Actas Dermosifiliogr. 2013;104(7):623-627

ACTAS Dermo-Sifiliográficas

Full English text available at www.elsevier.es/ad

CASE REPORT

Purpura Fulminans Associated with *Streptococcus pneumoniae* Septicemia in an Asplenic Pediatric Patient ☆

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KEYWORDS
Purpura fulminans;
Hemorrhagic necrosis;
Streptococcus pneumoniae;
Sepsis;
Asplenia;
Immunosuppression;
Disseminated intravascular coagulation;
Thrombosis

Abstract Purpura fulminans is a rapidly progressive syndrome of small-vessel thrombosis and hemorrhagic necrosis of the skin accompanied by disseminated intravascular coagulation. We describe a case of *Streptococcus pneumoniae* septicemia in an asplenic 5-year-old boy on oral tacrolimus, with a past medical history of multivisceral organ transplantation and subsequent development of purpura fulminans on his chest and distal extremities. The acute infectious form of purpura fulminans is usually caused by gram-negative bacteria. Cases secondary to gram-positive encapsulated bacteria usually occur when individuals are immuno-suppressed or have anatomic or functional asplenia. Our patient had both, which likely increased his susceptibility, and he responded well to antimicrobial therapy in addition to prophylactic coverage in the setting of his immunosuppression. We review the literature for similar cases due to *S. pneumoniae* in the pediatric population and discuss the etiology and treatment of purpura fulminans.

PALABRAS CLAVE
Púrpura fulminante;
Necrosis hemorrágica;
Streptococcus pneumoniae;
Sepsis;
Asplenia;
Inmunosupresión;
Coagulación intravascular diseminada;
Trombosis

Púrpura fulminante en relación con septicemia por *Streptococcus pneumoniae* en un paciente pediátrico asplénico

Resumen La púrpura fulminante es un síndrome rápidamente progresivo de trombosis de pequeños vasos y necrosis hemorrágica de la piel que se acompaña de coagulación intravascular diseminada. Describimos un caso de septicemia por *Streptococcus pneumoniae* en un niño de 5 años de edad tratado con tacrolimus oral, con una historia médica previa de trasplante de múltiples vísceras y sin bazo, y el desarrollo subsiguiente de púrpura fulminante en su pecho y la parte distal de sus extremidades. La forma aguda infecciosa de púrpura fulminante es debida habitualmente a bacterias gramnegativas. Los casos secundarios a bacterias grampositivas encapsuladas ocurren por lo general cuando los individuos están inmunosuprimidos o presentan asplenia funcional o anatómica. Nuestro paciente presentaba ambas condiciones, lo cual con seguridad aumentó su susceptibilidad, y respondió bien a la terapia antimicrobiana

☆ Please cite this article as: Konda S, et al. Purpura Fulminans Associated with *Streptococcus pneumonia* Septicemia in an Asplenic Pediatric Patient. Actas Dermosifiliogr. 2013;104:623-7.

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1578-2190/$ - see front matter

además de a la cobertura profiláctica en el contexto de su inmunosupresión. Revisamos la literatura buscando casos similares debidos a *Streptococcus pneumoniae* en la población pediátrica y discutimos la etiología y el tratamiento de la púrpura fulminante.

Case report

A 5-year-old Hispanic boy with a past medical history of premature birth at 24 weeks gestation and multivisceral organ transplantation (MVTx) at 2 years of age due to a hepatoblastoma with extensive abdominal compromise was admitted at the ICU with a fever of 103.4° F (39.7 °C). He complained of pain in his neck and right leg. He denied the onset of dysuria, cough, or ear pain. The MVTx included the stomach, pancreas, liver, and small and large bowels. He was on oral tacrolimus 1.0 mg twice daily to prevent rejection, which was held on admission, and ganciclovir prophylaxis to prevent cytomegalovirus infection secondary to immunosuppression. The transplant did not include a spleen and consequently he received a pneumococcal conjugate vaccine and was on penicillin prophylaxis due to increased risk of infection. He also had a right lower quadrant ileostomy with normal output and denied any gastrointestinal changes.

