Clostridial Myonecrosis in an Adolescent Male

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ABSTRACT. Extremity pain is a common complaint in adolescents. Pain out of proportion to examination findings should raise suspicion of deep tissue infection. Clostridial myonecrosis is a rapidly progressive disease consisting of muscle necrosis and systemic toxicity. It is usually seen in elderly and immunocompromised patients. Here we report a case of clostridial myonecrosis in an adolescent male. Pediatrics 2005;116:e735–e737. URL: www.pediatrics.org/cgi/doi/10.1542/peds.2004-2876; infection, pain.

Clostridial myonecrosis is a rare disease in the United States, with an estimated incidence of 900 to 1000 cases per year.1 However, its virulent course and high mortality rate necessitate a high degree of suspicion among treating physicians. Here we present a case in an otherwise healthy teenager and a review of the recent literature.

CASE REPORT

A previously healthy 17-year-old male presented to the emergency department with a 2-day history of bilateral leg pain. The day before admission he was seen by his pediatrician and found to have fever, facial swelling, and oral ulcers. He was diagnosed with a presumptive viral illness and treated with supportive measures. He returned the next day with increasingly severe leg pain, and additional evaluation was undertaken.

His past medical history consisted of a diagnosis of Epstein-Barr virus 1 month before presentation from which he still complained of fatigue. He had done some light weight-lifting several days before the onset of pain but had no recent participation in organized sports activities. There was no history of trauma, and he denied drug or steroid use.

His initial vital signs were notable for a temperature of 39.1°C, respiratory rate of 32 breaths per minute, heart rate of 132 beats per minute, and a blood pressure of 141/62 mm Hg. He was pale, diaphoretic, and unable to stand secondary to his pain. Mild orbital edema was present as well as gingival swelling and a single oral ulceration. There was severe tenderness involving both of his thighs and his right calf muscles.

Urinalysis revealed hematuria and proteinuria and the absence of red blood cells. Blood counts showed a white blood cell count of 2700/µL (57% bands, 5% segmented neutrophils, 13% lymphocytes, 19% monocytes), and his creatine phosphokinase was elevated at 3594 IU/L. His electrolytes and coagulation profile were within normal limits. The erythrocyte sedimentation rate was 42 mm/hour.

Despite intravenous hydration and multiple doses of morphine and ketorolac, there was no improvement in the patient’s pain, and he was admitted to the hospital with a presumptive diagnosis of viral myositis for pain control, continued hydration, and monitoring of renal function. Within 2 hours of admission, he developed a violaceous rash over his legs and became hypotensive. On examination, he was noted to have subcutaneous emphysema in both lower extremities. His legs were noted to be markedly enlarged, and distally they were cool and pale. The symptoms soon spread to involve his right arm. Surgery was consulted and an emergent computed tomography scan was obtained, which showed gas extending from his calves to his psoas muscles bilaterally and also in his right arm (Fig 1).

Based on these findings, a diagnosis of necrotizing fasciitis was made. He received penicillin, clindamycin, cefotaxime, and vancomycin. The patient was taken to the operating room for surgical debridement. He was then transferred to an outside institution for hyperbaric oxygen therapy. On arrival after transport, he was noted to have massive necrosis of his extremities and underwent bilateral disarticulations at the hips and removal of his right bicep muscle. Blood and wound cultures grew Clostridium septicum. He was given a final diagnosis of clostridial myonecrosis.

After amputation, he received antibiotic and hyperbaric oxygen therapy. His clinical course also necessitated intubation and vasoressor treatment. Limited diagnostic testing for both immunodeficiency and malignancies, including HIV testing, abdominal computed tomography scan, and immunoglobulin and complement levels were negative at the outside institution. One month after his presentation, he was stable and transferred to a rehabilitation facility. He has had no additional infections, signs of immunodeficiency, or medical problems.

