



Salvage of an Immunosuppressed Girl with Necrotizing Fasciitis by Serial Soft-Tissue Necrosis Excisions and Immediate Coverage by Integra

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Abstract

We describe the presentation, diagnosis and management of an 8 year old immunosuppressed girl with extensive necrotizing fasciitis (23% TBSA); this is the first case report describing a step by step procedure of carefully planned life-saving surgical treatment of debridement and immediate coverage of dermal-substitute Integra with complete resolution with no complications

Keywords: Necrotizing fasciitis; Dermal matrix; Reconstructive surgery; Debridement; Immunosuppression

Introduction

Hirschsprung's disease is a congenital disorder involving bowel motility, characterized by signs of intestinal occlusion due to the presence of an aganglionic segment, variable in size, in the last portion of the colon. The annual incidence is 1/5000 of newborns [1].

The 22q11.2 deletion syndrome (Di George Syndrome) is due to a chromosomal aberration that causes a congenital malformation. The global incidence is 1/2000-1/4000 of newborns [1].

The syndrome consists of thymic aplasia, hypoparathyroidism and congenital heart diseases. Immunodeficiency affects up to 75% of children with Di George syndrome due to thymic aplasia/hypoplasia and consequently impaired T cell production and function. Rare but important complications of this syndrome include intestinal malrotation, imperforate anus, Hirschsprung disease.

Necrotizing Fasciitis (NF) involves the skin, subcutaneous tissue, deep fascia and muscle that appear with necrosis. The infection carries a high mortality rate. NF is classified after the causing microbial. Type I infections represent 70% of all NF cases and it is polymicrobial (*S. Aureus* or *Streptococcus*, Gram negatives, anaerobes). Its course is less violent with fewer systemic complications.

Type 2 is less common and is caused by mono microbial infections. The causative microbes can be *S. Pyogenes* and *S. Aureus*. Type II NF may be associated with toxic shock syndrome.

The diagnosis of NF is clinical (fever, tachycardia, diaphoresis); in children, it is often misdiagnosed as cellulitis [2]. The skin shows patchy ill defined margins discolorations with swelling and pain followed by tense edema, a grayish-brown discharge, vesicles, necrosis, and crepitus.

We report an eight-year-old girl with Di George Syndrome treated in intensive care unit for toxic shock syndrome and severe abdominal, chest wall, lower limbs and perineal region NF, resulting in massive soft-tissue necrosis, following surgical treatment of Hirschsprung's disease.

The importance of prompt carefully planned life-saving surgical treatment consisting of staged aggressive wound excisions (debridement of soft-tissue necrosis) and prevention of infection and fluid loss by evaporation from debrided areas, by immediate wound coverage with the dermal-substitute Integra are highlighted.

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Case Presentation

The patient is an 8-year old girl with Di George syndrome and a history of Hirschsprung's disease presenting with NF with extensive areas of soft-tissue necrosis involving the abdominal and chest wall, perineal and genital area, hips and internal portion of the thighs bilaterally (23% TBSA approximately) following intestinal resection and subsequent temporary colostomy.

The patient was hemodynamically unstable with raised blood white cell count and inflammation indices, for which she was admitted to the intensive care unit.

The cause and the evolution of NF are unclear; apparently, communication between the inside and outside occurred during bowel surgery with consequent bacterial colonization first and infection later.

After the patient's hemodynamic stabilization, it was decided to remove necrosis step-by-step with weekly debridements in the operating room.

At each surgery, a large piece of necrotic tissue was excised until vital bleeding tissue was reached, and the excised wound was covered immediately with double-layer Integra dermal matrix despite the proximity of necrotic, potentially infected tissues (Figure 1).

The use of Integra double-layer with its silicone membrane on the top of the dermal matrix was justified, in our opinion, because it minimized evaporative loss from raw surfaces, prevented contamination and infection from surrounding necrosis and allowed a fast vascularization of the dermal matrix followed by definitive wound closure by STSG after 14 days. Microbiological examination, revealed positivity for *P. Aeruginosa*, for which specific antibiotic therapy was set.

