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Varicella Gangrenosa of Abdominal Wall: Rare but Fatal Complication of Varicella Even in Immunocompetent Healthy Children

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Introduction: Varicella gangrenosa is an uncommon but serious complication of chicken pox in young children. It should be suspected in any child with a history of varicella infection and increasing complaints of pain and swelling in an extremity or other body area, along with increasing fever, erythema, lethargy, and irritability. Early surgical intervention with intensive antibiotic therapy is essential to prevent fatal consequences.

Case Presentation: We describe a case of a previously healthy child who presented with sepsis due to varicella gangrenosa. While she initially responded well to a conservative antibiotic and acyclovir treatment, her subsequent rapid deterioration required urgent and repeated debridement.

Conclusions: This report highlights the significance of prompt diagnosis and early surgical intervention for management of varicella gangrenosa.

Keywords: Debridement; Fasciitis; Sepsis

1. Introduction

Varicella, commonly known as chickenpox, is a common disease of childhood that results from primary infection with varicella zoster virus. It starts with the appearance of a characteristic exanthema and symptoms including mild fever, malaise, nausea, and headache. Varicella usually has a benign course; however, complications may occur and can be life threatening, causing sepsis, osteomyelitis, encephalitis, and skin and soft tissue infection. Dermatological complications are commonly superficial superadded bacterial infections, sometimes leading to necrotizing fasciitis, varicella gangrenosa, and hemorrhagic chicken pox. Although the incidence of varicella complications is several-fold higher in adults, adolescents, and immunocompromised children, healthy immunocompetent children may also experience complications related to varicella (1). Hospitalization rates for varicella are about 3 - 6 per 1,000 cases, and the complication rate ranges between 2% and 4%. Mortality rates are between 0 and 0.05 deaths/100,000 population per year in Europe (2). Rivest et al. (1), Choo PW et al. (3) reported complication rates of 29.2 cases/10,000 cases of varicella and 0.82 cases/100,000 children/year, respectively, including necrotizing fasciitis and other skin infections, pneumonia, bacteremia, encephalitis, ataxia, aseptic meningitis, Guillain-Barre syndrome, hepatitis, thrombocytopenia, dehydrations, febrile convulsions, congenital varicella, disseminated intravascular coagulation [DIC], stomatitis, synovitis, purpura fulminans, empyema with bacteremia, and death. Varicella gangrenosa is a type of necrotizing fasciitis; it is a rare but life-threatening dermatological complication with a frequency of 0.05% - 0.16% (4). We report a case of previously healthy 5-year-old female child with varicella who developed varicella gangrenosa associated with severe sepsis, pneumonia, severe hypoalbuminemia, and thrombocytopenia.

2. Case Presentation

A previously healthy 5-year-old girl was admitted five days after she developed a typical chickenpox rash. She was referred to our hospital for high fever and necrosis of several skin lesions. On examination, the child had high-grade fever with rigors and chills, lethargy, irritability, and a large (5-cm diameter) black necrotic patch with marked erythema, swelling, and pain on the left lateral side of her abdominal wall. Varicelliform lesions were present over her face, scalp, and extremities; several were purplish and surrounded with diffuse erythema. The child appeared sick and was febrile and lethargic, with a respiratory rate of 20 breaths/minute, heart rate of 128 beats/minute, good pulse volume, and a blood pressure of 110/68 mmHg. Blood tests revealed a hemoglobin concentration of 10.8 g/dL, a total leukocyte count of 30,000 with 90% polymorphs, 178 platelets/nL, and C-reactive

protein concentration of 88 mg/dL. Coagulation studies and chest X-ray were normal. The child was admitted to the pediatric intensive care unit and managed conservatively with broad-spectrum intravenous antibiotics, acyclovir, intravenous fluids, and local wound care. The child initially responded well to treatment, with improvement in her general condition, decreased fever frequency and intensity, and drastic decrease in total leukocyte count (14,000) and C-reactive protein level (27 ng/dl). However, 48 hours after this initial improvement, her high-grade fever returned, along with respiratory distress, generalized anasarca, and increasing pain and swelling over her trunk. On examination, the child was lethargic and appeared ill, in severe respiratory distress with bilateral crepitations and decreased air entry on the right side on auscultation associated with marked edema all over her body. There was increase in her erythema and the swelling around the necrotic patch, with crepitus and blister formation. Repeated blood investigations revealed leukocytosis, with a total leukocyte count of 49,000 with 91% polymorphs, a platelet count of 16/nL, hemoglobin (Hb) concentration of 6.0 g/dL, C- reactive protein concentration of 102 mg/dL, and serum albumin levels of 1.6 mg/dL. A coagulogram and liver function tests were within normal range; however, a chest X-ray showed bilateral pneumonia with right-sided pleural effusion. Blood and swab cultures were sterile. The child was managed with intensive intravenous antibiotics and non-invasive ventilation; she was also supported with packed cells, single donor platelets, and albumin transfusion. Urgent surgical consultation and emergent debridement were performed. The child improved dramatically after debridement; however, repeated debridement was necessary, which required later skin grafting. Initial blood and swab cultures all were sterile. The child was discharged after the 14th day of admission.

