

# A Case of Acne Fulminans



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Acne fulminans (AF) is a rare form of acne vulgaris. The diagnosis can be mistaken for other skin infections and delay treatment, placing patients at risk for scarring and psychological distress. This case report presents a patient previously treated for COVID-19 and subsequent pneumonia. COVID-19 restrictions in the clinic setting, including mask-wearing and isolation protocols, accounted for the suboptimal assessment of the patient's skin, which ultimately prolonged the diagnosis. This case report reviews AF from a primary care perspective and highlights clinical manifestations, physical examination findings, and management of AF. *J Pediatr Health Care.* (2022) 36, 603–606

## KEY WORDS

Acne vulgaris, acne fulminans, COVID-19, pediatric primary management of acne

## CASE REPORT: ACNE FULMINANS

### Chief Complaint and History of Present Illness

A 17-year-old male presented to the office with complaints of worsening fatigue, low-grade fever, and joint pain. His history was notable for a severe acute respiratory syndrome coronavirus 19 diagnosis 6 weeks prior with a course complicated by bilateral, interstitial, radiograph-confirmed pneumonia with asthma exacerbation approximately 3 weeks after diagnosis. He was treated with augmentin, albuterol, and

Symbicort per his asthma action plan. His pneumonia resolved, and the asthma was controlled. He represented to the clinic 5 weeks after a COVID-19 diagnosis with fatigue and joint pain and was diagnosed with post-COVID-19 long haul syndrome. One week later, he presented to the clinic with reported difficulty focusing on his schoolwork because of whole body aches, recorded average body temperatures of 38.0–38.2°C for the previous 6 days, and decreased appetite and dull headache for the past several days. On a complete review of systems, he denied cough, shortness of breath, nausea, vomiting, diarrhea, and swelling of his extremities.

At the time of the visit, the parents had been in close contact with a COVID-positive patient and were asked to stay in their car. The parents permitted the child to be seen in the office without their presence. The practitioner reviewed the chief complaint and history of present illness with the parent in the parking lot. In addition to the child's reported symptoms, the mother reported they had a dermatologist appointment the next week for his acne breakout.

### Medical History

This patient's care was established at the clinic. Besides his recent COVID-19 diagnosis, he had no known medical or surgical history. He had been treated for mild-intermittent asthma and seasonal allergies, typically well controlled during the spring and fall seasons. He was not taking any medication at the time of the visit. He had no known allergies to medications or food. Family history was noncontributory.

### Social and Developmental History

At the time of the visit, the patient lived in a private home and shared a room with his twin brother. Both parents were working outside of the home. He reported being a senior in high school and, at the time of the visit, classes were a hybrid of live and virtual instruction. He was applying to colleges with the intent to study medicine. He denied using street drugs, alcohol, and/or vaping. He identified as a heterosexual male and denied having been sexually active. He denied anxiety, depression, suicidal ideation, and recent travel.

### Pertinent Physical Findings

The patient wore a mask throughout the visit to the office because of COVID-19 pandemic protocols. The patient was well-appearing, alert, and cooperative. He had a mildly

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**TABLE 1. Classification of acne by severity**

Classification of acne	Clinical features
Mild	Comedones present with or without the presence of inflammatory papules and pustules. Typically < 20 lesions on the face and or trunk. Scarring is unlikely
Moderate	Comedones are present with inflammatory papules and pustules. Greater than 20 lesions were noted on the face and trunk. Temporarily disfiguring
Severe	Nodulocystic acne was noted with confluent lesions. Comedones are severe, with inflammation noted as well as papules and pustules. Scarring is common

Note. Adapted from Ashton and Weinstein (2019).

elevated temperature of 38.2°C, and all other vital signs were appropriate for his age. The patient’s skin assessment was remarkable for severely painful, ulcerative, hemorrhagic nodules with pustular-like lesions. The lesions were diffuse and noted on his forehead, bilateral temporal areas, bilateral cheeks, chin, submandibular region, chest, and back. Lesions on the chest were oozing mucopurulent discharge. He was noted to have multiple palpable lymph nodes, 1–2 cm in size, that were mobile and nontender. Tender lymph nodes were palpable and noted bilaterally in the axillae, 1–2 cm in size, and mildly painful to palpation. There was no hepatosplenomegaly. The rest of his examination was noncontributory.

