Acne Conglobata and Rarer Forms of Acne

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

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 cosmetic: 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]

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.

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.[6]
- 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.[7]

Presentation
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[8]

- 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

- One study found that a low-glycaemic-load diet improved patients with acne vulgaris.\[10\] Presumably, similar considerations apply to acne conglobata. Regular face washing and the use of antiseptic gels may reduce the amount of \textit{P. acnes}.
- Emotional support is essential.
- People who have a severe variant of acne should be referred urgently (to be seen within two weeks) to a dermatologist.\[11\]
- People who have severe acne, such as painful, deep nodules or cysts (nodulocystic acne), or other people who could benefit from oral isotretinoin, should be referred as ‘soon’.\[11\]

Drugs \[5\]

- The therapy of choice is oral isotretinoin. Simultaneous use of oral prednisolone is also sometimes tried.
- There is some evidence to support the use of oral antibiotics in combination with azelaic acid. The guidelines of the European Dermatology Forum failed to identify any conclusive evidence supporting the choice of a first-line option. Specialist opinion supports the use of oral tetracycline or erythromycin.\[13\]
- Oral contraceptives: Dianette® (ethinylestradiol with cyproterone) may be particularly effective.\[14\] Dianette® is an effective contraceptive but it is not licensed as such and the patient should be warned that pregnancy whilst taking this can result in the cyproterone causing ambiguous genitalia in a male fetus.
- Dapsone is recommended for treatment-resistant cases.
- Acne conglobata has been successfully treated by carbon dioxide laser combined with topical tretinoin therapy.
- Modern external beam radiation has been used with some success.\[15\]
- Infliximab has been tried but not with good results.\[16\] However, one study reported the successful use of adalimumab.\[17\]
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

- The psychological effect of severe acne on the developing adolescent must not be underestimated.
- Renal amyloidosis has been reported.[20]
- 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

Acne fulminans is an uncommon, immune systemic disease in which the triggering antigen is thought to be *P. acnes*. Acne fulminans predominantly affects young males with a history of acne. High levels of testosterone (e.g., during therapy for Marfan's syndrome) and anabolic steroids appear to be trigger factors. Isotretinoin is also a precipitant, possibly due to an increase in *P. acnes* antigens in the patient's immune system.[23] Acne fulminans can be the only feature of late-onset congenital adrenal hyperplasia in males. Genetic factors may play a part and concordance in twins has been reported.

Epidemiology

It is a rare condition and becoming rarer, due to improved treatment of acne. The typical patient is a young male aged between 13 and 22.[24]

Presentation

- 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.[25]

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
- Oral steroids should be started and gradually reduced over six weeks.
- Oral isotretinoin should be started after four weeks and gradually increased to achieve complete clearance.
- Response to broad-spectrum antibiotic treatment is poor.
- One study reports the successful use of ciclosporin in combination with prednisolone.
- Infliximab may be used if other treatments are ineffective.
- Pulsed dye laser is effective for granulation tissue associated with acne fulminans.

Prognosis
- The prognosis is good in patients treated appropriately and recurrence of acne fulminans is rare.
- Scarring and fibrosis may occur.

Further reading & references
- Acne vulgaris: NICE CKS, September 2014 (UK access only)
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