Plastic surgery management in pediatric meningococcal-induced purpura fulminans

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Purpura fulminans is a rare but potentially devastating complication of septic shock caused by viral, rickettsial, or bacterial infection. In children, the most common causative organism is Neisseria meningitidis, an aerobic Gram-negative encapsulated diplococci [1]. It occurs first as a petechial rash that spreads rapidly. Once sepsis occurs, the skin rash quickly evolves into a full-thickness skin necrosis. Together with the disseminated intravascular coagulopathy (DIC), the necrosis can extend beyond the skin into the soft tissue and bone.

Children with these severe clinical presentations have had high mortality—up to 80% in several studies [2,3]. It is only in recent times, with the advances in critical care and antibiotics that allow these patients to survive, that concerns about their quality of life have become an issue. Plastic surgeons are frequently consulted regarding management of the soft tissue or the open wounds once debridement or amputation has been performed [4].

This article reviews the pathophysiology of meningococcal purpura fulminans and suggests a strategy for managing the wounds in these difficult cases.

Background

N meningitides is commonly present in the nasopharynx of asymptomatic adults. Thirteen specific serotypes exist; serotype A is most common worldwide, whereas serotypes B and C account for most cases in the United States. The organisms become a pathogen in susceptible individuals, such as the young or the immunocompromised, by penetrating the oral or nasal mucosa [5].

The disease can manifest itself in meningitis, pericarditis, pneumonia, or arthritis. It can also occur in a mild form of acute meningococcemia characterized by malaise, myalgia, fever, and diarrhea. A rarer chronic form can last several weeks with migratory symptoms.

Fulminant meningococcemia is most severe and is associated with soft tissue necrosis. It occurs in approximately 10% of all patients with meningococcemia [6]. It presents with high fever, chills, severe myalgia, headache, and mucosal petechiae. These symptoms can progress rapidly and quickly lead to septic shock with hypotension, DIC, and severe Adult Respiratory Distress Syndrome (ARDS). The skin manifestations worsen to purpura and frank tissue necrosis (Fig. 1). This whole cascade of events has been termed Meningococcal Septic Shock (MSS) [7].

Children with MSS have the best chance for an optimal outcome in a critical care center. The care of this complex and difficult situation is best managed by a team that includes specialists in critical care medicine/ pediatrics, infectious disease, plastic surgery, orthopedic surgery, physical medicine/ rehabilitation, psychology, and nutrition service, along with dedicated nurses. Aggressive treatment has improved survival rate. Antibiotics (penicillin is still the mainstay when the organism is isolated) and inotropic and ventilatory support during the early critical period have been shown to save lives [8]. During this time, the skin and soft tissue damage is evident but may...
Pathophysiology of purpura fulminans

Hemorrhagic, purpuric skin lesions with grayish centers associated with pustules or bullae are typical with MSS (Fig. 2). In the animal model (rabbits), the Shwartzman reaction—in which localized purpuric skin necrosis occurs after exposure to antigens—can mimic the skin purpura fulminans of MSS [9]. When this is associated with massive adrenal hemorrhage, it is known as the Waterhouse-Friderichsen syndrome, which has a mortality of close to 100%. Purpura fulminans has been shown to correlate with an acquired decreased level of proteins C and S, which, together with the DIC state, leads to a hypercoagulable state in the small vessels and capillaries [10].

N meningitides also releases a powerful endotoxin that causes inflammatory endothelial damage, increased vessel wall permeability, microthrombi, and ultimate occlusion. The powerful inotropic agents used to maintain the blood pressure of critical patients further contribute to tissue ischemia.

Fig. 2. Hemorrhagic, purpuric skin lesions with pustules and bullae on the right upper extremity of a child with meningococcemia.

The affected areas tend to be those of less perfused skin, such as the extremities. The lower extremities tend to be more severely affected (Fig. 3). The affected areas of the upper extremities are more distal and may not follow any vascular pattern. Occasionally, the soft tissues of the face or trunk are affected. The tissue losses not only affect the superficial and dermal skin layers but can also extend deeply into muscles and bones [11].

General care

Discussions should be held with the critical care specialists or team in charge. The following suggestions should be considered and instituted if agreed on by the team. They are listed in order of their relative effectiveness (with regard to tissue preservation) and of their reward-to-risk ratio as documented in the literature:

Use more cardiac inotropic and fewer vasoconstrictive agents to maintain adequate perfusion.
If possible, add vasodilators to augment tissue oxygenation [12].
Maintain normovolemic or even slightly hypervolemic state if pulmonary status is stable.
Administer purified protein C. One study [13] has reported the correction of organ dysfunction in all four patients after receiving protein C, with two patients suffering no tissue loss. Given the widely accepted premise that decreased protein C level contributes to the hypercoagulable...
Institute blood or plasma pheresis. This procedure can improve DIC and vessel-wall integrity by removing the circulating inflammatory cytokines. Its effectiveness is anecdotal, but the risk is low [14,15].

Use hyperbaric oxygenation. Improvement of limb perfusion with this treatment has been reported [16]. The need for separation or isolation in the treatment chamber during the critical illness poses a danger.

Administer steroids. Their use has limited the hearing loss in meningitis, but it has not proved beneficial in limiting the skin loss [17]. Steroid administration in septic shock remains very controversial.

Infuse heparin. Even though heparin can decrease and mitigate the damage of vascular occlusion, the attendant risk of bleeding (already increased in septic patients) renders this treatment less acceptable [18].

Other investigational therapies, such as the administration of prostacyclin, tissue plasminogen activator, sympathetic blockage, topical nitroglycerin, or extracorporeal membrane oxygenation, have not proved very effective [19]. They also have significant potential complications.