DISCUSSION

Clostridial myonecrosis, also known as nontraumatic or spontaneous gas gangrene, is an extremely rare occurrence in pediatric patients. The infection is characterized by muscle necrosis and systemic toxicity caused by several clostridial exotoxins, the most notable of which is α-toxin (phospholipase C).2,3 Clostridium septicum, an anaerobic Gram-positive rod, is the causative organism in the majority of cases, followed by Clostridium perfringens.4,5 Unlike typical cases of gas gangrene, which follow trauma and are caused by C perfringens, the majority of cases of spontaneous gas gangrene have no obvious external portal of entry. The presumed source is mucosal defects of the intestinal tract.6 Clostridium septicum is substantially more aerotolerant than C perfringens, and the inoculum required for infection is 300 times smaller, which may explain the absence of a portal of entry.1

In the adult population, gas gangrene is often associated with trauma, underlying immunodeficiency states, venous insufficiency, or malignancy.7 Similarly, in children, it has been associated with malignancies, leukemia, neutropenia, and immunosuppression.8,9 Spontaneous gas gangrene has a particular association with colon cancer.3,4,10 In a review of spontaneous Clostridium septicum infections, 34% of the patients were found to have colorectal cancer, and 40% were found to have a hematologic malig-
nancy. Those survivors of *C. septicum* gas gangrene who are found to be immunocompetent will produce antibodies to α-toxin.

The classic presentation of gas gangrene is that of severe pain and underlying crepitus. However, these are frequently late symptoms; early findings are often nonspecific. Other symptoms include a sudden onset of fever, abdominal pain, vomiting, diarrhea, and rapid progression to shock. Up to 25% of patients have myonecrosis at metastatic sites.

The sudden onset of severe pain is often the first symptom experienced by the patient. Typically, the pain is out of proportion to physical examination findings and is minimally relieved by medications. Initial physical examination of the site may be normal; within minutes to hours, localized tense edema, pallor, and tenderness are seen.

Systemic findings include diaphoresis, tachycardia disproportionate to temperature, and extreme anxiety on the part of the patient, who often remains exceedingly alert until the very terminal stages. Late complications include intravascular hemolysis, hemoglobinuria, hypotension, renal failure, and metabolic acidosis. The disease progresses rapidly over the course of hours and is frequently fatal.

The severity of the pain and presence of systemic toxicity help to differentiate *Clostridial* myonecrosis from other soft tissue infections including necrotizing fasciitis and crepitant cellulitis. Necrotizing fasciitis is frequently caused by group A β-hemolytic *Streptococcus*, but polymicrobial infections are also common. It is characterized by a rapid progression and significant systemic toxicity. Skin findings are often present early in the course of illness, and pain is present but to a lesser degree. Similar to myonecrosis, crepitant cellulitis can be caused by *Clostridium* species; however, there are minimal skin changes, pain, and systemic toxicity associated with this disease process. Aggressive surgical debridement is the cornerstone of treatment for all of these soft tissue infections.

The diagnosis of gas gangrene is based on clinical findings, demonstration of myonecrosis at surgery, and supporting microbiological data. Bacteremia is documented in only 10% to 15% of patients with clostridial myonecrosis. Ultimately, gas gangrene is a surgical diagnosis made when involved muscle is visualized. The extent of myonecrosis is often greater than the skin changes indicate.

Treatment includes early operative debridement and antibiotic therapy. Because of the polymicrobial nature of some necrotizing soft tissue infections, empiric antibiotic coverage is implemented. Antibiotic combinations used to treat gas gangrene usually include penicillin and clindamycin. Although high-dose penicillin was once the drug of choice for clostridial infections, it has been found to be less effective in severe disease. Clindamycin is beneficial because it has the ability to reduce the production of exotoxins produced by *Clostridium* species.

In some instances, hyperbaric oxygen therapy is also useful. The rationale for the use of hyperbaric oxygen includes the observations that raising local tissue oxygen concentrations arrests clostridial replication and α-toxin production and that hyperbaric oxygen treatment of experimental animals reduces mortality. The use of hyperbaric oxygen therapy remains controversial. Clinical experience with hyperbaric oxygen therapy suggest that, when used in conjunction with surgery and antibiotics, it may reduce morbidity and mortality in cases of clostridial infection. Hyperbaric oxygen therapy, however, is less useful in cases of *C. septicum* than *C. perfringens* because of the aerotolerant nature of this species.
The progression of *C septicum* spontaneous gas gangrene may be even more fulminant than that of traumatic *C. perfringens* gas gangrene. Despite optimal management, the mortality rate is 67% to 100%, with most dying within 24 hours after onset.\textsuperscript{5}

This case of gas gangrene in a healthy adolescent demonstrates that soft tissue infection can occur in the absence of precipitating events and risk factors. In summary, nontraumatic gas gangrene is a rare but life-threatening disease for which physicians must retain a high index of suspicion. Early and aggressive treatment is essential to reduce the mortality associated with this infection.

**REFERENCES**

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*Pediatrics* 2005;116;e735
DOI: 10.1542/peds.2004-2876 originally published online September 30, 2005;

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