

Extramammary Paget's Disease of the Axilla with an Underlying Apocrine Carcinoma

Sir,

The axilla is a rare but recognised site of extramammary Paget's disease (EMPD). Few cases have been fully documented. We here describe a case of axillary EMPD related to an underlying apocrine carcinoma and review the frequency and implications of this association.

CASE REPORT

A 71-year-old man with severe chronic obstructive airways disease (COAD) was referred with a 3-year history of a plaque in the left axilla, which had steadily enlarged and had become increasingly tender. It had failed to respond to treatment with imidazole creams. Examination of the left axilla showed a discrete red plaque measuring 6 by 3 cm, with an exudative surface (Fig. 1). The rest of his skin was unremarkable and no lymphadenopathy was detected. Histology of a punch biopsy showed rounded pale malignant cells scattered individually and in groups throughout the epidermis, particularly in the basal layer, consistent with EMPD. There was also secondary chronic inflammation. The intra-epidermal cells stained positively with periodic-acid-Schiff and showed positive immunoreactivity with antibodies against carcino-embryonic antigen and low molecular weight cytokeratin, confirming the diagnosis of EMPD. Screening investigations for an underlying malignancy, including serum prostate-specific antigen, liver function tests and full blood count, were normal and a chest radiograph showed no recent changes.

Before definitive excision could be performed, the patient died of an exacerbation of his COAD. His general practitioner obtained permission from the relatives to excise the whole of the lesion 12 h



Fig. 1. Exudative red plaque of extramammary Paget's disease in axilla.

after death. Histology confirmed EMPD and in addition some sweat glands in the dermis were filled with malignant cells similar to those in the epidermis, indicative of an apocrine carcinoma underlying the EMPD.

DISCUSSION

EMPD was first reported by Crocker in 1889 (1), in a case affecting the penis and scrotum. Subsequently the condition has excited much interest, mainly because of the difficulty in determining whether the Paget cells seen in the epidermis arise from migration of tumour cells from an underlying carcinoma or arise *de novo*. These distinctive cells are large (10–20 μm), with abundant pale vacuolated cytoplasm and a prominent nucleus, often placed eccentrically. Positive staining for carcino-embryonic antigen has established their glandular origin, and other specific immunoreactants, such as anti-gross cystic disease fluid protein antibody, have been used to prove their relationship to apocrine, not eccrine, sweat glands (2). It remains uncertain whether the cells originate in the epidermis and extend into adnexal structures or whether they represent direct extension into the epidermis of an adenocarcinoma from either an adjacent site, as in this case, or a distant site, such as the gastro-intestinal tract. Jones et al. (3) suggest that EMPD is more than one disease. They also theorise that, in some cases, pluripotential germinative cells within the epidermis respond to a neoplastic stimulus and attempt to form apocrine glands. Such a theory would explain the occasional case of multifocal EMPD (4–6).

Axillary EMPD is rare, with only one previous United Kingdom report by Gibson et al. (4). They described a case of multifocal EMPD of groins, scrotum and axilla, with an underlying apocrine carcinoma in the right groin, but not in the axilla. Kawatsu & Miki from Japan have described a similar case (5). We are aware of only three previous reports documenting 5 cases of axillary EMPD associated with an underlying apocrine carcinoma (6–8). In the largest recent series of EMPD, Jones et al. (3) reviewed the histological features of 55 cases. The numbers of sites affected were: vulva 39, scrotum 5, axilla 4, perianal 3, groin 2, buttock 1, pubis 1. In none of the 55 cases was there a neoplasm arising in an underlying adnexal structure, and it was concluded that such a situation must be "exceedingly rare".

Of the 11 cases of axillary EMPD previously recorded (3–8), 5 have been associated with an immediately underlying carcinoma (6–8). It is therefore important for such a lesion to be excised completely and for the specimen to be serially sectioned and carefully examined microscopically for a local sweat gland carcinoma. We report this case to increase awareness that EMPD can occur in the axilla and to highlight its frequent association at this site with an adjacent underlying apocrine carcinoma.

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