### Differential Diagnosis

The initial diagnosis considered was acne vulgaris with a secondary staphylococcal infection. Pustular nodules on the face, jawline, back, and chest guided the diagnostic approach. According to Giuliano, Patel, and Kale-Pradhan (2019), patients with skin lesions associated with purulent drainage are likely to be infected with *Staphylococcus aureus*. Superficial skin and soft tissue infections (SSTIs) have been studied over the past decade, and practice protocols for treating these lesions are widely accepted. Practitioners must consider resistant *Staphylococcus aureus* within the setting of an SSTI; however, this patient’s history and physical examination did not reveal lesions consistent with an SSTI. This patient’s lesions were nonindurated and diffuse, whereas SSTI lesions are usually individual with extensive induration. The patient’s lesions did appear impetiginous; therefore, the clinician followed the Infectious Diseases Society of America recommendations to obtain a wound culture to exclude methicillin-resistant *Staphylococcus aureus* (Liu et al., 2011). Because of the acute nature of the infection, additional differentials such as atopic dermatitis, psoriasis, rosacea fulminans, and telangiectasia were considered but promptly excluded because of the acute and inflammatory nature of the skin. Although the initial diagnosis of acne with secondary *Staphylococcus aureus* infection was a reasonable place to start in this patient, the number and characteristics of the lesions, and the presence of systemic symptoms, led the practitioner to consider more severe and rare forms of acne.

Acne vulgaris, the most common form of acne in the adolescent age group, is an inflammation of the pilosebaceous glands. The lesions should be classified as mild, moderate, or severe when assessing acne on the basis of clinical

features. See Table 1 for the classification and associated clinical features of acne. The more complex the lesions are at presentation, the more challenging the diagnosis and treatment are for the clinician. The patient presented with severe acne. The lesions were ulcerative, painful, and oozing with purulent discharge. His face alone had > 50 lesions confluent to the temporal and chin areas. The lesions on his chest and back were similar, with a distinct, painful, erythematous base.

Although acne is seen frequently in the pediatric population, and in most cases, it is routine, some cases are challenging and difficult to diagnose and treat (Dessinioti & Katsambas, 2017). Acne vulgaris, if wrongly used as a working diagnosis, can delay treatment of more rare forms of acne. Therefore, the clinician must understand how to differentiate benign acne vulgaris from the more severe rare acne conditions. The more rare and complex forms of acne include nonresponding acne, acne conglobata, and acne fulminans (AF). Table 2 notes the key history and physical findings of these conditions.

**TABLE 2. Rare and complex forms of acne**

Type of acne	Key history and physical findings
Nonresponding acne	<ul style="list-style-type: none"> <li>• Comedones, papules, and pustules on physical examination</li> <li>• Does not respond to conventional forms of treatment</li> <li>• Benefits from isotretinoin</li> </ul>
Acne conglobata	<ul style="list-style-type: none"> <li>• Groupings of pustules found on the trunk, rarely on the face</li> <li>• Comedones common</li> <li>• Chronic, slowly progressive</li> <li>• Typically, it affects males in the second to third decades of life</li> <li>• Not associated with systemic complaints</li> <li>• Difficult to treat because of antibiotic resistance</li> </ul>
Acne fulminans	<ul style="list-style-type: none"> <li>• Acute presentation</li> <li>• Often associated with severe, hemorrhagic, and ulcerative acne</li> <li>• Comedones uncommon</li> <li>• Face, chest, and back are affected</li> <li>• Systemic complaints (joint pain, fever, fatigue) are common</li> </ul>

Note. Adapted from Dessinioti and Katsambas (2017).

