Managing patients with hidradenitis suppurativa

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Managing patients with hidradenitis suppurativa

How do patients present in primary care?

What are the potential complications?

What are the evidence-based management options?

Hidradenitis suppurativa

Hidradenitis suppurativa is a painful inflammatory condition affecting apocrine-bearing areas of the skin. It is primarily a disease of follicular occlusion, associated with an autosomal dominant mode of inheritance.

Epidemiological studies conducted in the USA, Denmark and France give an average prevalence of 1%. Women appear to be more than twice as likely to be affected as men. Historically, hidradenitis suppurativa has been the subject of relatively little research, despite the fact that it is common and often significantly impacts on quality of life.

‘Pain is a significant complication as is the psychosocial impact’

Presentation

Hidradenitis suppurativa forms part of a tetrad of follicular occlusion, the other components are acne conglobata, pilonidal disease and dissecting cellulitis. The onset of hidradenitis suppurativa is variable but usually occurs in the second and third decades of life, coinciding with development of the apocrine glands. Hence it is rarely seen in pre-pubertal children.

The most commonly involved anatomical sites are the intertriginous...
(inverse) areas of skin such as the axillae (see figure 1, p25 and figures 5 & 6, p27), groin (see figure 3, p27), and inframammary regions, although the buttocks (see figure 2, below), nuchal scalp (see figure 4, p27) and retroauricular areas are notable atypical sites, more commonly seen in men.6,7

Hidradenitis suppurativa varies in clinical presentation and severity, and is often associated with significant pain and morbidity. The condition is characterised by painful, inflammatory papules and nodules which frequently progress to form abscesses, sinus tracts and hypertrophic scars. A useful physical sign is double-headed comedones. Pressure on inflammatory lesions may lead to serosanguinous or purulent discharge. Postinflammatory scarring is frequently hypertrophic or rope like, and multiple interconnected sinus tracts may lead to honeycomb patterning.6,8

The complications of hidradenitis suppurativa include lymphatic obstruction or contractures from scarring, sacral osteomyelitis, anaemia and formation of pyogenic granulomas. Squamous cell carcinoma is another well recognised but rare risk in these patients, and may present difficulties in diagnosis if mistaken for inflammatory lesions.8,9

Pain is a significant complication as is the psychosocial impact. Patients with hidradenitis suppurativa have a mean score on the Dermatology Quality of Life Index that is higher than other skin conditions associated with high morbidity including eczema, acne, psoriasis and even chronic urticaria.10

PATHOGENESIS
The primary event in the pathogenesis of hidradenitis suppurativa appears to be occlusion of the apocrine glands caused by epidermal hyperplasia. Subsequent follicular rupture leads to inflammation and abscess formation with consequent scarring.1,11 The ruptured follicles may reform to create the sinus tracts and fistulas seen in the disease.1,11

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‘There appears to be a correlation between smoking and more severe disease’

A number of different factors have been implicated in the aetiology of hidradenitis suppurativa, including genetics, obesity and smoking. Recent studies have found mutations in the gamma-secretase genes, which could possibly contribute to the follicular occlusion pathway.12,13 Metabolic syndrome is significantly more common in hidradenitis suppurativa patients than those without the condition (P < 0.001), and obesity is also well recognised as an independent factor. Almost half of hidradenitis suppurativa patients are overweight.14,15

The effects of weight on pathogenesis could relate to increased frictional forces and a low-level pro-inflammatory effect.15,16 The prevalence of smokers within the hidradenitis suppurativa...
population is significantly higher than in the normal population. A recent Dutch study found that not only were 71% of hidradenitis suppurativa patients current smokers, but there appears to be a correlation between smoking and more severe disease.17

Another study reported a lower remission rate in those who were current smokers.18 The mechanism of smoking on the pathogenesis of hidradenitis suppurativa may be associated with nicotine causing epidermal hyperplasia and subsequent follicular occlusion.19

Bacteria are not felt to have a primary role in lesion formation, and abscesses are often sterile. No obvious relationship between hidradenitis suppurativa and hormonal factors has been established.20,21

CONFIRMING DIAGNOSIS
A European guideline published in 2015 gives primary and secondary diagnostic criteria, see table 1, opposite.22

The Hurley staging system provides a classification of severity, see table 2, opposite.23

The diagnosis of hidradenitis suppurativa is clinically based, without a specific diagnostic test. In primary care it is prudent to take a bacterial swab from any discharging abscesses to exclude superadded infection or colonisation.24 Blood tests are helpful to screen for underlying disease such as metabolic syndrome or anaemia of chronic disease. Skin biopsy is not routinely required.

