Familial acne inversa with acne conglobata in three generations

Sir,

Acne inversa (Hidradenitis suppurativa) is a chronically relapsing inflammatory disease that is characterized

by recurrent draining sinuses and abscesses occurring predominantly in skin folds that carry terminal hairs and apocrine glands.[1] Healing occurs with substantial scarring. We report here a family with acne inversa and acne conglobata occurring in three generations.

A 33 year-old male with no history of consanguinity, presented to the outpatient department with multiple painful swellings of eleven years' duration over the face, chest, axillae and gluteal region. He had taken various courses of antibiotics without much benefit. His younger brother also was suffering from similar complaints, but with less severity. Both his father and grandfather also had similar lesions in the axillae, but had not taken any treatment. Local examination revealed multiple, hyperpigmented, firm nodules of varying sizes measuring one to several centimeters in size. Nodules on the chest wall coalesced to form large plaques with sinus-discharging pus. Larger nodules were seen on the gluteal region [Figure 1]. The inguinal lymph nodes were enlarged, discrete and nontender. General and systemic examinations were unremarkable. His younger brother aged 30 years also showed similar lesions of ten years' duration. On examination, he also revealed similar nodules and cystic lesions, but the lesions were less severe. There was no sinus formation at the time of examination [Figure 2].

Investigations revealed normal blood counts, liver and renal function tests and the ESR was 110 cm in the first hour. Glucose tolerance test was normal. Bacterial culture showed heavy growth of *Staphylococcus aureus*. Serology for HIV-1 and 2 gave negative results. Biopsy revealed multiple epidermal inclusion cysts and perifolliculitis with neutrophilic and histiocytic infiltrates. He was treated successively with oral doxycycline, minocycline, isotretinoin, acitretin and intralesional corticosteroids. He had some relief with oral acitretin combined with intralesional steroids, but there was no evidence of complete remission.

Investigations revealed similar findings in the younger sibling. He was treated with rifampicin 600 mg per day for eight weeks, and showed good improvement with prolonged remission.

Both our patients suffered from acne conglobata with acne inverse, which was evident from the history and clinical examination findings. There was no evidence of the follicular occlusion triad as they did not suffer from dissecting cellulitis of scalp.

In some families, acne inversa may show a single gene dominant inheritance. Fitzsimmons *et al.* studied three families with a total of 21 affected members and reported that the pattern of transmission and the number of affected individuals were consistent with autosomal dominant inheritance. Later, they studied the families of 266 subjects with acne inversa, comprising a total of 62 affected individuals. They reported that 34% were first-degree relatives in 11 families.[2,3] There was a history of disease in three generations in our patients' family, suggesting an autosomal dominant inheritance.

More recent studies have identified acne inversa as a disorder of follicular, rather than apocrine occlusion. Thus, apocrine gland involvement is only incidental or secondary to the primary developments that involve the terminal hair follicle. Unlike acne vulgaris, acne inversa is localized in nonfacial regions where there are terminal pigmented coarse hairs. Acne inversa is a segmental rupture of the follicular epithelium, followed by spilling of foreign body material, such as corneocytes, bacteria, sebum products and hairs into the dermis. The dumping of foreign products initiates an inflammatory response provoking foreign body granuloma and epithelial



Figure 1: Shows multiple nodules in the gluteal region



Figure 2: Acne conglobata in siblings

strands, which then try to encapsulate the necrotic tissue. The apocrine glands are not involved in the earliest stage of follicular hyperkeratosis. Once rupture of the follicular epithelium has occurred, the disease spreads rapidly.[4] The draining sinus is a late complication of acne inversa.

Acne inversa has to be differentiated from furuncles, carbuncles, vegetating pyoderma, cutaneous tuberculosis, actinomycosis, lymphogranuloma inguinale and Crohn's disease.[5] Treatment options include antiseptics, antibiotics and corticosteroids.[6] Tetracycline, clindamycin or rifampicin have all been tried with limited success. Isotretinoin is effective in some cases only. Etretinate and acitretin have been more successful.[7] Incision and drainage or exteriorization of individual lesions may be useful in some instances.[8] Radical surgical excision at the earliest recognized stage remains a mainstay of therapy. Inadequate excision is the main reason for recurrence. Carbon dioxide laser excision is another treatment option for acne inversa.

Our patients were treated with adequate courses of antibiotics. The elder sibling did not respond to the various treatments given. Hence, he was given acitretin at a dose of 25 mg per day for four months. Large nodular lesions began decreasing in size. Intralesional steroid therapy was also given to large nodules on the chin and the lip. These lesions also partially responded to therapy. Some of these lesions recurred after stopping the treatment. Large nodules on the gluteal region were treated by surgical intervention earlier but there was recurrence at that site as well. On the other hand, the younger sibling showed an adequate response with rifampicin. There was no evidence of recurrence after stopping the therapy.

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