The patient got better after every debridement with a final resolution of the sepsis. The double-layer Integra gradually covered all excised areas, including the peristomal ones. Its silicone film, for its consistency, composition and hydrophobia, allowed fixation to the borders of ileostomy with 1.0 silk suture without fecal contamination of the wound bed. The patient underwent weekly dressing changes

in the OR under general anesthesia. There were no infections of the dermal matrix. After three weeks, the dermal matrix was progressively covered with STSG (meshed 1:1.5) taken from the lateral part of the thighs and legs. The graft take was 100%. During the last surgery, the stoma was repositioned by general surgeons to the left abdominal wall through healthy tissue, while the primary stoma breach was closed by a suture in layers, after which Integra covered it. Three weeks later, Integra was taken and was therefore covered with STSG from the left leg (Figure 2). After 26 operations, nine months of ICU hospitalization and several months of physical therapy, the patient was discharged completely recovered.

Discussion

The pathophysiology includes a local release of bacterial toxins and inflammatory cytokines provoking erythema, vasculitis, and thrombosis leading to tissue ischemia followed by tissue necrosis.

The inflammatory response then becomes systemic, leading to shock followed by multiorgan failure and death.

Two types of NF are known. Type I usually involve the abdominal wall, perineal and groin areas. Gram-positive or Gram-negative growth and anaerobes always cause it. Type II infections are usually caused by *Streptococcus pyogenes* infection. Our patient had a typical type I infection. In any case, once the blood and wound cultures have been obtained, broad-spectrum antibiotics should be administered. In our patient, NF resulted from a combination of shock and septic emboli secondary to surgery. It is possible that during surgery, there was bacterial colonization of soft tissue of the abdomen where the trocars were inserted for the laparoscopic operation, which explains *P. aeruginosa* positive microbiological culture from fluid collections and tissue obtained later.

Integra is the most widely used dermal regeneration template (Integra Life Science Corporation, Plainsboro, N.J.) [3]. It consists of a bilayer matrix composed of bovine collagen cross-linked with glycosaminoglycans from shark chondroitin sulfate with an overlying protective silicone layer. Integra's fibers form a collagen matrix with a specific pore size to maximize cellular in growth while inhibiting granulation tissue formation [4].

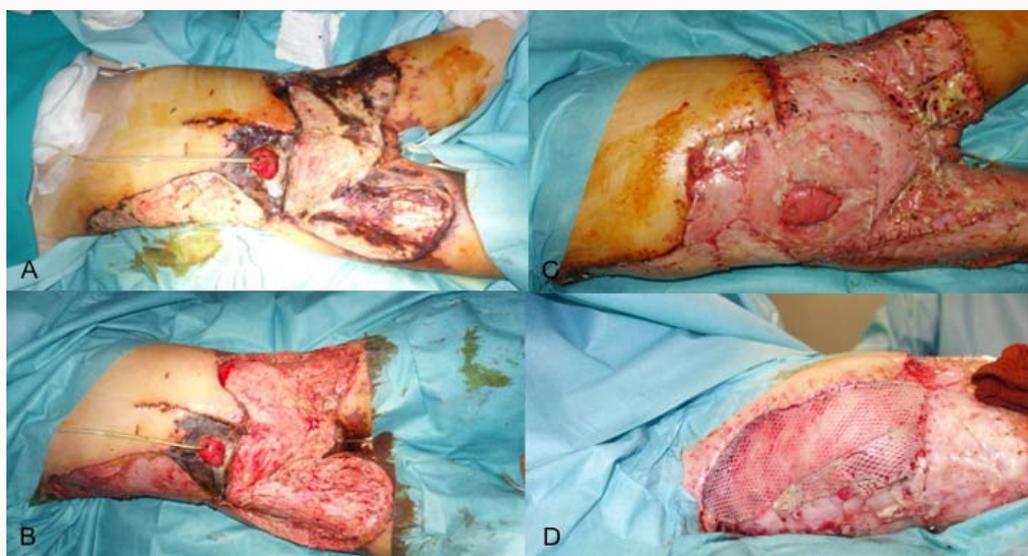


Figure 1: Before debridement (A), after debridement (B), after positioning of Integra (C) during positioning of STSG (D).



