MULTIFOCAL PILOMATRICOMA

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A 10-year-old male child developed four tumours of multifocal pilomatricoma.

Key words : Pilomatricoma

Pilomatricoma is an uncommon benign skin tumour of the hair matrix cells[1] seen as a single or multiple, asymptomatic, deeply-seated, firm nodule, covered by normal or pink skin which on stretching may show the 'tent sign' with multiple facets and angles.[2] Fifty percent of all lesions are seen on head and neck, 25% occur on the arms and the remainder are distributed over the trunk and lower extremities.[1] Multiple tumours, especially four or more are extremely rare.[1,3,4] Forbis and Helwig[5] in their study of 228 patients with 240 tumours found only 1 case with 4 tumours, 1 case with 3, and 7 patients who had 2 tumours. The occurrence of four pilomatricomas at atypical sites including the ear-lobule prompted us to report this case.

Case Report

A 10-year-old male, sikh had developed four nodules distributed over the external ear, neck and upper extremity during the last 2 years. The first lesion started spontaneously on the right ear without antecedent history of trauma, burning or itching in the lesion. Soon, three more lesions appeared on the neck, arm and hand. The lesions were asymptomatic except for tenderness on pressure, were deep seated, 1 cm-1.5 cm in size, firm, rounded and attached to the overlying skin. The overlying skin was slightly bluish but there was no punctum. On stretching the skin over the lesion, the 'tent sign' with multiple facets and angles could be demonstrated. General physical and systemic examination of the patient did not reveal any abnormality. Routine investigations were within normal limits. Histopathological examination revealed principally two types of cells. One group comprised of basophilic cells consisting of closely aggregated nuclei with a negligible amount of cytoplasm around them. The other, larger group was made up of shadow cells (Fig. 1). These comprised of close aggregation of cells with pinkish cytoplasm and central unstained area indicating the site of their nuclei. Focal areas of stromal keratinization and giant cell reaction were clearly evident (Fig. 2). However, calcification within the tumour masses was not seen.

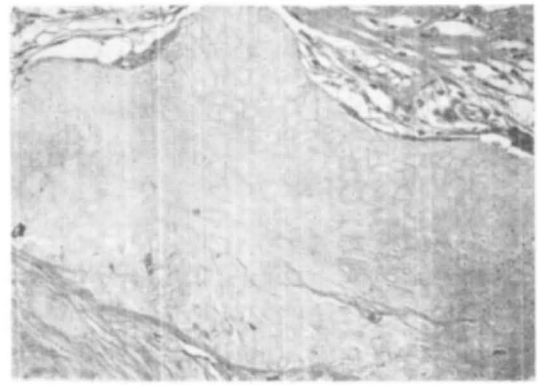

Fig. 1. Closely aggregated shadow cells showing centrally unstained area.

Comments

Pilomatricoma is a benign skin tumour which was first described by Malherbe and Chenantais[5] in 1918 under the name 'epithe' liome calcifis'des glandes se'bace'ces. They believed it to be of sebaceous gland origin. In 1961, the initial name of calcified epithelioma

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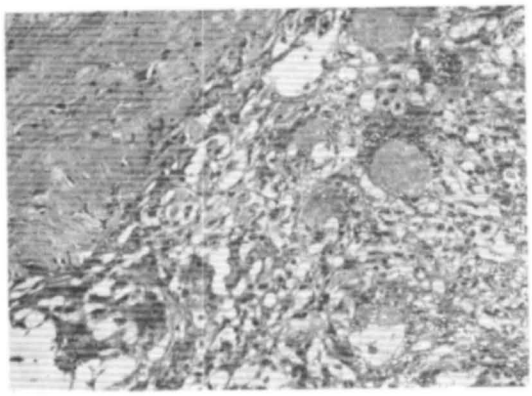

Fig. 2. Focal area of stromal keratinization and giant cell reaction.

of Malherbe was changed to pilomatrixoma by Forbis and Helwig[5] as it was found that the tumour originates from the hair matrix.

Diagnosis of pilomatricoma is rarely made on clinical examination. The characteristic deep-seated firm nature of the nodule with attachment to the overlying apparently normal skin and demonstration of the 'tent sign' are points in favour of pilomatricoma. In our case, the unusual features were multiple number of lesions and the presence of a lesion at an unusual site—the ear-lobule. Pilomatricoma has been reported to occur in associations with Gardner syndrome,[6] myotonic dystrophy[7,8] and hypercalcemia with skull dystrophy,[9] but our case had no associated abnormality.

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