

*\*\*no patient handout*

## Acne fulminans

### Synopsis

Acne fulminans is a rare, highly inflammatory, immunologically induced form of acne that occurs mainly in male patients between 13 and 22 years old. Individuals of Northern European descent are predominantly affected. Individuals of East Asian descent may experience a milder form of acne fulminans. Risk factors include chronic severe acne (several months or years), a positive family history, high testosterone levels, and history of anabolic steroid use.

The inciting antigen is believed to be from *Propionibacterium acnes*. Testosterone may play a role in the pathogenesis as well, as this steroid hormone increases sebum excretion and the population density of *P. acnes*. Circulating immune complexes have been found in patients with acne fulminans.

In acne fulminans without systemic symptoms (AF-WOSS), patients present with a sudden onset of large inflammatory cysts, nodules, papules, pustules, and abscesses. In acne fulminans with systemic symptoms (AF-SS), patients are febrile with systemic complaints, which may include polyarthralgia, myalgias, malaise, anorexia, and weight loss. Splenic tenderness, erythema nodosum, and bone pain from aseptic osteolysis have been reported.

Isotretinoin treatment, especially if given at high doses, may induce acne fulminans in individuals with severe acne. The diagnosis is known as isotretinoin-induced acne fulminans without systemic symptoms (IIAF-WOSS), since systemic symptoms are typically absent. In rare cases, systemic symptoms accompany, and the diagnosis is known as isotretinoin-induced acne fulminans with systemic symptoms (IIAF-SS).

Acne fulminans can be the dermatologic manifestation of **SAPHO** (synovitis, pustulosis, hyperostosis, and osteitis), PAPA (pyogenic arthritis, pyoderma gangrenosum, and acne), PASH (pyoderma gangrenosum, acne, and suppurative hidradenitis), and PAPASH (pyogenic arthritis, pyoderma gangrenosum, acne, and suppurative hidradenitis) syndromes.

### Codes

ICD10CM:

L70.8 – Other acne

SNOMEDCT:

4659007 – Acne fulminans

### Look For

Multiple inflammatory cysts, nodules, papules, and pustules covering broad areas on the back, chest, and buttocks, as well as the abdomen, shoulders, and upper arms. The face and neck may also be affected. Fluctuant tender cysts often drain serosanguineous fluid or pus. The severity of

inflammation can result in erosions and ulcerations with hemorrhagic crusts. Eventual healing with deeply pitted scars follows ulceration. Keloids often form after ulceration.

Systemic symptoms include fever, arthralgias, myalgias, malaise, anorexia, weight loss, aseptic osteolysis, splenic tenderness, and erythema nodosum.

## Diagnostic Pearls

The neck is usually spared, and the face is involved less often than the trunk.

## Differential Diagnosis & Pitfalls

- **Acne conglobata** lacks many of the systemic manifestations of acne fulminans, and onset is typically at a later age. Acne conglobata exhibits polyporous comedones and noninflammatory cysts.
- **Steatocystoma multiplex** may become infected, but usually does not display the systemic findings.
- **Acne vulgaris**
- **Chloracne**
- Staphylococcal or gram-negative **folliculitis**
- **Rosacea**
- **Pyoderma faciale**
- **Hidradenitis suppurativa**

## Best Tests

The following laboratory and/or imaging abnormalities may be present if systemic findings are present:

- CBC with differential: leukocytosis (common), anemia, neutrophilia.
- Erythrocyte sedimentation rate and C-reactive protein: elevated.
- Urinalysis: proteinuria.
- Plain bone radiographs: osteolytic lesions are in areas of bone tenderness (common sites include the sternum, clavicles, sacroiliac joints, and hips) in patients with AF-SS and IIAF-SS. Hyperostosis of the anterior chest wall, spine, pelvis, or limb may be seen in SAPHO.

- Technetium bone scan: increased uptake and osteolytic cysts ("hot spots") in AF-SS and IIAF-SS.
- Blood cultures: negative.
- Culture of lesions will most likely yield skin commensals, such as *P. acnes* and *Staphylococcus epidermidis*.

## Management Pearls

The disease characteristically does not respond to oral antibiotic therapy alone. Similarly, topical antibiotics are unhelpful in acne fulminans.

Oral glucocorticoids are usually essential for control of the severe inflammatory response. Isotretinoin is also of benefit in some cases (although oral glucocorticoids must first be administered to prevent explosive flares and should be tapered slowly to avoid flares).

The scarring from acne fulminans may be disfiguring. Pulsed-dye laser may be used to improve scarring.

## Therapy

Systemic glucocorticoids and isotretinoin with adjunctive oral antibiotics are the standard of therapy. Intralesional steroids and dapsone therapy also may be effective in some cases.

- Recommended dosages: 0.5-1 mg/kg per day of prednisone (usually 20-60 mg every 24 hours) for at least 4 weeks for AF-SS and at least 2 weeks for AF-WOSS. Then, isotretinoin 0.1 mg/kg every 24 hours can be initiated while continuing prednisone for 4 additional weeks. At this time, if the crusted lesions have healed, continue isotretinoin, and taper prednisone treatment over a 4-week period by halving the dose every week until a daily physiologic dose is reached. Then continue the physiologic dose every other day for 2 weeks. After that, slowly increase the dose of isotretinoin as tolerated, and monitor for crusting and erosions, until a dose of 1 mg/kg every 24 hours is reached. The cumulative goal dose for isotretinoin is 120-150 mg/kg. The total duration of treatment is at least 3-5 months.
- If the patient flares after isotretinoin is added to prednisone treatment, stop isotretinoin, and continue prednisone at 1 mg/kg per day.
- Possible additional therapies that have been reported to be beneficial include dapsone (100 mg a day), cyclosporine, tumor necrosis factor alpha inhibitors, anakinra, or canakinumab.
- Alternatively, erythromycin (1-2 g every 24 hours), doxycycline (100 mg every 12 hours), minocycline (100 mg every 12 hours), or tetracycline (500 mg to 1 g every 12 hours) can be added.

Arthralgias generally respond well to salicylates.

## Drug Reaction Data

Below is a list of drugs with literature evidence indicating an adverse association with this diagnosis. The list is continually updated through ongoing research and new medication approvals. Click on Citations to sort by number of citations or click on Medication to sort the medications alphabetically.

Medication	Citations
androgen	<a href="#">3</a>
dapsone	<a href="#">3</a>
Isotretinoin	<a href="#">10</a>
Leprostatic	<a href="#">3</a>
Retinoid	<a href="#">10</a>
testosterone	<a href="#">3</a>