

Unprotected People #43 Varicella (Chickenpox)

Periorbital varicella gangrenosa necessitating orbital exenteration in a previously healthy adult

We usually try to choose stories that are written for a general audience. Sometimes, however, a powerful story appears in a technical medical journal that we think almost all readers can appreciate. We believe that today's story about an adult case of varicella infection is indeed powerful. For those who don't have a medical dictionary handy, "orbital exenteration" means complete removal of the contents of the eye socket.

Most of us think of chickenpox, or varicella infection, as a childhood disease. It's true that many more children than adults get chickenpox. However, a higher proportion of adults than children die from chickenpox or complications from the disease. The following medical case report tells us about one adult male, previously healthy, who apparently caught chickenpox from his infected children. He died after a year of suffering from pneumonia and severe group A Streptococcus infection that led to varicella gangrenosa, a form of necrotizing fasciitis.

Adults, if you have not had chickenpox in the past, it's not too late to get vaccinated now. Ask your doctor.

"Periorbital Varicella Gangrenosa Necessitating Orbital Exenteration in a Previously Healthy Adult" was written by William O. Thomas, M.D.; James A. Parker, M.D.; Bonnie Weston, M.D.; and Christine Evankovich, M.D. The full text, excluding two figures, is republished here by permission of the Southern Medical Association, which originally published this article in July 1996 in the Southern Medical Journal. (Copyright Southern Medical Association.)

Abstract

A previously healthy 31-year-old man had profound neurologic compromise and necrotizing periorbital infection due to a complication of varicella infection. Despite aggressive treatment, he required orbital exenteration and radical debridement of the involved tissues. He survived in a vegetative state for

almost 1 year before succumbing to progressive neurologic deterioration. We present and discuss this complicated case of varicella gangrenosa to show the devastating nature of complications that can occur from varicella infection in the unsensitized adult.

Although varicella is generally considered a benign disease of childhood, complications can occur. Our report of devastating complications from varicella infection in a previously unsensitized adult is presented to alert the readership to the potential for development of varicella gangrenosa in adults who have chickenpox.

Case Report

A 31-year-old white man was seen initially at an outlying hospital, where a diagnosis of chickenpox was made. His children had recently been infected with chickenpox, and he had no antecedent history of having had the disease as a child. Previously, he had been otherwise healthy. Despite conservative measures, he continued to have high fevers and went to a local emergency room with evidence of dehydration and septicemia. While he was in the emergency room, neurologic deterioration and coma developed. The patient was transferred to the University of South Alabama and was admitted to the medical intensive care unit, where mechanical ventilation was begun because of pulmonary insufficiency and respiratory arrest due to sepsis and airway swelling. He had hypotension with clinical signs of septicemia, shock, renal failure, and encephalitis and was treated with intravenous acyclovir (800 mg every 12 hours) and appropriate dosages of nafcillin, ceftazidime (Fortaz), and metronidazole for broad spectrum antibiotic coverage because of the renal compromise. Evaluation in our institution showed necrotic eyelids, as well as significant surrounding facial and neck erythema and cellulitis.

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Blood cultures and vesicular cultures were positive for group A [beta]-hemolytic *Streptococcus* sp. Computed tomography of the neck, face, and brain showed significant diffuse brain swelling and cerebritis with impending herniation. Neurosurgical consultation was obtained. High-dose steroid therapy was recommended and instituted to prevent herniation from the brain swelling and edema. The plastic surgery service saw this patient in consultation with the ophthalmology service for evaluation of the periorbital swelling and lid necrosis, and immediate operative debridement was scheduled.

After neurosurgical clearance, the patient was brought to the operating theater for debridement of the necrotic lids and decompression of the tense fluid accumulation in the neck. At the time of debridement of the necrotic lids, we found subcutaneous fat necrosis with some purulence throughout all planes of the incised left side of the face. Frozen section examination of debridement specimens was negative for mucormycosis. Additionally, there was some necrotic bone of the left zygoma, which was also debrided. The left side of the neck was incised, yielding about 500 mL of serosanguineous fluid from the left and 300 mL of fluid from the right side. The left globe appeared marginally viable but was left in situ at the initial debridement, in hopes that the retinal arterial blood supply would allow preservation of left eyesight. After the left globe was observed for a few more days, it became obviously necrotic. Orbital exenteration was recommended and done along with further debridement of the periorbital structures, which had developed further gangrenous changes despite our initially debriding back to bleeding apparently viable tissues, including periorbital bone. Wound cultures were positive also for group A [beta]-hemolytic *Streptococcus* sp.

