Therapeutic options for the follicular occlusion tetrad.

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Acne conglobata, hidradenitis suppurativa, dissecting cellulitis or folliculitis decalvans, and pilonidal sinus have in common the occlusion of sebaceous or apocrine glands, with subsequent inflammation and scarring. These disorders may be seen in isolation or in various combinations, and their treatment is often challenging for both the patient and the physician. Medical and surgical therapies for each disease are reviewed with an emphasis on recent developments and trends in the care of patients.

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Introduction

The follicular occlusion tetrad comprises four clinical entities: dissecting cellulitis of the scalp, acne conglobata, hidradenitis suppurativa, and pilonidal sinus. The similarity of their etiopathogeneses was first suggested by Goeckerman [1], and Brunsting [2] described the anatomic, pathophysiologic, and clinical similarities of the first three disorders (the follicular occlusion triad), stating that they appeared to be fulminant forms of acne vulgaris. He pointed out that they had features in common, including 1) glandular hyperplasia of the pilosebaceous apparatus or apocrine gland with hyperactivity; 2) follicular orifice occlusion and double comedo formation; 3) bacterial invasion with suppuration and undermining of loose areolar tissue; and 4) cicatricial healing (Figure 1). The subsequent inclusion of pilonidal sinus led to the designation follicular occlusion tetrad [3]. The clinical characteristics of these four entities have been previously well described and are outlined in Table 1.

Treatment options have traditionally involved the use of topical and systemic antibiotics and retinoids, either alone or in concert with surgical therapy. Other less commonly employed treatment modalities such as orthovoltage radiation, ultraviolet irradiation, and hormonal manipulation have also been used, but with varying degrees of success.

With so many options available, no one modality has demonstrated clear superiority. This article attempts to summarize the current trends, both medical and surgical, in the approach to these rather difficult-to-treat disorders.

Folliculitis decalvans or dissecting cellulitis of the scalp

Folliculitis decalvans, or dissecting cellulitis, is an unusual, chronic, and progressive purulent folliculitis that leads to follicular atrophy, alopecia [4,5], and scarring. It occurs primarily in adult men and African Americans, and it initially affects the vertex and occiput of the scalp [6]. Cystic papules or discrete abscesses are less common than a diffuse phlegmon or abscess that forms from the coalescence of multiple nodules. Sinuses discharging seropurulent material may be found at the base of individual nodules or within collections of nodules. Alopecia occurs over the nodular swellings, and following “burnout” of the disease, atrophic scarring and convoluted ridges can be seen. The latter is similar in appearance to cutis verticis gyrata.

The treatment of folliculitis decalvans or dissecting cellulitis has encompassed radiation therapy, incision and drainage, excision and grafting, antibiotic use, and cauterization. The acute suppurative phase is often treated with topical and systemic antibiotics and local cleansing, but this practice may not halt the progression of the disease process.

Efforts have recently focused on retinoids and anti-inflammatory agents in an attempt to induce longer lasting remissions. A good response to isotretinoin (0.5 to 2 mg/kg/d) alone has been recently reported [7-9]; however, the long-term clinical response needs to be confirmed.

Both the combination of isotretinoin with radical excision and tissue expansion [10], and the combination of isotretinoin with intralional triamcinolone acetate (40 to 80 mg) [11], have also been recommended as therapeutic options in dissecting folliculitis. The anti-inflammatory effects of isotretinoin have been touted as being of benefit in the treatment of this disorder. Oral zinc therapy (400 mg/d), either alone [12] or in combination with oral (1500 mg/d) and topical (1.5%) fusidic acid [13], has also been reported as a therapeutic option with good results observed after follow-ups of up to 2 years. Zinc presumably has an anti-inflammatory effect, whereas fusidic acid is an effective antimicrobial.

Surgical excision with or without grafting is reserved for recalcitrant cases that are unresponsive to medical management. Reconstructive options for the resultant alopecia are difficult if the disease process remains active. Hair transplantation, grafting, or local flap operations, with or without tissue expansion, and fractional excision are available if the chronic, progressive nature of the disease can be suppressed.

Acne conglobata

Spitzer [14] is credited with giving the first description of acne conglobata, and other authors have added to its clinical [15,16] and histologic description [17,18]. Histologically, involvement of the pilosebaceous apparatus is observed, differentiating acne conglobata from hidradenitis suppurativa. In addition, it was noted that all hair-bearing areas, including the entire trunk, the buttocks, and the extremities, were susceptible to this eruption. Florid acne conglobata does not occur until the late teens or early twenties, and it is almost exclusively limited to men. The age of onset has been attributed to a postadolescent functional hyperplasia of sebaceous glands incited by hormonal influences.
Tropical acne, described by Sulzberger et al. during World War II in military personnel stationed in the Pacific, closely resembles acne conglobata. However, its rapid onset and resolution, dependent on tropical climatic exposure, led Novy and others to disclaim it as a variant of acne conglobata.

