co-existing literature and was considered of interest to be placed on record, being the first case report from Marathwada region, Maharashtra State.

This unusual benign neoplasm of the upper dermis was first described by Malherbe and Chenantis in 1880. Since then several reports have appeared in the literature (Chin, 1933; Cote, 1936; Ferve et al, 1938; Highman and Ogdan, 1944 and King, 1947). In 1961 Forbis and Helwig noted that about 300 cases were then in the literature. In view of the obvious inadequacies of the term 'calcifying epithelioma' the name pilomatrixoma was suggested by Forbis and Helwig. This name may be offensive to the language-purist, but it has the advantage of conveying the histogenesis of the tumour and avoids the use of the word 'epithelioma', which generally indicates a malignant tumour. The case described below was considered of interest to be placed on records, being the first case report from the Marathwada region, Maharashtra State.

Case Report

A 40 year old male was admitted in the Medical College Hospital, Auranga-

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bad, with a small firm mass in the right lumbar region for the past 3 He noticed a small pea-sized firm but painless nodule which had progressively increasing in size. examination a hard lobulated mass measuring about 9 cm. in diameter was present in the right lumbar region. It was freely mobile over the deep seated muscles. The skin over the mass was tense but not adherent. It was not tender. The lymph nodes were not enlarged. A clinical diagnosis of fibroma was made. Patient was reported to be in good health one year after the operation.

Macroscopic Pathology

The specimen consisted of an excised mass measuring 2x1x1 cm. in size. It was completely enclosed within a fibrous capsule. The cut surface showed the tumour mass to be greyish white in colour and firm in consistency. A few smaller satellite calcified nodules varying from 0.2 to 0.5 cm. in size were found enclosed within the same fibrous capsule but were separated from the main tumour mass by thin fibrous septa.

Microscopic Pathology

The tumour mass had a definite fibrous capsule and was composed of epithelial masses of variable sizes separated by fibrous stroma (Fig. 1).



Fig. 1
Shows epithelial masses of variable sizes separated by fibrous stroma.

Two types of cells—basal cells (small. round, closely packed compact cell resembling hair matrix cells of the hair follicle) and shadow cells (Polyhedral, lightly eosinophic, faded cells, which contain shrunken, unstained or pyknotic nuclei). The Two cells types blend with one another (Fig.2), and about against histiocytic giant cells, masses of cornified cells and amorphous and cornified debris. The fibrous stroma showed poor vascularity and was densely infiltrated with lymphocytes and a few plasma cells and giant cells (Fig. 3).

Discussion

As the patient presented with a mobile firm mass in the lumbar region, clinical diagnosis of fibroma was made.

The presence of large sheets of squamous epithelial cells with areas of cornification and masses of keratin surrounded by large number of giant cells ruled out this possibility on histological examination. The histologic appearance of the tumour was, however, characteristic for calcifying epithelium.



Fig. 2

Shows sheaths of basal cells and shadow cells blend with one another.

This tumour is a relatively infrequent one. The incidence reported in the literature has been-10 calcifying epithelium out of 22000 consecutive surgical specimens (Ch'in1), 12 out of 24000 specimens (Cote,2) and 9 out of 7500 specimens (King⁷). Most frequent site has been head and neck. Till 1933, out of 116 cases reported in literature, 57 were situated in head and neck, 19 in upper extremity, 15 in the trunk, 4 in the lower extremity and 21 at unspecified sites (Ch'in,1). Of the 15 cases in the trunk, in this series 3 were found situated in breast and nipple region and 1 in the pectoral region. Mehrotra et al (1967) reported a case

in the male breast. The tumour varied between 0.5 to 5 cm. in diameter in majority of the reported cases, however Mehrotra et al (1967) reported a large mass of 6x5 cm. in size.

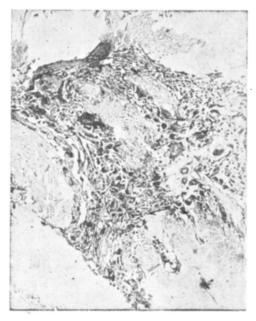


Fig. 3

Shows stroma densely infiltered with lymphocytes plasma cells and giant cells.

The current consensus favours the idea of pilomatrixoma arising from a primitive cell differentiating toward a hair matrix cell (Lever and Griesemer, 1949; Lever, 1967; Forbis and Helwig, 1961). Histochemical studies reveal in calcifying epitheliomas, especially at the

sites of transition of basophilic into shadow cells, a strong positive reaction for sulphydryl and disulfide groups, just as is seen in the keratogenous zone of normal hair (Forbis and Helwig.4 Hashimoto et al², Lever⁸). That the shadow cells contain keratin is indicated by their strong birefrigence in polarized height. Peterson and Hutt (1964) studied 26 cases of pilomatrixoma. All the tumours were found to show positive reactions with one or more of the histochemical procedures (PAS, Hale's, dihydroxyl-dinaphthyl disulfide, congo red, methyl violet and Purssian blue). Presence of amyloid was suggested by them as number of staining methods for amyloid was positive.

Electron microscopic examination reveals in the basophilic cells only few desmosomes but a moderate number of tonofilaments, largely in perinuclear arrangement (McGavran, 1965; Hashimoto et al, 1966) and thus the current concept regarding the histogenesis of pilomatrixoma is that it arises from primary epithelial germ cells with a tendency to differentiate into keratotic hair cells (Lever⁸). The histological findings in the case under record would support this concept.

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