Figure 2: STSG completely taken (E), all STSG healed and Integra on the peristomal area (F), patient fully healed (G).

The matrix is covered with a semipermeable polysiloxane layer that serves as a temporary epidermis and after vascular in growth (2 to 4 weeks), separates, allowing a split-thickness skin graft to be applied.

Cases of NF in children have been described in the literature. They are caused by bowel perforation after abdominal surgery (abscesses, appendicitis, necrotizing enteritis). Hsin-Lin Tsai et al. [5] published a case report in which a 18 month-old child with necrotizing enteritis ended up in peritonitis followed by NF involving an extended area similar to ours. The patient was treated by serial debridement's and immediate positioning of Integra dermal substitute adjacent to the not removed necrotic tissue as in our patient. However, our patient was immunosuppressed and shocked.

In our patient, Integra placement avoided immediate skin grafting on muscle tissue leading to debilitating fixed scarring and lymphedema for lack of subcutaneous tissues. Moreover, its silicone membrane cover prevented the fluid loss, particularly important during the septic shock phase of treatment in the ICU and served as protecting barrier against the spread of bacteria from non-excised necrotic soft tissues.

Immediate split-thickness skin grafting was not indicated because of stoma in the middle of the abdominal soft-tissue defects that made the medications placement challenging.

The rationale of our step-by-step approach has developed around the following key points:

Integra can serve as an intermediate dressing in a patient with large wounds involving both skin and subcutaneous tissue, avoiding the negative pressure therapy (VAC) because of the wound size and the presence of the stoma.

Further, the silicon lamina of the Integra was used as a stoma attachment as it is waterproof and tight and allowed us to keep the stoma isolated from the wound and prevent feces contamination. The lamina also prevented contamination from outside, keeping the wound underneath isolated.

Once the Integra was taken, we proceeded to positioning split-thickness skin grafts using a carefully planned "step by step" procedure until all soft tissue defects were covered with a percentage of taking close to 100% and percentage of infection both of Integra and grafts close to 0%.

Conclusion

In conclusion, with this case we want to emphasize how it is possible to manage a huge loss of skin e subcutaneous tissues due to NF in children through an accurate timing of surgical therapy.

The step-by-step excision of necrosis was fundamental for minimizing the fluid losses and homeostasis in a patient after septic shock. Immediate wound coverage after excision of necrosis with Integra double-layer dermal substitute prevented wound infection. Furthermore, it enabled a 100% split-thickness skin graft take and good cosmetic result.

References

1. Orphanet.
2. Abass K, Saad H, Abd-Elsayed AA. Necrotizing fasciitis with toxic shock syndrome in a child: A case report and review of literature. *Case J.* 2008;1(1):228.
3. Gonzalez SR, Wolter KG, Yuen JC. Infectious complications associated with the use of integra: A systematic review of the literature. *Plast Reconstr Surg Glob Open.* 2020;8(7):e2869.
4. Papa G, Pangos M, Renzi N, Ramella V, Panizzo N, Arnez ZM. Five years of experience using a dermal substitute: indications, histologic studies, and first results using a new single-layer tool. *Dermatol Surg.* 2011;37(11):1631-7.
5. T sai HL, Liu CS, Chang JW, Wei CF, Lin JT, Chin TW. Severe necrotizing fasciitis of the abdominal wall secondary to colon perforation in a child. *J Chin Med Assoc.* 2008;71(5):259-61.