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Hidradenitis suppurativa

Jan von der Werth
MRCP Consultant Dermatologist
Lincoln County Hospital

Hidradenitis suppurativa (HS) was first described by the French physician Velpeau in 1839. Some patients may feel that over the past 160 years we have not made much progress in our attempts to successfully treat this condition.

Is this so? Have we really failed to advance significantly on this disease despite our powerful array of modern medicaments and surgical techniques? I guess in the eyes of many who suffer from HS the answer would be: yes. Hands up all those who feel comfortable when confronted with a hidradenitis patient. So, what is the cause for this lack of progress and do we really have so little to offer for this condition?

A history of confusion

First of all, there was the 'apocrine misunderstanding'. In 1833, six years before the first clinical description of hidradenitis, Purkinje discovered sweat glands in human skin. Twelve years later, in 1845, Robin followed with the first description of the structure and location of apocrine glands, so that in 1854 another Frenchman, Verneuil, having studied Velpeau's work, became the first person to relate the disease he termed 'hydrosadenite phlegmoneuse' to apocrine glands. He went on to publish four papers on this condition in which he confirmed his belief that hidradenitis represented a disease of apocrine glands, a view that became generally accepted with time.

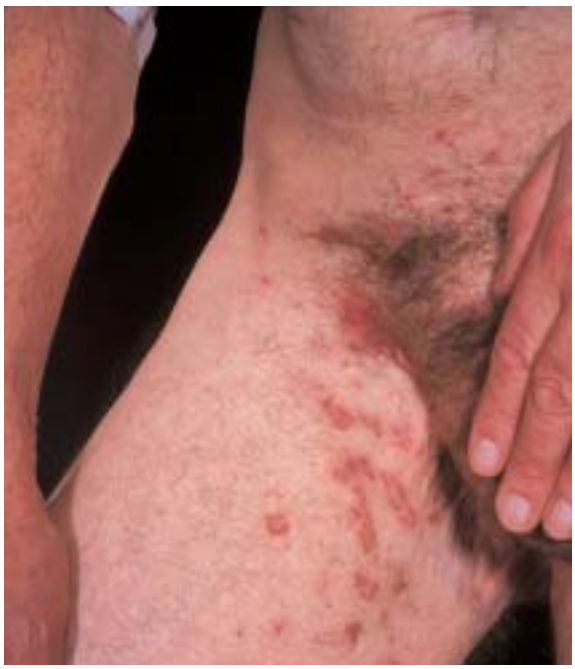

Figure 1. *The typical lesion is a painful inflammatory papule, nodule or abscess*

It took a series of detailed histopathological studies in the 1980s and 1990s to prove that hidradenitis is primarily an acne-like disease of follicular occlusion where apocrine glands only become involved in the context of an intense perifollicular inflammation. The initial process was found to be one of cornification of the follicular infundibulum, followed by an early inflammatory reaction and ultimately the rupture of the involved hair follicle. This leads on to the formation of abscesses which can either subside spontaneously over a matter of days or which occasionally progress into a chronic process of cutaneous or subcutaneous tunnelling, referred to as sinus tract formation.[1–4]

Knowledge of this process has given us a better understanding of the disease and has helped to point us towards treatment attempts resembling those successfully used in acne vulgaris. Before these studies had emerged many hidradenitis patients had received treatments for presumed apocrine gland infections – with virtually no benefit at all.

Patients in hiding

A further reason for our lack of progress rests with the patients themselves. Although it has never been scientifically studied or quantified, any researcher in the field will confirm the reluctance of patients with hidradenitis to come forward with their disease. Patients frequently conceal their condition even from their closest relatives. Such seclusion is openly acknowledged by the international hidradenitis self-help group which has adopted the name HIDE (Hidradenitis Information Development and Exchange) in reference to this behaviour. The patients' evasiveness coupled with the lack of diagnostic laboratory tests have probably led to significant underreporting of HS, betraying its true significance and condemning it to a fringe existence among over 1,000 other dermatological conditions.

