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Acne conglobata and rarer forms of acne

See also the separate Acne Vulgaris and Neurotic Excoriation and Acne Excoriée articles.

Rare forms of acne

Acne vulgaris in the form of 'teenage spots' is very common in adolescence but other rarer forms of acne may occur. Severe forms of acne can affect many aspects of a person's life, causing a great deal of embarrassment and stress. Severe acne may significantly limit social life and even interfere with opportunities for employment. Rarer variants of acne include:[1]

- **Acne conglobata**: very severe form of nodulocystic acne in which inflammatory lesions predominate and run together and often form exudates or bleed. Acne conglobata may cause extensive scarring.
- **Acne fulminans**: sudden, severe inflammatory reaction which causes deep ulcerations and erosions; may be associated with fever and arthralgia.
- Acne excoriée**:** mainly affects young women and is characterised by self-inflicted wounds associated with a psychological or emotional problem.
- **Acne mechanica**: caused by pressure, friction or rubbing from clothing.[2]
- **Acne cosmetica**: caused by contact comedogenic products with the skin. One study found the link between acne and cosmetics was weak but conceded that it was possible with some products.[3]
- **Chloracne**: caused by occupational exposure or military exposure to halogenated hydrocarbons. It presents with many large comedones.[4]

Acne conglobata[5] [6]

Acne conglobata is a rare and severe form of acne found most often in men. It presents with extensive inflammatory papules, suppurative nodules (may coalesce to form sinuses) and cysts on the trunk and upper limbs.[1]

Important information
Refer people to a consultant dermatologist-led team if they have acne conglobata.[7]

How common is acne conglobata? (Epidemiology)

- Acne conglobata is uncommon and may develop as a result of a sudden deterioration of existing active papular or pustular acne, or may occur as a recurrence of acne that has been inactive for many years.
- Males are affected more often than females.
- The onset is usually between the ages of 18-30 but infants can be affected as well.

What causes acne conglobata? (Aetiology and risk factors)

- The primary cause of acne conglobata remains unknown.
- Changes in reactivity to *Propionibacterium acnes* may be important.
- Androgen-producing tumours and anabolic steroids used for medical or other purposes may induce severe acne.
- There is a tendency for it to run in families and there is an association with certain HLA antigens. There is a familial link with pyoderma gangrenosum, aseptic arthritis and hidradenitis suppurativa.[8]

Presentation of acne conglobata

Acne conglobata is a chronic and severe form of acne vulgaris showing:

- Deep abscesses.
- Inflammation.

- Severe damage to the skin.
- Scarring.
- Comedones (blackheads) which are obvious and widespread, often occurring on the face, neck, trunk, upper arms and/or buttocks.

Inflammatory nodules may form around multiple comedones and grow until they break down and discharge pus. Deep ulcers may form under the nodules, producing keloid-type scars, and crusts may form over deeply ulcerated nodules. Abscesses can form deep, irregular scars.

Acne conglobata may be preceded by acne cysts, papules or pustules that do not heal but instead rapidly deteriorate. Occasionally, it flares up in acne that had been dormant for many years.

Rarely, acne conglobata can be associated with pyogenic arthritis and pyoderma gangrenosum (known as PAPA). This is thought to be a genetic condition (a defect of chromosome 15). Another variant is pyoderma gangrenosum, acne and suppurative hidradenitis (PASH) syndrome.

Differential diagnosis

- Acne rosacea.
- Folliculitis.
- Acne fulminans.
- Acne vulgaris.
- Acneiform eruptions.
- Sporotrichosis.

Investigations [1]

Diagnosis is usually clinical with no investigations required for diagnosis. However, underlying conditions must be considered:

- Total and free testosterone for polycystic ovary syndrome (PCOS) or ovarian cancer. The androgen producing arrhenoblastoma is rare.
- Serum dehydroepiandrosterone sulfate (DHEAS) for adrenal tumour or congenital adrenal hyperplasia.

- Ratio of LH/FSH for PCOS.
- 17-hydroxyprogesterone for congenital adrenal hyperplasia.
- Prolactin in case of pituitary adenoma.
- 24-hour urinary free cortisol for Cushing's syndrome.
- If isotretinoin is considered, baseline blood tests such as LFTs and fasting lipids are required.