3. Discussion

Chickenpox caused by varicella zoster is an extraordinary contagious childhood illness. Concomitant bacterial infection is a common complication in previously healthy children with chickenpox. Varicella gangrenosa is an uncommon but serious complication of chickenpox infection in young children (5, 6). It constitutes less than 1% of cases. In 1881, Jonathan Hutchinson first described the condition and coined the term 'varicella gangrenosa' (7). Its incidence however, has been increasing in recent years due to invasive *Streptococcus pyogenes*. The most common sites of initial involvement in adults are the lower extremities; however, in the pediatric population, it is most commonly reported in the abdominal wall, followed by the gluteal region and thighs, head and neck, and upper and lower extremities. The initial skin presentations include induration or cellulitis with progression to skin discoloration and bullae formation. Intense pain out of proportion with skin findings, leukocytosis, elevated C-reactive protein levels, fever, and tachycardia are important associated clinical findings. Ulcerations, ecchymosis, crepitus, anesthesia, and necrosis are indicative of advanced disease.

Three types of varicella gangrenosa have been described in the literature: [a] moist gangrene, which is believed to be infective; [b] dry gangrene, secondary to arterial thrombosis; and [c] purpura fulminans, associated with DIC (8). Moist gangrene is characterized by erythema that changes within 24 - 48 hours into wet, blue lesions. Lesions can appear as small as spots arranged diffusely on the body and rapidly progress into larger, crusted lesions. It can be difficult to distinguish moist gangrene from other skin infections. However, some subtle differences offer diagnostic clues. Dry gangrene generally involves the tips of fingers and toes and is characterized by cool, dry, discolored lesions. Purpura fulminans is characterized by thrombocytopenia and bleeding of mucous membranes and gastrointestinal tract. Necrotizing fasciitis is characterized by red/violet, swollen skin with crepitus, and blisters may form. The most typical feature is pain out of proportion to the physical findings. There is no evidence that laboratory tests or imaging techniques (computed tomography [CT] and magnetic resonance imaging [MRI]) are useful for establishing a diagnosis, and have not been associated with decreased morbidity or mortality associated with these skin infections. Varicella-associated gangrene generally presents between the third and seventh days from the day of eruption. It is characterized by pain and erythema of the affected part, which changes within 24 - 48 hour into wet, blue lesions and is associated with increasing fever, erythema, lethargy and, irritability. Severely affected patients may present with severe sepsis, septic shock, DIC, thrombocytopenia, and multiple organ dysfunction syndrome [MODS].

Group A beta hemolytic streptococcus [GABHS] and *Staphylococcus aureus* are the major causative organisms. GABHS manifests as deep-seated infections and is often the most severe and difficult to treat (9). Several factors contribute to the pathogenesis of necrotizing fasciitis in children with chickenpox. The vesicle creates a full-thickness dermal lesion that provides a route for bacteria to spread from the skin surface to the subcutaneous tissues (10). GABHS has numerous virulence factors. The M surface protein repels macrophages and complement factors. Streptococcal pyogenic exotoxins A, B, and C initiate release of tumor necrosis factor-α and interleukin-1, resulting in fever, rash, direct toxic effects to the endothelium, septic shock, and T-cell activation. Streptococcal M protein and exotoxins A and B have been reported to be associated with more severe infections (11). The microorganism also secretes hyaluronidase and streptolysin, which lead to tissue invasion and damage. Surgery is necessary to remove necrotic tissue, which is the source of exotoxins that may cause circulatory collapse, multiple organ failure, and eventually lead to death (12), Deep-seated infections are associated with thrombocytopenia,

bacteremia, persistent fever, prolonged hospitalization requiring intensive care management, and the potential for fatal outcomes. Diagnosis of complications secondary to chickenpox in children may be difficult in the early stages of disease. Contributing factors include high, persistent fever; recurrent fever after an afebrile period; localized swelling; induration; erythema; disproportionate pain; lethargy; refusal to bear weight; toxic appearance; hypotension; and tachycardia. Necrotizing fasciitis can be distinguished from cellulitis on the basis of diffuse erythema, toxic appearance, fever, and thrombocytopenia. Once diagnosis of varicella gangrenosa is suspected, intensive care and early and aggressive surgical debridement with appropriate antibiotics is warranted (12, 13). In one large published series, diagnosis of necrotizing fasciitis was missed in 85% - 100% of the reviewed cases due to the paucity of cutaneous findings early in the disease course, which resulted in significant morbidity and mortality rates of approximately 25% - 40% (14). Early recognition, emergent surgical debridement, and intravenous antibiotics are essential to prevent significant morbidity and mortality.

In the present case, a 5-year-old female child presented on the seventh day after eruption with high-grade fever, irritability, pain, and a large necrotic patch surrounded by marked erythema on her trunk. The child's condition was initially managed and she responded well to conservative treatment with intravenous antibiotics, acyclovir, and local wound care. However, after her initial improvement, she again developed high-grade fever with increasing erythema, blister formation, and crepitus. These symptoms were also associated with severe respiratory distress, severe thrombocytopenia, and hypoalbuminemia. The child was managed with urgent surgical debridement, intensive intravenous antibiotics, and also was supported with non-invasive ventilation and blood products. This case emphasizes the need for prompt diagnosis of necrotizing fasciitis secondary to varicella infection even in immunocompetent children; furthermore, this diagnosis requires early and urgent surgical interventions along with administration of intravenous antibiotics and acyclovir in order to prevent fatal consequences.

The complications we have described are preventable with vaccination for chickenpox, which has been shown to be effective in preventing and decreasing infection severity. Numerous studies have advocated vaccination to prevent infection and consequently eliminate chickenpox complications (11, 15, 16). Vaccination should therefore be standard practice since chickenpox complications seem to be increasing in severity, with an increased frequency of GABHS infections. Current guidelines recommend vaccination for varicella in all children between 12 and 18 months of age and in children between 19 months and 12 years of age who have not previously been vaccinated or who have no history of varicella-zoster infection.

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Authors' Contributions

All authors had equal roles in study design, practical work, and manuscript writing.

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