Local care

In general, affected limbs should be elevated to decrease edema and carefully examined for compartment syndrome. This measure is especially important if the wound is circumferential or if digital examination reveals significant edema. Classically, compartmental pressure measurements of 30 mm Hg or greater have been used to indicate the need for fasciotomies. However, the mean arterial pressure must be considered; in hypotensive patients, compartment syndrome may result with intracompartmental pressures of less than 30 mm Hg. One should have a low threshold for compartment release in these patients.

The affected skin should also be examined for purulence. If purulence or abscess is present, the debridement should be performed early when the medical condition permits. During the period before debridement, the purulence should be opened to drain as much as possible, and dressing changes should be initiated. After debridement, the subsequent open wounds can be treated with wet to dry dressing changes until coverage is performed.

When there is no purulence, and the skin is ischemic but not frankly necrotic (ie, purpuric skin), the authors recommend local skin care with an antibiotic ointment.

When there is eschar or skin necrosis without purulence (ie, dry gangrene), then the skin should be treated with a bactericidal solution such as diluted Betadine (Purdue Frederick Co., Norwalk, CT) or Dakin’s solution to decrease the risk of infection. The nonviable area should be allowed to demarcate before debridement. Patients with aggressive, early debridement have been shown to have a more proximal amputation level and more need for repeat surgeries than those who are allowed to wait [20,21]. The questionable areas are given a chance to recover as long as there is no active infection.

However, there is a possible benefit to early debridement of dry necrotic tissue when present in large volume: the improvement of renal function. One report exists of renal improvement after debridement of a substantial volume of nonviable tissue—an arm and a leg—in a pediatric patient [22]. This effect may be secondary to the inflammatory byproducts associated with necrotic tissue. It should be kept in mind if the patient’s renal function continues to deteriorate despite optimal medical management.

The use of leeches can be helpful to salvage digits. The Hirudin effect can vasodilate blood vessels and locally decrease the hypercoagulable state, thereby increasing blood flow [23]. This treatment would be most appropriate in a patient with isolated digital ischemia.

Progressive management

After the patient has recovered from the initial critical period and the medical condition has stabilized, surgical management of ischemic tissue and open wounds can become more aggressive. The goals are still to preserve as much tissue as possible, especially the joints.

Debridements are performed to remove the nonviable skin, muscle, and bone. Even at this stage, the margins of tissue survivability can be difficult to delineate with precision. MRI, with or without concomitant angiography, can give the surgeons an approximate level of debridement or amputation [24]. During surgery, intraoperative muscle or bone biopsy can demonstrate viability of tissue. The authors’ strategy is to be aggressive with debridement of skin and
soft tissue (with the exception of the face), but more conservative in the resection of functional bone length and articular surfaces.

Cutaneous defects can be covered with skin grafts. However, there are reports of graft failure when grafting is performed in the early clinical course, even when the recipient beds appear ready [25]. This failure could be due to residual inflammatory response or to continued perfusion dysfunction. In the early period, when questions of tissue perfusion persist, but coverage is needed to assist in the fluid, electrolyte, and nutritional management, the authors have found that allograft skin can serve very well as a temporary physiologic dressing [22]. The allograft skin can be maintained for weeks at a time. The attachment of allograft skin can also signal the readiness of the wound to accept an autologous skin graft.

More complex defects with exposed bone or joint will require coverage with flaps. The timing of reconstruction is important. As with skin grafts, reports exist of early flap failure with both local and free tissue transfer [4,26]. Based on the literature, the authors believe that the chance of success can be increased by postponing the major surgery until recovery of local tissue perfusion and vascular integrity has occurred. This recovery may take up to 4 weeks after the initial injury. At this time, the patient should have a normal blood pressure without pressor support, and the clinical condition should be stable both in terms of infection and tissue loss. During this waiting period, careful dressing changes or temporary allograft skin coverage can keep the exposed bone and joint protected. Local and free vascularized muscle, musculocutaneous, or fascial flaps have been used with success at the authors’ center and at other institutions [4,27].

Late management

Most of these patients require extensive rehabilitation and further surgical revision. One of the late sequelae of the disease is the damage to the epiphyseal growth centers of the long bones. This damage may result in asymmetric growth that will require orthopedic intervention [28]. Alternatively, the bone growth may stretch the overlying soft tissue coverage and necessitate revision. The treatment of these problems should be individualized to allow the child to lead a normal life.

Scar contracture is another frequent problem. Early treatments include pressure garments and physical therapy. Surgical scar release is frequently required and should be done early if the contracture is across a joint, so that physiotherapy can be started early. Scar release surgery can include simple procedures, such as Z-plasty to local flap rotation to free-tissue transfer [22]. The principles for surgical intervention in these patients are the same as in burn patients.

Surgical treatment for cosmesis becomes more important as the child grows. The benefits of these procedures are obvious, as demonstrated in the care of burned children. Major procedures, including the use of free flap, are indicated to allow these patients to wear a properly fitting, cosmetically acceptable prosthesis [29].

Growth and development patterns in surviving children are slow in the first 12 months after the injury. However, these patients rapidly catch up with their peers and can function well independently within their respective disabilities [21].

Summary

Purpura fulminans associated with meningococcal infection [29]. Growth and development patterns in surviving children are slow in the first 12 months after the injury. Many children survive the disease but suffer major morbidities such as amputation. The authors suggest a strategy to manage these wounds with the goal of preserving as much tissue and function as possible. At the present time, conservative therapy to the wounds appears to be the best course in the initial, critical phase, as long as no active local purulence is found. Debridement or amputation is performed when the nonviable tissue margins are delineated. Temporary coverage with allograft may be required, and definitive coverage is accomplished when the local tissue perfusion has recovered. Future revisions are often necessary and should be pursued aggressively to improve these children’s quality of life.

References