Management of acne conglobata

- Regular face washing and the use of antiseptic gels may reduce the amount of *P. acnes*.
- Emotional support is essential.

Drugs

- Oral isotretinoin commenced early to prevent scarring is recommended. Treatment is required for at least five months, and further courses are sometimes necessary.
- Intralesional steroids following cyst drainage, can be used for individual persistent or large inflammatory nodules or cysts.
- Additional treatments are usually required, including:
    - Oral antibiotics for secondary bacterial infection.
    - Systemic corticosteroids to reduce inflammation.
    - Adalimumab for resistant severe disease.
- Topical treatment is usually ineffective.

Surgery

- Large haemorrhagic nodules may be aspirated.
- Intralesional triamcinolone or cryotherapy may be effective.
- Surgical excision of interconnecting large nodules may occasionally be beneficial.

Complications [9]

- The psychological effect of severe acne on the developing adolescent must not be underestimated.
- Renal amyloidosis has been reported.
- Scars remain for life.

Prognosis

The disease has a chronic course, leading to extensive scarring and psychological distress.

Prevention

There is nothing that can be done to prevent this disease but it needs to be treated energetically to minimise the psychological impact and to reduce scarring.

Acne fulminans[10] [11] [12]

Acne fulminans is a rare and severe form of inflammatory acne presenting abruptly with painful, haemorrhagic pustules and ulceration, that may or may not be associated with systemic symptoms, such as fever and polyarthritis. It typically affects male teenagers with pre-existing acne.

Important information
Urgently refer people with acne fulminans on the same day to the on-call hospital dermatology team, to be assessed within 24 hours.[7]

Presentation of acne fulminans

- Sudden onset of severe and often ulcerating acne, associated with fever and polyarthritis.
- Acne fulminans causes many inflammatory nodules on the trunk. Large nodules tend to become painful ulcers with surrounding exudative necrotic plaques which become confluent.
- Erythematous neovascular nodules may also be seen.
- Painful splenomegaly, inflammatory arthralgia (this especially affects the hips and knees), bone pain, erythema nodosum and chronic aseptic multifocal osteomyelitis may be present.

- Acne fulminans can be the dermatological manifestation of the synovitis-acne-pustulosis-hyperostosis-osteitis (SAPHO) syndrome. [13]

Investigations

Abnormal findings include the following:

- FBC: anaemia leukocytosis (with increased polymorphs).
- Raised ESR.
- Circulating immune complexes.
- Proteinuria.
- Blood culture will be sterile.
- X-rays: approximately 50% of patients have lytic bone lesions. Destructive lesions resembling osteomyelitis may be seen.
- Technetium scintillography: multifocal osteolytic cysts may be detected as hot spots.

Differential diagnosis

- Acne conglobata.
- Acne vulgaris.
- Acneiform eruptions.
- Pyoderma gangrenosum.

Management of acne fulminans

- Systemic corticosteroids (prednisolone) and retinoids (isotretinoin) are usually the first choice of treatment.
- Dapsone, ciclosporin A, methotrexate, azathioprine, levamisole, and biological agents (eg, anakinra, infliximab or adalimumab) may be considered as alternative therapies.
- Adjunctive topical and physical therapies may also be considered..
- Pulsed dye laser is effective for granulation tissue associated with acne fulminans.

Prognosis

- The prognosis is generally good in patients treated appropriately and recurrence of acne fulminans is rare.
- However, scarring and fibrosis may occur, and may be severe.

Management of the rarer forms of acne

- **Acne mechanica**: reducing heat and moisture helps (eg, by changing clothing and showering after exercise).The obvious treatment is to avoid the aggravating trauma but, if this is not possible, topical treatment with salicylic acid or benzoyl peroxide is helpful.[2]
- **Acne cosmetica**: this was common in the 1970s and 1980s but is now rare due to changes in formulation of cosmetics. Treatment includes a review of cosmetic products to exclude any that potentially block skin pores ('comedogenic'). Further management options are as for acne vulgaris.[14]
- **Chloracne**: the only known treatment is to avoid exposure to chloracnegens (eg, occupational exposure, contaminated industrial waste, contaminated food products).[15]

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