The clinical history and physical examination are generally what a clinician needs to make a formal acne diagnosis. This patient's history revealed systemic complaints of fatigue and generalized joint pain. When considering the hallmark findings within the acne vulgaris subgroups, the patient's systemic symptoms and clinical manifestations suggest AF. The aggressive nature of hemorrhagic ulcerative lesions involving the back, chest, jawline, and face, along with fever, arthralgia, and general malaise, led the practitioner to further evaluate him with blood work and wound culture, begin treatment, and consult with dermatology.

Initial diagnostics performed in the office included a complete blood cell count, comprehensive metabolic panel, and wound culture. The complete blood cell count revealed a mild leukocytosis of 13,000  $\mu$ /L (normal range, 4,500–11,000  $\mu$ /L), relieving bacteremia concern. There was no evidence of anemia. The patient's comprehensive metabolic panel was normal. The wound culture report would reveal, in time, that the specimen was positive for *Propionibacterium acnes* with multiple additional organisms present. This confirmed a diagnosis of AF.

### Acne Fulminans

Most reported cases of acne, 86%, are in the adolescent population, with persistent disease noted in patients aged between 20 and 30 years (Ashton & Weinstein, 2019). AF, an uncommon and severe form of inflammatory acne, is more commonly noted in Caucasian males aged between 13 and 22 years (Greywal et al., 2017). Patients who present with AF typically have had mild-to-moderate acne for approximately 2 years before the development of AF. The sudden onset of hemorrhagic ulcerative lesions involving the back, chest, and face is pathognomonic for AF. If untreated, the condition then progresses to include systemic findings such as fever, leukocytosis, anemia, hepatosplenomegaly, myalgia, and arthralgia (Dessinioti & Katsambas, 2017). The extremely painful nodules and ulcerative lesions will crust and heal, leaving severe scars behind (Greywal et al., 2017). According to the literature, no specific laboratory abnormalities are diagnostic of AF (Dall'Oglio, Puglisi, Nasca, & Micali, 2020). The most notable bacterial organism within AF lesions is *Propionibacterium acnes* (Dessinioti & Katsambas, 2017).

The terminology used to both diagnose and classify AF has been inconsistent. AF is a condition with a spectrum of clinical and historical findings and severity. It can be limited to skin lesions alone, but systemic symptoms might also be present. It is important to note that AF can also be associated with high-dose isotretinoin (Greywal et al., 2017). It has been referred to as acne maligna, acute febrile ulcerative acne, and acute febrile ulcerative acne conglobata with polyarthralgia (Siadat, Bostakian, Abtahi-Nacini, & Shahbazi, 2017). This spectrum of findings and confusing terminology has contributed to inconsistent diagnosis and management of the condition. Greywal et al. (2017) convened a group of experts to clarify this confusion and developed a standard of care for the classification, diagnosis, and treatment of AF.

The group differentiated AF on the basis of the presence or absence of systemic symptoms and recent use of high-dose isotretinoin (Greywal et al., 2017). This patient fits the subtype of AF with systemic symptoms without high-dose isotretinoin use.

The clinician must recognize acne associated with AF as inflammatory. Alterations in innate immunity, autoimmunity, adaptive immunity, and autoinflammation have been proposed to be causative; however, the evidence for AF is inconclusive. Autoinflammatory syndromes associated with AF have been related to elevations and activation of interleukin  $-1$  (Greywal et al., 2017). This suggests that the nature of the illness may be from an inflammatory process already present in the body. Recall that the patient was 6 weeks post-COVID-19 infection that was complicated with interstitial pneumonia. This might have contributed to his development of AF, but more data are needed from patients infected with COVID-19 to determine the true relationship between these two conditions.

### Management of AF

Medical management of AF needs to be swift to reduce the inflammatory process and decrease the risk of scarring. The treatment significantly differs from severe acne because of the severity of clinical presentation and possible systemic involvement (Dall'Oglio et al., 2020). It is imperative that the clinician complete a thorough history and physical examination. Arthralgias are common; when AF is severe, the sacroiliac and knee joints can be compromised. Radiographs can aid the clinician in determining if there are lytic bone lesions, with approximately 50% of cases having these radiographic findings (Dessinioti & Katsambas, 2017). In rare instances, management of severe cases of AF may require hospitalization for intravenous antibiotic administration and further management. It is important to note that few cases become severe, most likely because of the advancement and safety in the medical treatment of acne (Siadat et al., 2017).