The pathogenesis of acne conglobata is similar to that of hidradenitis suppurativa, except that in the former follicular occlusion causes rupture of the pilosebaceous unit. In acne conglobata, in contrast to hidradenitis suppurativa, single comedones, inflammatory papules, and cystic lesions are frequently found. However, double comedones can occur, and clusters of 10 to 20 or more comedones may be observed. Abscesses, communicating fistulas, and discharging sinuses are also characteristic findings. Brunsting stated that acne conglobata, more than hidradenitis and dissecting cellulitis, resembled a severe form of acne with superimposed pyogenic infection. He regarded hidradenitis and dissecting cellulitis as primarily pyogenic diseases that have little resemblance to acne and occur in structurally predisposed anatomic areas.

The association of acne conglobata, as well as the other members of the follicular occlusion tetrad, with musculoskeletal manifestations such as arthritis, myalgias, muscle weakness, and enthesopathies has led investigators to propose a genetic or an immune system basis for the disorder. Surgical treatment of acne conglobata is not practical as the sole modality because of the widespread distribution of involved skin. Medical management has been used more frequently for this reason. The results were generally poor until the recent release and use of isotretinoin (Accutane; Hoffmann-LaRoche, Nutley, NJ). This agent is presumed to have a direct inhibitory effect on the sebaceous glands. The side effects are common and vary from dryness of the skin and mucous membranes to osteophyte formation. When medical management fails, surgical extirpation has provided significant alleviation of problematic regions. Skin grafting has been the most common method for resurfacing these areas. Finding donor sites free of disease can, however, be difficult.

**Hidradenitis suppurativa**

Hidradenitis suppurativa is believed to be caused by the occlusion of the follicular orifice of an overactive apocrine gland, resulting in periglandular inflammation. When two adjacent follicles are obstructed and their respective glands coalesce, a double comedo is formed. Retrograde infection produces suppuration, which invades the loose subcutaneous areolar plane, producing a phelegmon. Healing occurs with substantial scarring.

The axillary, inguinal, and anogenital regions are the primary sites of involvement. There is a slight female predominance, and the disease process does not occur before puberty and is rare in the postmenopausal patient. An increased incidence is observed in obese patients and those with a family history. Recently, Chlamydia trachomatis has been implicated in perineal hidradenitis suppurativa.

Bacterial invasion of the obstructed gland is common and must be treated appropriately with local care and systemic antibiotic therapy. The mildest forms of the disease process are often palliated with local care, adequate hygiene, and avoidance of irritating agents. Retinoid therapy, either alone or in combination with erthyromycin (1 g/d), has shown promise in moderate cases of hidradenitis. Controlled studies are needed to confirm the effectiveness of these agents, however.

The recent work implicating hidradenitis suppurativa as an androgen-dependent disorder has led to attempts at hormonal therapy. Treatment using gonadotropin-releasing hormone agonists in combination with dexamethasone or total abdominal hysterectomy and bilateral salpingo-oophorectomy have been shown to eradicate persistent hidradenitis suppurativa in case reports. The theoretical basis for these treatments as well as others for antiandrogens, eg, flutamide, makes this an interesting future field of investigation.

Surgical excision of established hidradenitis suppurativa, in conjunction with local medical therapy, remains a mainstay of therapy. Various surgical techniques have been employed for treating hidradenitis suppurativa. The complete excision of all involved skin and tissues is necessary to ensure eradication of the condition. A comparison of techniques is difficult because of the variations in the methods of excision and reconstruction used by different authors. Banerjee however, attempted to compare various methods of excision and reconstruction. It appears from his data that inadequate excision is the main reason for recurrence and that radical excision with healing by secondary intention is the best treatment in most cases.

Skin graft failure remains significant in the convoluted areas involved with hidradenitis suppurativa, and in many instances the use of skin grafts adds little benefit over healing by secondary intention. Our experience has been that patients who have skin grafting heal more quickly and comfortably than those who do not. If purulent discharge is copious, a two-stage procedure can be done, with excision followed by the use of local dressings. When the infection has resolved, grafting can be performed. The time interval is usually 5 to 10 days after excision. Advancement, local, and fasciocutaneous flap coverage are alternative options for difficult cases and produce excellent results in the proper hands.