Hidradenitis suppurativa and quality of life

Three studies have put the disease prevalence at between 1:100 and 1:600.[5–7] Even the conservatively estimated prevalence figure of 1:500 would translate into approximately 100,000 hidradenitis sufferers in the UK alone. What makes this number more significant is the high degree of dis-

ability suffered by the average hidradenitis patient. A recent study found the impairment of life quality as measured through the Dermatology Life Quality Index (DLQI)[8] – a widely used dermatological quality-of-life inventory – to be higher in hidradenitis than that seen in a variety of other skin diseases, including acne, eczema or psoriasis.[9] An internet survey of patients visiting the Dermatology Online Atlas of the University of Erlangen (www.dermis.net/doia/) came to a similar result. From over 10,000 patients with more than 600 different skin diseases who returned an online DLQI questionnaire, hidradenitis sufferers were found to have the highest levels of disability.[10]

These figures should suffice to place the disease onto the front row of national and international dermatological research. Unfortunately, in reality only a trickle of new work on HS emerges in the dermatological literature from time to time. This is only likely to change when patients can be encouraged to come forward with their complaints more frequently.

Clinical features

Still, over the past four decades, a number of dedicated authors have given us a reasonably clear picture of the most common aspects of the disease. Hidradenitis begins at an average age of about 23 years and affects women two to five times more often than men. It most commonly erupts in the groins and axillae, less often in the perineum, the perianal region, suprapubic region, inframammary region or in the abdominal folds. In severe cases lesions can erupt in any hair-bearing part of the body. The typical lesion is a painful inflammatory papule, nodule or abscess (Figure 1). Occasionally, comedones (Figure 2) or dermal contractures, which lead to rope-like elevations of the skin, (Figure 3) can occur.

As many as one-third of all boils remain blind, failing to burst or discharge. In mild cases, boils remain painful for only two to three days. However, on average, boils stay tender for about one week. Verneuil himself observed that boils often took two weeks to subside.

In advanced disease, as many as two-thirds of all patients have one or more chronic lesions, which never completely settle. In these cases the abscess has led to the development of intradermal or subcutaneous epithelium-lined sinus tracts, which remain a constant source of inflammatory activity. A recent survey of HS patients seen in secondary care showed that patients suffered an average of 4.8 boils each month and continued to have active disease an average of almost 20 years after the onset of disease.[11]

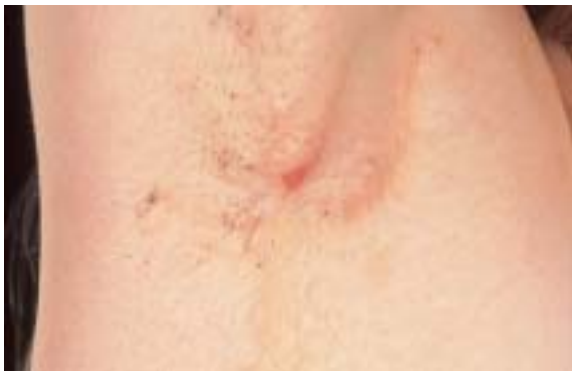

Figure 2. *Occasionally, comedones can occur*

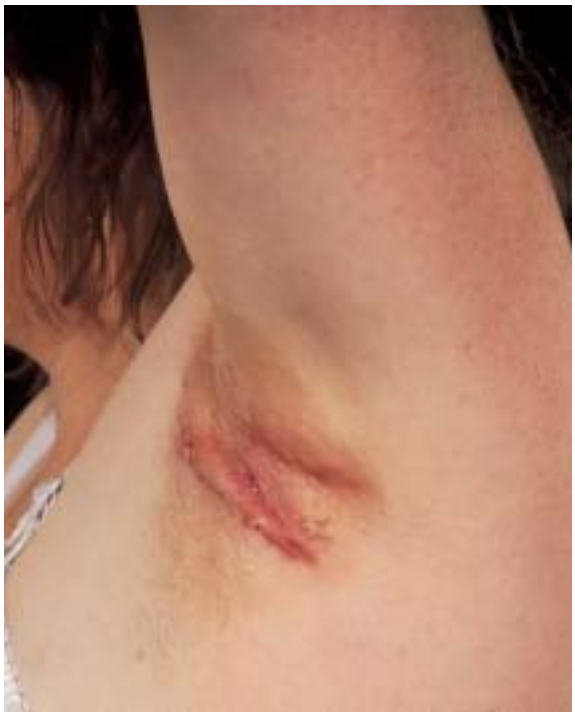

Figure 3. *Dermal contractures can occur which lead to rope-like elevations of the skin*

