Clinical features

Toxic erythema of the neonate (erythema toxicum, erythema toxicum neonatorum) is a very common, self-limiting disorder that presents as an asymptomatic erythematous macular rash usually in the first few days of life. Very rarely, the eruption occurs a week or more after birth. It affects up to 50% of neonates. In survey studies from Japan, Australia, China and India, toxic erythema was found in 40.8%, 34.8%, 33.7% and 20.6% of infants, respectively. It may be associated with papules, and occasionally pustule formation is evident. It most often involves the forehead, face, chest, trunk and extremities. Lesions usually resolve in a few days. Its etiology is obscure.

Histological features

Early erythematous lesions show a somewhat nondescript perivascular inflammatory cell infiltrate with conspicuous eosinophils, which may be seen penetrating the epidermis in close proximity to hair follicles. The pustules are characteristically intrafollicular, may be subcorneal or intraepidermal, and contain large numbers of eosinophils and occasional neutrophils.

Differential diagnosis

Toxic erythema of the neonate must be distinguished from incontinentia pigmenti. The latter, however, is characterized by eosinophilic spongiosis, a feature not seen in toxic erythema. In miliaria rubra the vesicles are related to sweat ducts rather than hair follicles and typically contain mononuclear cells rather than eosinophils.

References


Hidradenitis suppurativa

Clinical features

Hidradenitis suppurativa is a common disease which is also known as acne inversa and more rarely apocrine acne. Studies from Denmark have found the prevalence to be around 4%, 1,2,3 Hidradenitis suppurativa is a chronic relapsing suppurrative inflammation of regions where apocrine glands occur, i.e. the axilla, inguinal folds, perineum, genitalia and perianal region (Fig. 14.50).4,5 It occurs postpubertally in both sexes, but is more common in women. The disease is seen most frequently in young adults, although its first presentation may be in older individuals and even before puberty.5,6 Initially, there is a firm painful nodule in the groin or axilla. The nodule may involute slowly or else discharge pus through the skin; the discharge of pus is not copious, but is chronic and often foul smelling. In the late stages a complex interconnecting system of sinuses extends deeply into the dermis and subcutaneous fat with extensive dense fibrosis (Fig. 14.51).5

Fig. 14.50
Hidradenitis suppurativa: early lesion presenting as an erythematous nodule discharging clear fluid. The axilla is a commonly affected site. By courtesy of R.A. Marsden, MD, St George’s Hospital, London, UK.

Fig. 14.51
Hidradenitis suppurativa: in this very severe example there is marked scarring and numerous sinuses are present. By courtesy of R.A. Marsden, MD, St George’s Hospital, London, UK.
Axillary lesions are more common in women and genitoinguinal lesions are more common in men. Changes may be confined to one region or occur in both, but the axillary region is involved in over 70% of cases. Some reports have attached etiological importance to axillary shaving and the use of deodorants, but this is not generally accepted. In women, obesity appears to be a predisposing factor and smoking is closely associated, but it is not clear whether this is a cause or effect. In one study, nearly 90% of German patients were smokers (expected prevalence rate 27%). Whether cessation of smoking improves the course of the disease is unknown. Patients with the hidradenitis suppurativa appear to be at increased risk of developing non-melanoma skin cancer.

The lesions are clearly maintained by bacterial infection as various organisms are often grown. Symptomatic improvement can be achieved with long-term antibiotics. Perineal lesions are often severe and complicated by abscesses, fistulae and draining sinus tracts. Lesions may also rarely be seen on the malar region of the face and even on the eyelids (glands of Moll), sites with modified apocrine glands.

Hidradenitis suppurativa can be seen in association with conditions which are said to be pathologically similar, namely acne conglobata and dissecting folliculitis of the scalp. These three conditions have been referred to collectively as the ‘follicular occlusion triad’. Any one condition, however, may occur separately. Acne conglobata, an extremely severe nodulocystic variant of acne, occurs extensively on the trunk, buttocks and limbs with predilection for males. The disease has been described in association with HIV and following pregnancy. Dissecting folliculitis (folliculitis capitis abscedens et suffodiens) is centered on the vertex of the scalp and is characterized by boggy tender lesions that tend to become confluent with formation of draining sinuses and suppuration. The disease presents more commonly in black males and it is very rarely familial. Radical surgery is often the only satisfactory means of terminating the process. All the diseases in the follicular occlusion triad can occasionally be complicated by progression of the infective process to cellulitis and septicemia. Squamous carcinoma (including the verrucous variant) is a rare and late additional complication. As in Marjolin’s ulcer–cancer, the carcinomas are capable of aggressive invasion and metastasis (50%) and are generally associated with a poor prognosis. Such tumors arise most frequently on the buttock and are more often seen in males. Hidradenitis has been shown to be rarely associated with systemic granulomatous lesions, in particular Crohn’s disease. An association with spondyloarthopathy, Dowling–Degos disease and lithium therapy has also been documented.