The patient continued to fare poorly with signs and symptoms of continued sepsis and was maintained by vigorous supportive measures including tube feeding, mechanical ventilation, a pressure prevention bed, and wet-to-dry dressing changes for the orbital exenteration defect. A left frontal lobe brain abscess developed and was drained by stereotactic craniotomy. The neurologic status did not improve,

and the patient remained in a vegetative state with essentially no organized cortical brain wave activity on serial electroencephalograms. Pulmonary support was difficult but aided by tracheostomy for pulmonary toilet, and with frequent suctioning he survived.

Intermittent pneumonia and atelectasis of both lungs with low systemic arterial oxygen saturations precluded a safe anesthetic for the performance of additional elective operations. Approximately 3 months after hospital admission, he was transferred to a chronic care and rehabilitation facility with the left periorbital wound manifesting continued contraction and slow wound healing. The periorbital wound epithelialized eventually without requiring skin grafting. He survived in a vegetative state at the rehabilitation facility with no sign of any higher cortical function and died approximately 1 year after the onset of his illness.

Discussion

Varicella is generally considered a benign disease in childhood. In a 1935 study of hospitalized children, a 5.2% complication rate was reported among more than 2,500 patients with chickenpox.[1] In ambulatory patients, the complication rate must be significantly lower. In adults, the complication rate of varicella infection is severalfold higher, and results of these complications tend to be disastrous for the afflicted patient, as this case illustrates.

Secondary infections or superinfections of the skin are the most common complication of varicella viral infections and include abscess, lymphadenitis, cellulitis, erysipelas, and gangrene. Group A [beta]-hemolytic *Streptococcus* is the most commonly isolated bacterial pathogen contributing to those complications.[2] Gangrenous complications of varicella occur at an estimated frequency of only 0.05% to 0.16% of cases.[3] The original description of this dreaded complication is credited to Stokes,[4] who in 1807 reported on an eruptive disease of children. Hutchinson[5] coined the term varicella gangrenosa, a more apt description of this morbid and potentially fatal complication of varicella infection. Varicella gangrenosa likely represents one of the many forms of necrotizing fasciitis consequent to subcutaneous

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streptococcal infection that can be seen in clinical practice. Such cases of fasciitis are variously called Fournier's gangrene, synergistic gangrene, and Meleny's gangrene. Differentiation of this entity from mucormycosis, which can produce rapidly progressive gangrenous changes in patients with diabetes, is important. Antifungal therapy with intravenous amphotericin B would be indicated if hyphal elements were found on frozen or permanent sections of involved necrotic tissue.[6]

Oral and intravenous acyclovir may reduce the incidence of complications if given early in the course of the disease in immunocompetent hosts, but particularly in immunocompromised patients.[7] Ideally, therapy with acyclovir should begin as soon as possible after the first vesicle appears at a dose of 800 mg orally five times a day for 7 days in uncomplicated varicella. If patients manifest complicated varicella, intravenous acyclovir should be administered at a dose of 10 mg/kg for 7 to 10 days. Our patient had multiple complications, including cerebritis, encephalitis, brain abscess, and facial/orbital necrosis despite aggressive intravenous treatment with acyclovir and broad spectrum antibiotic medications before and after surgical treatment.

The early surgical management of our case with eyelid and soft tissue debridement did not prevent ongoing damage to the vital eye structures despite subfascial debridement, which included some bony structures. The secondary debridement necessary in this case bespeaks of the potent effects of subintegumentary infection with group A [beta]-hemolytic streptococci complicating varicella viral infections. Poor penetration of antimicrobial medication into the areas of relative hypovascularity may explain the poor response of this infection to traditional treatment with intravenous antibiotics, and therefore aggressive surgical debridement is mandated. Earlier diagnosis and surgical therapy with

prompt debridement may help reduce the extreme morbidity and potential mortality incurred by varicella gangrenosa. Our patient's severely compromised neurologic status precluded safe skin grafting or periorbital reconstruction with free tissue transfers. The disfigurement induced by debridement of gangrenous periorbital structures is probably best remedied by manufacture of a prosthesis for aesthetic considerations.

For more information on varicella, see IAC's Varicella Information web page at:<http://www.immunize.org/genr.d/varicel.htm>

To see the Centers for Disease Control and Prevention (CDC) Varicella and Herpes Zoster webpage, go to: <http://www.cdc.gov/nip/diseases/varicella/faqs-clinic-disease.htm>

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