INTRODUCTION

The term pemphigus stems from the Greek pemphix (blister) [1] and describes a group of chronic blistering skin diseases in which autoantibodies are directed against the cell surface of keratinocytes, resulting in the loss of cohesion between epidermal cells, through a process called acantholysis. Pemphigus is divided into pemphigus vulgaris (PV) with a suprabasal acantholysis and pemphigus foliaceus (PF) with acantholysis in the upper parts of the epidermis. With these definitions in mind, we could classify pemphigus vegetans and pemphigus erythematous, types of pemphigus with certain unique clinical features, as subtypes of PV and PF, respectively.

4.2 CLINICAL FEATURES

4.2.1 Presentation

Pemphigus has a worldwide distribution with an annual incidence of approximately 0.1–0.5 per 100 000 population [2–5]. The prevalence of PV in patients of Jewish origin is increased, and the annual incidence of pemphigus has been reported to be about 1.6–3.2 cases per 100 000 Jewish population. The disease has a peak incidence of occurrence in patients between the fourth and sixth decade [6–8].

4.2.2 Cutaneous distribution, mucous membrane involvement and morphology of lesions

PV is the most common type of pemphigus and comprises about 80% of patients with pemphigus [6, 7]. In more than 50% of cases the disease begins with oral lesions, which may precede the cutaneous lesions by several months or may be the major, if not only, manifestation in some patients [6–9]. Even though 10–15% of patients present with cutaneous lesions only, the mucous membranes are ultimately involved in almost all cases [4, 6]. Mucous membrane lesions in PV usually are seen as painful erosions. Presumably because of trauma, intact bullae are rarely seen in the mouth. Lesions in the oral cavity are usually eroded, painful and heal slowly with peripheral extension through shedding of the epithelium. These lesions are of different sizes with irregular ill-defined borders. Most frequently buccal and palatine mucosa are involved [10]. Extensive erosions and painful lesions in the mouth may result in decreased food intake which in turn may contribute to hypoproteinaemia and hypalbuminaemia. Other mucous membranes may be involved and ulcerations of the vulva [11, 12] or the conjunctiva [13] may result in an initial misinterpretation of the diagnosis. Lesions of pharynx, larynx [14], oesophagus [15, 16], urethra, cervix [11, 12] and rectal mucosa [17] have been reported. If mucous membranes other than the oral cavity are involved, the course of the disease may be...
more severe [18]. A more generalized bullous phase occurs in many patients, if untreated, 5 or more months after the onset of oral lesions [19].

Cutaneous lesions in PV can be localized or generalized and usually present primarily as flaccid vesicles or bullae varying in size from less than 1 cm to several cm (Figure 4.1(a)). The scalp, face, axillae and groin are frequent sites of involvement. The content of the lesions is initially clear. The blisters may develop on normal skin or on an erythematous macule or plaque. In a few days the vesicular contents may become turbid, but often the blisters rupture easily and produce painful raw denuded areas. These eroded areas often extend at the edges due to continual peripheral loss of epidermis. Thus, relatively large areas of erosion are the main cutaneous lesions seen in many PV patients. Crusted verrucous lesions may also often develop at sites of ruptured bullae (Figure 4.1(b)) [18, 19].

Pemphigus vegetans, a subtype of PV, has been classically divided into two clinical types, Neumann and Hallopeau [20]. These two differ in their clinical presentation but it is probably accurate to think of PV, pemphigus vegetans Neumann and pemphigus vegetans Hallopeau as a spectrum from most to least severe. In the Neumann type of pemphigus vegetans the denuded areas often develop hypertrophic granulations, so-called vegetations, and in the periphery of extending vegetations pustules may be seen. Older

Figure 4.1 Clinical appearance of PV. (a) Patient’s ankle shows crusted erosions and flaccid blisters. (b) Patient’s sideburn area shows crusted verrucous lesions.