**Pilonidal sinus**

Pilonidal sinus is a common disease process of young adults in which persistence and recurrence are significant problems. Pilonidal sinus is now thought to be due to an acquired hair-containing deformity, as seen in Figure 2. The etiopathogenesis of pilonidal sinus is therefore somewhat different from that of the other disorders of this tetrad. Pilonidal sinuses usually present from the time of puberty (when androgens begin to act on the pilosebaceous glands) to middle age. Cavitities become secondarily infected; this process may result in the formation of multiple subcutaneous sinus tracts, which can require immediate drainage.

Therapeutic options for pilonidal sinuses remain mostly in the surgical realm. Any therapy that does not include excision of the involved areas results in a very high recurrence rate. A recent comparison of open versus closed treatment of chronic pilonidal sinuses supported excision and primary closure, citing the lower morbidity and improved cost-effectiveness of this method. The economic benefit of primary closure was confirmed by Khawaja et al. in their prospective study of the economic and medical outcomes with primary closure. The authors cited a lower complication rate and a more rapid return to normal activities in the group with primary closure. A follow-up longer than the 1-year follow-up provided in this study is necessary to evaluate recurrence rates fully.

Advancement flap operations, which address and eliminate the causative process of hair insertion, have also shown excellent results. Karydakis' series of 6545 cases, which had 95% follow-up rate and only a 1% recurrence rate, is to be commended. The concept of addressing the underlying cause rather than just treating the problem is the basis for this preferred treatment modality and success. In our hands, complete excision with Z-plasty closure to eliminate the deep cleft has provided the most successful result in eradicating disease and minimizing recurrence.

**Conclusions**

The disorders of the follicular occlusion tetrad-folliculitis decalvans or dissecting cellulitis, acne conglobata, hidradenitis suppurativa, and pilonidal sinus—may be seen singly or in various combinations. The medical and surgical options for these disorders remain rather limited in their effectiveness. Aside from the treatment of secondary infections with topical and systemic antibiotics, the use of isotretinoin and judicious surgical intervention remain the mainstays of treatment. Recent
hormonal and immunologic studies may contribute to the management of these disorders.

Acknowledgments

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References and recommended reading

Papers of particular interest, published within the annual period of review, have been highlighted as:

- of special interest.
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A good overview of the follicular occlusion triad of hidradenitis suppurativa, acne, and dissecting cellulitis of the scalp and of the relationship of these conditions to musculoskeletal manifestations. Information on the treatment course, prognosis, and evaluation is included. The etiology is also discussed.


Ornsby OS, Montgomery H: Acne. *Disease of the Skin, 6* Philadelphia, Lea & Febiger 1943 1216-1233


The authors follow up seven consecutive patients who have perineal hidradenitis suppurativa and compare them with 10 control patients who have cryptogenic perianal abscesses. They show that six of seven in the hidradenitis group and none in the control group demonstrate IgA antibodies to *C. trachomatis*. A possible link, but not proof of *Chlamydia*’s role in hidradenitis.


A correspondence outlining the possible usefulness of etretinate in the treatment of hidradenitis suppurativa. A case report demonstrates the success of the authors' treatment regimen.


Brief discussion proposing a link between hidradenitis suppurativa and adrenarche and the support it lends to the supposition that hidradenitis is an androgen-dependent disorder.


A startlingly radical concept of treating hidradenitis suppurativa with combined surgery and estrogen therapy. The theory and the results are presented briefly.


An excellent review of the surgical options available for hidradenitis suppurativa. The author outlines the pathogenesis, complications, management, and outcome of various options from multiple authors. This article is an excellent reference guide.


The authors present their series of axillary reconstructions with posterior arm fasciocutaneous flaps and outline how even extremely radical excisions may be covered with properly designed local flaps.


An excellent quick reference guide to pilonidal sinus. A great deal of information is packed into only three pages.


This excellent randomized study of open versus closed treatment of pilonidal sinus shows better outcome with closed treatment in terms of cost-effectiveness and morbidity than with the traditional open method.


A brief outline of the cost-effectiveness of closed treatment of natal cleft sinus. The authors conclude that primary closure should be widely used.

Fig. 1. Components of the follicular occlusion tetrad. A, Folliculitis decalvans with scarring alopecia. (Courtesy of J. L. Bolognia, New Haven, CT.) B, Hidradenitis suppurativa of the axilla. C, Acne conglobata of the back. D, Pilonidal cyst.

Fig. 1a

Fig. 1b

Fig. 1c

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Fig. 2. Pathogenesis of a pilonidal sinus. (From Jones [36]; with permission.)

Fig. 3. Z-plasty for excision of pilonidal sinus.

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