Cicatricial Pemphigoid

Synonyms: benign mucous membrane pemphigoid, Brunsting-Perry disease, epidermolysis bullosa acquisita

Cicatricial pemphigoid (CP) refers to a group of rare chronic autoimmune blistering diseases affecting mainly mucous membranes (including the conjunctiva). A few patients have skin involvement which is usually around the head and neck or sites of skin trauma. Some patients have only eye involvement and a few only skin involvement. It is characterised by scarring of mucous membranes which can lead to complications such as blindness and supraglottic stenosis.

There is variety in the way CP presents clinically and some variety in the pathophysiological detail between patients. It can be difficult to distinguish from other autoimmune blistering diseases which may have mucosal involvement (for example, bullous pemphigoid). It is important to recognise the distinctive pattern of clinical features when making the diagnosis.

Pathophysiology

CP is an autoimmune disease with autoantibodies found in the lamina lucida (occasionally extending into the lamina densa) at the epidermal-dermal junction. These antibodies recognise a variety of antigenic components of the epithelial membrane. Bullous pemphigoid antigens 1 and 2 (BPAG 1 and 2) have been identified in CP along with a variety of others. There is a poor correlation between the particular circulating autoantibody and the clinical presentation.

Epidemiology

- CP is rare although precise figures for incidence are unknown.
- There appears to be an association between human leukocyte antigens and CP.
- The reported incidence in European countries is about 1 per million per year.
- The estimated incidence of ocular CP is between 1 in 8,000 and 1 in 46,000. The real frequency of the disease is probably higher because of diagnostic difficulty.
- It appears to be about twice as common in women.
- It occurs in older patients with an average age of between 62 and 64 years at onset.

Presentation

CP typically causes persistent and painful ulceration on mucous membranes, with progressive scarring. Oral ulceration can present a diagnostic problem. The main sites and types of involvement are:

Mouth
- Painful and recurrent erosions present anywhere in the mouth, starting as blisters.
- The gingivae are most commonly involved, but palate and buccal mucosa are frequently affected too.
- Dysphagia and hoarseness may signal involvement of the oropharynx.
- Swallowing may be impaired by progressive scarring leading to oesophageal stenosis.
- Subglottic stenosis may compromise the airway (tracheostomy then required).

Eyes
- This may start with conjunctivitis and grittiness of the eye.
- Dry eyes may result from lacrimal gland involvement.
- Ocular involvement can lead to blindness because of inflammation and scarring involving the:
  - Lens
  - Cornea
  - Iris
  - Lobe
- Some patients with CP have only eye involvement.

Nose
- This may manifest first with epistaxis.
- Painful, scarring erosions persist and alert to underlying disease

Skin
- This occurs in as many as a third of patients with CP.
- Intense, often generalised, pruritus accompanies skin involvement (tense blisters or erosions on normal or erythematosus skin).
Head and neck involvement is common. This may occur without mucosal involvement in, typically, elderly men (Brunsting-Perry variant).

Vesicles may be tense and haemorrhagic with scars and milia forming afterwards. Alopecia follows scalp involvement.\(^8\)

**Genitalia and perianal mucosae**

- Typically painful with pruritus and scarring.
- Involves the clitoris, labia, glans and shaft of penis.

**Differential diagnosis**

- Bullous pemphigoid
- Epidermolysis bullosa
- Erythema multiforme
- Linear IgA dermatosis
- Pemphigus
- Desquamative gingivitis

**Investigations**

- Histology, direct immunofluorescence (DIF) and indirect immunofluorescence (IDIF) are used to diagnose CP. However, differentiation from similar blistering diseases (particularly bullous pemphigoid) is on clinical grounds.
- Imaging may be required to investigate the upper airway and oesophagus.

**Associated diseases**

There is an increased risk of malignant disease with CP - for example, T-cell lymphoma.\(^9,10\)

**Management**

Management of CP may involve different modalities of treatment depending on the pattern of involvement. It is a difficult disease to manage. This may involve:

- **Medical management:**\(^{11}\)
  - The aim is to suppress blistering and scarring.
  - Lowest doses of drugs should be used to minimise side-effects. Drugs used to suppress inflammation include:
    - Triamcinolone - used topically for mild gingival and buccal disease.
    - Dapsone - can be helpful in ocular and mucosal disease.
    - Prednisolone - often required with attendant complications.
    - Azathioprine - may be needed to spare steroid use.
    - Other agents (such as cyclophosphamide, ciclosporin, sulfasalazine, etanercept and mycophenolate) - are also used.

- **Surgical management may be required particularly for:**
  - Eye complications.\(^{12}\)
  - Airway problems (particularly tracheostomy).
  - Oesophageal dilatation.

- A co-ordinated team approach. Other referrals are helpful to a variety of specialties determined by the particular pattern of disease and complications:
  - Dietary advice: soft, non-acidic foods, adequate calcium and vitamin D intake.
  - Dental advice for gingival and buccal complications.\(^{13}\)
  - Physiotherapy. This should encourage and facilitate an active and as near-normal lifestyle as possible. Exercise, avoiding trauma, is helpful (typically swimming is recommended).
  - Ophthalmology for eye complications - for example, surgery and contact lenses may be indicated.\(^{14,15}\)
  - Specialist dermatology advice is usually required for what is a rare and difficult disease.
  - ENT referral may be required for diagnosis (endoscopy) and treatment.
  - Gynaecology referral may be needed when there is genital involvement.\(^{16}\)
  - Gastroenterology advice, usually for oesophageal complications.
  - Endocrinology referral may help with advice on prevention of steroid complications.

**Complications**

- Severe sight impairment
- Oesophageal strictures
- Airway obstruction

**Prognosis**
The disease is chronic, progressive and difficult to treat. There may be exacerbations and remissions with waning disease activity. The risk of malignancy, particularly with anti-epiligrin CP, is significant.

Further reading & references

13. Fatahzadeh M, Radfar L, Strois DA; Dental care of patients with autoimmune vesiculobullous diseases: case reports and literature review. Quintessence Int. 2006 Nov

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