Hidradenitis can be aggravated through stress, heat, sweating or friction. Approximately 50% of female patients report a disease flare before or during menstruation. Smoking may be a further trigger for the disease. Two studies have observed the number of smokers among hidradenitis sufferers to exceed 70%, compared with about 30% in the normal UK population.[12,13]

In 1985 Fitzsimmons *et al* reported a familial form of hidradenitis and proposed an autosomal dominant pattern of inheritance in affected families.[14] These findings were recently confirmed[15] and have led to the commencement of the search for the underlying genetic abnormality of hidradenitis. At the moment these studies are ongoing.

The course of hidradenitis is usually a chronic one. However, there is some evidence that in women the disease eases or may even subside following the menopause, which supports the view that hormonal influences are important in its pathogenesis. The precise role of hormones in the aetiology of HS remains unclear, and this also applies to the role of microbes.

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How to diagnose hidradenitis suppurativa

To diagnose hidradenitis one first and foremost has to think of it. The key feature of the disease is the occurrence of recurrent boils in flexural body sites, particularly the groins, axillae and perianal regions. The main differential diagnosis is that of bacterial abscesses. These, however, will hardly ever recur repeatedly in the regions affected by HS. Leach *et al* found that among 57 patients with axillary abscesses, only patients with hidradenitis developed recurrent lesions.[16]

A definition in which the occurrence of five or more boils either in the axillae or groins was tantamount to the diagnosis of hidradenitis suppurativa was used by the author in a recent study of the clinical genetics of the disease.[15] An excellent correlation between the diagnoses based on this definition and an earlier clinical diagnosis made by an experienced dermatologist was observed.

Thus, recurrent painful inflammatory eruptions in the groins or axillae should be considered to be hidradenitis until proven otherwise. In addition the presence of comedones – particularly double-ended comedones – or dermal contractures should also point the clinician to the existence of this disease.

Treatment

The difficult subject of treatment for HS becomes easier if one separates it into two categories:

- The treatment of acute flare-ups or very painful individual boils
- The long-term management of chronic disease.

Treatment of acute flare-ups

For acute painful boils the swiftest response is achieved with systemic corticosteroids. Prednisolone 30 mg for three or four days can provide quick and often profound relief of the symptoms. Where bacterial superinfection is contributing to the problem, systemic antibiotics such as clindamycin or a macrolide antibiotic like clarithromycin should be added. In addition, ordinary analgesics will usually be required. Many patients find a hot bath or flannel helpful and patients may need a few days off work to aid their recovery. Another option is the injection of intralesional corticosteroid, such as 1% triamcinolone.

Unfortunately, the treatments most commonly used today are the least effective and can be positively harmful. Incising a non-fluctuant boil is only going to make matters worse and should be avoided. Excising an acute inflammatory lesion and closing the resulting wound primarily is also wrong, as there is a very high rate of, often immediate, recurrences in such cases (approximately 80% recurrences).[17,18] When excision is performed, healing should be by secondary intention. Another frequent mistake is to prescribe penicillins or short courses of other antibiotics. Virtually every patient with at least moderately severe HS will testify to the fact that short courses of antibiotics on their own are unhelpful. What is worse is that many patients who are prescribed such treatments feel 'fobbed off' with less than useful medication and ultimately end up turning their backs on the medical profession. Such a step is crucial as many of these patients feel unable to confide their problems to anyone other than a general practitioner or specialist.

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Long-term management

The long-term management of HS patients is a particular challenge. No universally effective treatment for this disease is available. For that reason many papers written on this subject plead for early and radical surgery to prevent the disease from showing the relentless progression often attributed to it. I would like to argue with this view. Some medical treatments are undoubtedly effective and should be explored before the decision to proceed to surgery is made. In fact, a close co-operation between medical and surgical teams will greatly benefit most patients. In addition, in my experience, patients do by no means always relentlessly progress in their disease.

