



Crispian Scully

Aspects of Human Disease

This series outlines, briefly, the clinical presentation, diagnosis and management of a further 20 chronic medical problems which are most common and important in the developed countries, but space also precludes coverage of acute infections and most malignant disease.

45. Vesiculobullous disorders

Damage to proteins that bind epithelial cells one to another, or to the epithelial basement membrane, can lead to blistering diseases of the skin and/or mucosae; these are termed vesiculobullous disorders. Most are acquired autoimmune disorders, and pemphigoid and pemphigus are the most important.

Pemphigoid

Mucous membrane pemphigoid (cicatricial or ocular pemphigoid) is a group of sub-epithelial immune autoimmune blistering diseases with autoantibodies directed to different epithelial basement membrane proteins, causing loss of attachment of the epithelium to the connective

Crispian Scully, CBE, MD, PhD, MDS, MRCS, BSc, FDS RCS, FDS RCPS, FFD RCSI, FDS RCSE, FRCPath, FMedSci, FHEA, DSc, DChD, DMed (HC), Eastman Dental Institute, University College London, 256 Gray's Inn Road, London WC1X 8LD, UK.

tissue, and blister formation.

Clinical features

- Intact mucosal vesicles or bullae are frequently seen.
- The gingivae are often affected by 'desquamative gingivitis'.
- Scarring is a serious complication in the eyes, larynx, oesophagus or genitalia.

Diagnosis

- The diagnosis must be confirmed by biopsy and immunostaining.

Management

- If there is suspicion of ocular involvement, an ophthalmological referral is essential.
- Other lesions can often be controlled by dapsone or topical corticosteroids.

Dental aspects

- Orofacial manifestations are as described above.

Pemphigus

Pemphigus is an uncommon potentially fatal autoimmune reaction against proteins in stratified squamous epithelium. Autoantibodies in the common form (pemphigus vulgaris) are directed

against desmoglein-3 of intercellular attachments (desmosomes) of epithelial cells.

Clinical features

- Pemphigus is characterized by widespread intra-epithelial vesicles and bullae frequently first appearing in the mouth.
- The blisters rupture and, in the absence of treatment, pemphigus is usually fatal.
- Stroking the skin or mucosa with a finger may induce vesicle formation in an apparently unaffected area or cause a bulla to extend (Nikolsky's sign).

Diagnosis

- Diagnosis is confirmed by biopsy and immunostaining.

Management

- Patients should be under the care of a physician.
- Systemic corticosteroids plus a steroid-sparing agent such as azathioprine, or mycophenolate mofetil are typically needed.

Dental aspects

- Orofacial manifestations are as described above.