Midface Necrosis From Purpura Fulminans Secondary to Streptococcus Pneumoniae Sepsis

Aaron D. Robinson, D. Gregory Farwell, MD; Quang C. Luu, MD, Levi G. Ledgerwood, MD
University of California, Davis Department of Otolaryngology – Head and Neck Surgery

ABSTRACT

Introduction: Purpura fulminans is a rare syndrome associated with consumptive coagulopathy leading to infarction of terminal vessels, especially in the skin. This leads to purpuric skin lesions that progress to necrosis of skin and subcutaneous tissues. Head and neck involvement is rare in this disease, but when there is involvement, the subsequent cosmetic and functional deficits are usually extreme. Reconstruction of these defects can be challenging when donor sites are also diseased.

A multidisciplinary team of otolaryngologists, speech and language pathologists, prosthodontists, and dentists is essential when helping patients recover after extreme facial resection.

CONTACT

Aaron D. Robinson, MD
University of California, Davis Medical Center, Department of Otolaryngology – Head and Neck Surgery
Email: aaron.robinson@ucdmc.ucdavis.edu
Website: http://www.ucdmc.ucdavis.edu/otolaryngology/
Phone: (916) 734-2801

Case Presentation

A 42 year-old, previously healthy man presented to the emergency department after a 3-day history of generalized malaise, fevers and myalgia. He notes his children had recently been diagnosed with pneumonia. He became concerned when he noticed a purple rash developing on his arms and legs.

In the emergency department the patient became hypotensive, was found to be severely hypoglycemic and became obtunded. He was intubated and admitted to the intensive care unit (ICU). Initial workup revealed severe leukocytosis, renal failure, lactic acidosis and blood cultures positive for growth of S. pneumoniae. He was managed medically for S. pneumoniae sepsis with broad-spectrum intravenous (IV) antibiotics, inotropes to address hypotension and aggressive fluid resuscitation. He was also started on a heparin drip to address DIC.

Pertinent physical exam findings included a diffuse purpuric rash with subsequent bullae formation and sloughing of the skin resulting in 90% total body surface area desquamation. The maxillary teeth were loose and the upper lip, nose, nasal septum and portions of the lower lip were necrotic. The tip of the uvula and posterior oropharynx, hard palate and distal tongue also had some degree of necrosis. (See Figure 1)

After initial resuscitation and optimal medical management, the patient underwent multiple excisions of necrotic tissue with cadaveric skin grafting, four limb amputations and two debridement procedures on the midface structures including a total rhinectomy, anterior septectomy, bilateral inferior turbinectomies, partial maxillectomy, excision of upper lip and partial excision of the lower lip. (See figure 2)

After the initial surgical debridement, the patient was managed in the ICU and continued on IV antibiotics. With no normal skin to use for local advancement flaps or axially based flaps primary reconstruction of the defect has been deferred. With concomitant coagulopathy, reconstruction with a free-tissue transfer was not an option. Currently the patient is recovering at home. He wears an obturating mask and will have a facial prosthesis and a permanent dental appliance placed to allow for oral competence and improved cosmesis. (See Fig 3)

Discussion

- Trip of symptoms:
  - Peripheral Purpura
  - DIC
  - Bacterial Sepsis

- Biopsy shows non-specific vasculitis and micro-emboli

- PT, PTT, fibrinogen, D-dimer, Protein C and S can help diagnose DIC, but are non-specific

- CSF and blood cultures isolate causative organism of sepsis and direct therapy

Management

- Initial management:
  - Introtropic support
  - Ventilator management
  - Fluid resuscitation
  - Early, broad-spectrum antibiotics to treat underlying cause

- Treatment of DIC

- Wound care:
  - Silver sulfadiazine, topical antibiotic ointment
  - Surgical debridement of necrotic soft tissue

- Reconstruction:
  - Few donor sites for reconstruction available due to destructive nature of disease
  - Healing by secondary intention
  - Split-thickness or full-thickness skin grafts
  - Local tissue advancement
  - Axially based forehead flaps
  - Free tissue transfer

Conclusion

Purpura fulminans is a rare, but devastating disease. Head and neck involvement is rare in this disease, but when it is present, the subsequent cosmetic and functional deficits are usually extreme. Reconstruction of these defects can be challenging when donor sites are also diseased. A multidisciplinary team of otolaryngologists, speech and language pathologists, prosthetists, and dentists is essential when helping the recovery of these patients.

References