The decision to embark on long-term treatment should be discussed with the patient in some detail. Factors that need to be considered are the disease activity, the impact of the disease on the patient's life, the patient's family planning and the potential drug side-effects.

Treatment options include systemic tetracyclines or macrolide antibiotics in doses similar to those given for acne. The response to treatment needs to be monitored, for which a simple lesion-count estimated by the patient is a useful measure. An alternative is treatment with an antiandrogenic hormonal preparation like Dianette® (Schering Health, UK). Occasionally, the combination of this with an antibiotic may be beneficial. One of the most effective medical treatments for HS is unfortunately hazardous for the majority of HS sufferers due to its teratogenicity. Acitretin has been shown to lead to sometimes dramatic responses in a number of patients,[19] but it cannot be prescribed to many of the young female HS sufferers who make up the largest group among

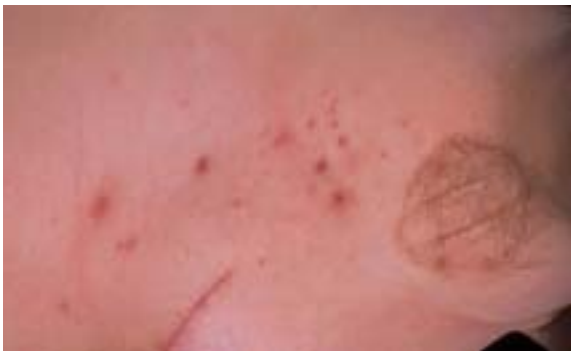

Figure 4. *Surgery can never be curative, as the disease may erupt outside the operation area at a later stage*

patients.

A variety of other drugs have been tried but whether their effects are more than just placebo remains unproven. Interestingly, the use of Roaccutane® (Roche, UK) is usually ineffective in patients with HS, although it may be helpful for those who have coexisting acne.

Surgery

When medical treatments fail, surgery remains the only alternative. However, there appears to be a right way and a wrong way of performing surgery for HS.

As mentioned above, the wrong way is by incision of an acute, non-fluctuant abscess or by excision of a lesion with subsequent primary closure. The right way is with wide excision and subsequent secondary healing or split-skin grafting.[20] The successful principle seems to be to scar the involved areas. As scars are free from hairs and glands the disease is deprived of its core structures. Scarring can also be achieved with aggressive cryotherapy or laser surgery. However, cryotherapy should be limited to areas of disease that are chronically inflamed. It should be avoided for the self-limiting boils that make up the majority of HS lesions. It should always be remembered that surgery – even if extensive – can never be curative, as the disease may erupt outside the operation area at a later stage (Figure 4).

Conclusion

Combinations of the measures discussed here will help to ease the suffering of most patients with this disabling disease. When used appropriately they will allow the clinician to prove that progress has been made in the management of hidradenitis within the past 160 years. There can be no doubt, though, that we have a long way to go and, in the meantime, we should not forget that patients also need our understanding and empathy. For this to be possible all of us need to appreciate the high degree of disability that HS causes for many patients all over the world ■

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Key points

- **Hidradenitis is a common disease with probably over 100,000 sufferers in the UK alone.**
- **Hidradenitis causes a high degree of disability, which is often concealed by patients.**
- **The cause of hidradenitis remains unknown but the pathogenesis resembles that of acne and puts the disorder among the follicular occlusion diseases. Hidradenitis is not a disease of apocrine glands.**
- **Recurrent boils in axillae or groins should be regarded as hidradenitis until proven otherwise.**
- **Acute flare-ups can be treated with a very short course of systemic corticosteroids or intralesional steroids. Short courses of antibiotics are generally useless in long-standing HS.**
- **Long-term management is difficult. Acitretin works well but needs to be prescribed with great care, particularly in young women.**
- **There is a right way and a wrong way of performing surgery for hidradenitis patients. The right way is by wide excision with secondary intention healing or split-skin grafting. The wrong way is by incision or narrow excision with primary closure.**