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Hidradenitis Suppurativa

Synonym: acne inversa

Hidradenitis suppurativa is a chronic persistent or recurrent suppurative disease of unknown cause occurring in the apocrine follicles, usually affecting the groin and axillae and also other apocrine-bearing sites such as the breasts, perineum and buttocks. Follicular occlusion may lead to chronic relapsing inflammation, mucopurulent discharge and progressive scarring. Induration, ulceration and also sinus and fistula formation may occur [1].

Epidemiology

- The prevalence in Europe has been estimated to be in the region of 1% [2].
- Overall it occurs more often in women. Submammary, axillary and inguinal involvement is more common in females but perineal involvement is more common in men.
- It does not start before puberty. Hormonal factors are thought to play a role. Average age of onset is 21 years [1].
- Hidradenitis suppurativa is more common in white and Afro-Caribbean populations and is rare in people from Asia.

Risk factors [1, 3]

- Cigarette smoking.
- Obesity.
- Lithium therapy.
- Possibly increased in patients with diabetes and polycystic ovary syndrome.
- Hormonal factors: tends to improve during pregnancy and when taking the combined oral contraceptive (COC) pill; tends to relapse after pregnancy and when stopping the COC pill. There may be premenstrual flares and it may remit after menopause.
- Genetic factors: there is a family history in 30%

Presentation

- Usually starts at puberty but can present at any time from puberty until middle age and is variable in severity and distribution.
- Predominantly occurs in the axilla, groin, perineum, buttocks, scrotum and submammary regions. The axilla and groin are the most commonly affected sites.
- Early lesions are solitary, painful and pruritic nodules and multiple sites may be affected at the same time. They may persist for weeks or months. Any subcutaneous extension appears as indurated plaques. Episodes of acute cellulitis may occur.
- The nodules develop into pustules and eventually rupture with discharge of purulent material.
- Healing occurs with dense fibrosis, so scarring is common.
- Recurrences tend to occur in the same region, leading to chronic sinus formation, with intermittent release of serous, purulent, or bloodstained discharge. Sinus formation and rupture may occur internally into adjacent structures as well as externally.
- Regional lymphadenopathy is usually absent.

Differential diagnosis

- Acne.
- Abscess (usually solitary lesions).
- Lymphogranuloma venereum.
- Cutaneous Crohn's disease.
- Furunculosis.
- *Actinomyces* spp.

Investigations

Diagnosis is clinical but investigations may include:

- FBC: underlying anaemia associated with chronic disease.
- Blood glucose: identify associated diabetes.
- Microbiology swabs (usually negative).

Associated diseases

Common comorbidities include [2]:

- Cystic acne

- Pilonidal sinus
- Crohn's disease
- Down's syndrome
- Metabolic syndrome
- Pyoderma gangrenosum
- Spondyloarthropathy

Staging

Hidradenitis suppurativa can be divided into the following three clinical stages (Hurley's classification)[1]:

- Stage I: abscess formation, single or multiple without sinus tracts and scarring.
- Stage II: recurrent abscesses with sinus tracts and scarring; single or multiple widely separated lesions.
- Stage III: diffuse or almost diffuse involvement or multiple interconnected tracts and abscesses.

Other scoring and staging systems exist which may be used to monitor interventions in clinical trials[2]:

- Sartorius score.
- Physician Global Assessment (PGA) score.
- Hidradenitis Suppurativa Severity Index (HSSI).

Management[2, 3, 4]

Early lesions are usually treated by medical therapy; however, long-standing, severe disease usually requires surgery. Dermatology referral should be considered for patients with exacerbations that are frequent enough to cause them distress despite treatment and those with active disease and scarring in at least one site (Hurley stage II disease)[1].

General and lifestyle

- Advice should include good hygiene (using non-soap or antiseptic cleansers, and non-fragranced antiperspirants) and wearing loose-fitting clothing. People with hidradenitis suppurativa should be advised to take measures to avoid heat, humidity, sweating, trauma, friction and irritation.
- Diet: advise weight reduction if appropriate, as there is a strong association with being overweight or obese. Dietary associations with the condition have been proposed, with some finding low glycaemic index and/or dairy-free diets helpful; however, the link has not yet been well established and research is ongoing[5, 6].
- Smoking cessation.
- Assess psychological impact: poor self-image and depression.

Pharmacological

Acute stage treatment for large painful lesions includes intralesional or oral corticosteroid, although these are contra-indicated if infection is suspected, in which case treatment would be a course of systemic antibiotic. For chronic and relapsing stages, the following are evidence-based treatment options:

- Topical antibiotic. Clindamycin 2% used topically twice daily for three months is first-line for mild disease (Hurley stage I or mild stage II).
- Systemic antibiotic. Options used include lymecycline 408 mg od, tetracycline 500 mg bd, doxycycline 100 mg od, or erythromycin 500 mg bd. Where these are not effective after three months, clindamycin 300 mg bd in combination with rifampicin 600 mg od or 300 mg bd may be used.
- Anti-TNF therapies have been shown to be effective; adalimumab has recently been approved by the National Institute for Health and Care Excellence (NICE) for treating moderate to severe hidradenitis suppurativa[7, 8]. Infliximab has also shown to be effective but is not currently approved for this indication in the UK.

Other medical therapies which have been used include:

- Anti-androgens. Dianette® is often used in women, particularly where a hormonally related cycle of flare-ups has been noted. Other COC pills, particularly those with an anti-androgenic progesterone such as drospirenone, are used.
- Dapsone.
- Retinoids (isotretinoin and acitretin).
- Ciclosporin.

Surgical[1]

- Only surgery can remove scarred areas. Surgery should be performed during a quiescent phase.
- Incision and drainage of painful boils is not effective, as over half recur. Medical therapy is preferable.
- Radical excision of the apocrine glands may produce temporary relief of symptoms but the disease process may recur in residual glands in the surrounding area. Recurrence is more common in inadequate excision, obesity, continued smoking and locally macerated skin.
- Surgery may be extremely complex for extensive disease, with large areas of skin needing to be removed and problems with skin closure. Skin grafting or rotation flaps may be required. Infections and delayed healing are common.
- Deroofing and skin-tissue-saving excision with electrosurgical peeling (STEEP) offers a more limited surgical option.
- Carbon dioxide laser and Nd:YAG laser are also options which have been studied with some success, although a Cochrane review concluded more controlled trials were needed, as for most treatments used for this condition[8].

Complications[2]

- Fistula formation into the urethra, bladder, rectum, or peritoneum may occur but is uncommon.
- Chronic infection may lead to anaemia, hypoproteinaemia or, rarely, amyloidosis.
- Chronic malaise, reduced quality of life, depression and poor sexual health[9, 10].
- Scarring may lead to lymphatic obstruction and lymphoedema.
- A number of arthropathies may occur in long-standing disease.
- Squamous cell carcinoma may develop in areas of chronic lesions.

Prognosis

- Variable but without treatment tends to be a relentless progressive disease with acute exacerbations and remissions, leading to sinus tract formation and scarring.
- Recurrence rate tends to be high with many treatment options. The quality of data on recurrence after wide excision is poor but some evidence suggests lower recurrence rates with wide excision where skin flaps or skin grafts are used as the closure methods[11]. A Cochrane review concluded more evidence was needed to make conclusions upon benefits of most treatment options[8].

Further reading & references

- Hidradenitis suppurativa; DermIS (Dermatology Information System)
- Hidradenitis suppurativa; DermNet New Zealand

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