Dependent on the severity of the illness, multidisciplinary care may be warranted. The patient was referred to dermatology and subsequently had an infectious disease. He was prescribed oral isotretinoin and oral prednisone. The recommended treatment is quick initiation of oral corticosteroids to decrease the inflammatory process. Corticosteroids should be continued as monotherapy at 0.5–1 mg/kg/day for 6 weeks, gradually decreasing over time. Once the lesions form crusts and/or scabs, low dose isotretinoin is often added starting at 0.25 mg/kg/day and increased slowly to 1 mg/kg/day to avoid unnecessary side effects. Treatment is usually required for 3–4 months for an optimal outcome (Greywal et al., 2017). The long-term prognosis with appropriate treatment appears good; generally, AF does not recur (Dessinioti & Katsambas, 2017). Other management options supported by literature include aspirin, cyclosporine when the AF is resistant or nonresponsive to isotretinoin, and other antibiotic treatments because of their anti-inflammatory properties (Greywal et al., 2017). Research demonstrates that treatment protocols using a combination of

prednisolone and isotretinoin lead to a faster control of systemic features and a faster clearance of the acne (Siadat et al., 2017). However, it must be noted that larger-scaled randomized controlled trials evaluating the treatment of AF are lacking in current literature (Greywal et al., 2017).

In addition to the medical management of AF, the psychological impact of the disease must also be assessed. Hazarika and Archana (2016) suggest acne can adversely affect a patient's physical, emotional, and daily life, resulting in poor self-esteem, poor body image, anxiety, and depression. The disfiguring elements of the lesions can have a profound negative impact on interpersonal relationships in adolescents (Hazarika & Archana, 2016). Although the literature does not correlate the duration or severity of the acne nor patient age with the severity of adverse effects on the quality of life (Eyüboğlu, Kalay, & Eyüboğlu, 2018), it stands to reason that the particularly disfiguring lesions of AF have the potential to significantly affect the psychological outlook of patients with the condition. Clinicians must consider not only the patient's psychological health but their ability to cope with the disfiguring elements of the disease and assess and support accordingly. Taking the time to evaluate the patient's emotional and psychological well-being should be considered the standard of care for the acne patient, particularly the patient with AF.

## IMPLICATIONS FOR PRACTICE

Pediatric primary care clinicians should be prepared to assess, diagnose, and treat acne. This has been complicated by infection control measures implemented during the COVID pandemic. Wearing face masks for prolonged periods has been shown to aggravate acne (Han, Shi, Chen, & Zhang, 2020). Furthermore, a decline in face-to-face encounters between health care providers and patients has been implicated in delayed identification of serious illnesses and failure to provide routine care for chronic conditions, including mental and behavioral diagnoses (Somekh, Pettoello-Mantovani, & Somekh, 2020).

Unfortunately, both these complications contributed to a delayed diagnosis of AF for this patient. With a focus on staff and patient safety and decreasing the risk of COVID-19 transmission, pediatric providers offered multiple car visits resulting in a limited physical examination. It was not until the pediatric nurse practitioner assessed his complaints during a face-to-face visit that the lesions around his mask were noted, prompting a thorough head-to-toe evaluation. Only then was the significant disease burden appreciated and the diagnosis of AF subsequently made. After treatment

with prednisone and isotretinoin, the inflammatory nature of his acne resolved.

There is much to learn from this case. Pediatric primary care providers should ensure they are comfortable treating mild-to-moderate acne to prevent progression to severe disease. That treatment should include not only management of the physical symptoms but also consideration of the psychological impacts of the disease, particularly in the more severe forms such as AF. Clinicians should continue to be acutely aware of the challenges mask-wearing and distanced encounters present and, despite them, be sure to complete thorough physical assessments. Altogether, these changes can improve the care of children with all forms of acne.

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