Treatment of this disease is difficult due to its chronic relapsing nature. Surgery is often used to remove affected areas but the cure rate in some studies is very low. Nevertheless, occasional patients are satisfied with the relief of symptoms, albeit temporary, afforded by surgery. Other studies have shown a low recurrence rate following wide excision. Early surgical treatment appears to increase the chance of success.

Pathogenesis and histological features

The pathogenesis of hidradenitis suppurativa is poorly understood. It has generally been thought that the earliest lesion is an acute inflammatory process involving the apocrine duct and gland, which extends into the surrounding connective tissue with subsequent abscess and sinus formation (Fig. 14.52). Some authors, however, believe that eccrine hidradenitis is more commonly found than apocrine involvement and others think that the primary event is follicular obstruction. The provocation to the initial ‘apocrinitis’ is believed to be keratin occlusion of the corresponding hair follicle. Certainly, keratin plugging of follicles and sinuses and inflammation in and around the hair follicle are regularly seen. In one study, follicular occlusion was present in all of 118 specimens examined in patients with disease duration that ranged from as little to 1 month to many years. The anatomic distribution of the lesions also supports the concept of an underlying apocrine gland defect. The condition has some similarity to Fox–Fordyce disease, which is more convincingly associated with an inflammatory process of the apocrine duct. Fox–Fordyce disease has the same sex, age incidence and

![Fig. 14.52](image)

(a, b) Hidradenitis suppurativa: early lesion showing acute inflammation involving the apocrine gland.
anatomic distribution and it too is alleviated by pregnancy. Interestingly, some cases of Fox–Fordyce disease have been reported to progress to hidradenitis suppurativa.

The other members of the follicular occlusion triad – acne conglobata and dissecting folliculitis – are both clearly associated with keratin plugging.

There is no doubt that the main cause of symptoms and chronic disability are the sinuses and fibrosis; these are largely due to the chronic secondary infection, since injection of sterile apocrine sweat into tissues does not induce an inflammatory response.

Organisms that may be found include Staphylococcus aureus, Streptococcus viridans, Escherichia coli, Proteus mirabilis, Klebsiella, Pseudomonas aeruginosa, Streptococcus milleri and anaerobic organisms. Coagulase-negative S. aureus is the most common bacterium isolated from the depth of the lesions. Anaerobic organisms are responsible for the offensive smell, which can be a major problem for the patient. Generally, no immune deficiency can be detected, but there have been occasional reports of a functional neutrophil deficiency.

In considering the pathogenesis of this condition it must also be noted that some cases clearly develop as an autosomal dominantly inherited tendency. Others have no suggestion of familial incidence.

The disease has been simulated in 3 of 12 normal volunteers by occlusion of axillary skin with atropine tape following depliation. The latter in itself could be expected to produce some pathology, which is clearly not seen in the normal individual. The absence of lesions in 75% of these volunteers shows at least that there is some variation in susceptibility to developing the disease. This experimental induction of the disease has not been repeated.

In a study of 42 women with hidradenitis suppurativa, the authors noted premenstrual exacerbation of symptoms in two-thirds of patients and over one-third of patients reported menstrual irregularities. In this same study, testosterone and free androgen index were higher compared to women. Pregnancy may relieve the symptoms of the disease.

Differential diagnosis

The main differential diagnoses are primary infection, a response to a ruptured epidermal inclusion cyst, or wounds. Clinical correlation is necessary to establish the correct diagnosis.

References

43. Luppa, J., Jarek, G., Em SST, L. (1999) Coagulase-negative staphylococci are the most common bacteria found in cultures from the deep portions of hidradenitis suppurativa lesions as obtained by carbon dioxide laser surgery. Br J Dermatol, 140, 90–93.