Risk factors and associated conditions
The most important non-genetic factors implicated in hidradenitis suppurativa are obesity and smoking.

There is also a possible link between hidradenitis suppurativa and inflammatory bowel disease, though cutaneous Crohn’s disease is also a common differential diagnosis. A Dutch study found that 17% of Crohn’s patients and 14% of patients with ulcerative

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The first instance, the next treatment option is oral antibiotics, initially, a tetracycline, such as lymecycline for four months.

More potent treatment options for recalcitrant disease include combination therapy of oral clindamycin (300 mg twice daily), and rifampicin (300 mg twice daily for three months), which leads to an improvement in about 80% of cases.27, 28 If this proves ineffective, retinoids are often used. The evidence shows that isotretinoin is less effective than acitretin. However, because of the ongoing teratogenic risk with acitretin (a woman should not conceive for two years after cessation of the drug), isotretinoin is often favoured in younger women.29

The advent of biological therapies has significantly improved the outcome for hidradenitis suppurativa patients. A systematic review in 2012 found that 89% of hidradenitis suppurativa patients treated with infliximab had a moderate or significant response, as did 79% of patients treated with adalimumab.29

In May 2016 NICE published final draft guidance on the use of adalimumab with the recommendation that the drug is used for people with active moderate to severe hidradenitis whose disease has not responded to conventional systemic therapy. The drug is recommended only if the company provides it at the price agreed in the patient access scheme. Final guidance is expected to be published in June 2016.30

Surgical management ranges from simple incision and drainage of single abscesses to deroofing of sinus tracts with secondary intention healing, through to extensive operations such as radical wide excisions. The latter is thought to be the only definitive curative treatment and can involve removal of an entire anatomical area of apocrine-bearing skin, with repair via skin grafting or flaps. Ablative laser treatments are also sometimes used.

REFERRAL
Patients with Hurley stage I disease can be effectively managed in primary care, with lifestyle advice, topical clindamycin, and oral lymecycline should lesions persist. Beyond this, referral to secondary care is appropriate. Should the patient have more severe disease at first presentation, rapid referral and prescription of tetracycline antibiotics pending the dermatology appointment would be advocated.

CONCLUSION
Hidradenitis suppurativa is a painful, debilitating and under-researched condition that has a high psychological and physical burden on the people it affects. Its pathogenesis is complex and multifactorial and there is a comparative lack of evidence-based treatments.
Hidradenitis suppurativa affects the apocrine-bearing areas of the skin. The onset is variable but usually occurs in the second and third decades of life, coinciding with development of the apocrine glands.

The condition is characterised by painful, inflammatory papules and nodules which frequently progress to form abscesses, sinuses and hypertrophic scars. The complications of hidradenitis suppurativa include lymphatic obstruction or contractures from scarring, sacral osteomyelitis, anaemia and formation of pyogenic granulomas. Squamous cell carcinoma is another well recognised but rare risk in these patients.

A number of different factors have been implicated in the aetiology of hidradenitis suppurativa, including genetics, obesity and smoking. Recent studies have found mutations in the gamma-secretase genes, which could possibly contribute to the follicular occlusion pathway. Metabolic syndrome is significantly more common in hidradenitis suppurativa patients than those without the condition (P < 0.001), and obesity is also well recognised as an independent factor.

Bacteria are not felt to have a primary role in lesion formation, and abscesses are often sterile. The diagnosis of hidradenitis suppurativa is clinically based, without a specific diagnostic test. The most important non-genetic factors implicated in hidradenitis suppurativa are obesity and smoking.

Locally recurring lesions can be treated surgically and more widespread disease may be better managed with a combination of medical treatment and surgery. However, first-line treatment in the form of topical clindamycin 1% is first-choice for mild disease. The next treatment option is oral antibiotics, initially, a tetracycline, such as lymecycline for four months. More potent treatment options for recalcitrant disease include combination therapy of oral clindamycin 300 mg twice daily, and rifampicin (300 mg twice daily for three months), which leads to an improvement in about 80% of cases. If this proves ineffective, retinoids are often used.

The advent of biological therapies has significantly improved the outcome for hidradenitis suppurativa patients. A systematic review in 2012 found that 89% of hidradenitis suppurativa patients treated with infliximab had a moderate or significant response, as did 79% of patients treated with adalimumab. Hidradenitis suppurativa is a painful, debilitating and under-researched condition that has a high psychological and physical burden on the people it affects.

Available, particularly in the form of randomised placebo-controlled trials. However, the European guidelines have provided a helpful treatment pathway, with evaluation of all current available research.

Taking a holistic approach when assessing and treating these patients is vital, and referral to dermatology services is important should simple measures fail to be effective.

**REFERENCES**


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