On initial examination he was in moderate distress, vasodilated, and hypotensive. After a sepsis workup, he was started with broad spectrum of empiric antibiotics including vancomycin, piperacillin, tazobactam, and ceftriaxone. Venous blood gases revealed a severe mixed acidosis and he was started on fluid resuscitation, pressor support, shock doses of hydrocortisone, and subsequently intubated. Lung edema worsened secondary to extracellular fluid leakage and worsening acute respiratory distress syndrome (ARDS) so transplant surgery placed bilateral pleural pigtail drains to improve ventilation. Hypocalcemia was addressed with continuous calcium infusion and renal failure secondary to sepsis was managed with continuous veno venous hemodiafiltration (CVVHDF).

The day after admission, ecchymoses and petechiae with consolidated irregular areas of blue-black hemorrhagic necrosis and a surrounding erythematous border were noted on his chest and distal extremities (Fig. 1). He had cyanotic nailbeds with delayed perfusion. We did not have baseline quantitative or functional coagulation assays and levels of protein C, protein S, and antithrombin III were not measured. The rash progressed to numerous tense bullae, which were drained without debridement of the epidermis, and topical mupirocin was applied with Telfa dressing. The drained fluid was sent for culture and had no microbial growth. The dermatology service was consulted and a skin biopsy from the right arm revealed intraepidermal vesicles with focal epidermal necrosis, dermal hemorrhage, and multiple thrombosed vessels in the superficial and reticular dermis without evidence of vasculitis (Figs. 2 and 3).

Blood cultures came back as predominantly gram positive cocci in chains consistent with *Streptococcus pneumoniae*. The antimicrobial regimen was adjusted to linezolid, vancomycin, meropenem, abelcet, and gancyclovir for active coverage of *S. pneumoniae* septicemia and prophylactic coverage of other flora. He remained afebrile with a resolving leukocytosis and negative blood cultures and the affected areas continued to resolve with scattered hemorrhagic vesicles and crusts. Heparin or clotting factors were not administered because the patient was improving clinically with the adjusted antibiotic regimen by the time the skin biopsy results were obtained. He was weaned to lower settings on the ventilator, his heart rate and blood pressure stabilized, and tacrolimus was restarted due to subtherapeutic levels. Both pleural pigtail drains were removed without complications and CVVHDF was discontinued. He continued to have intermittent hemodialysis and was eventually extubated without complications.

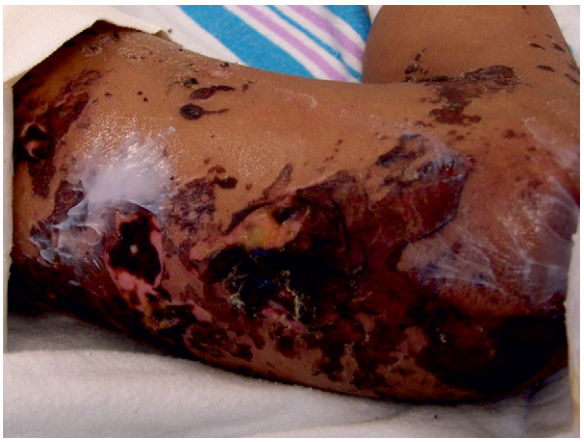

Figure 1 Widespread areas of purpura and necrosis on right lower extremity with lysed bullae and crusting.

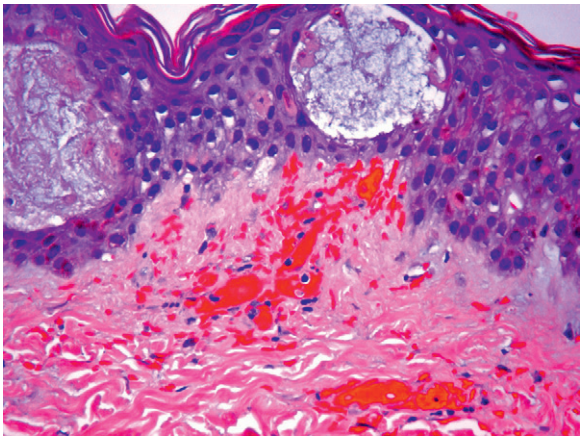

Figure 2 H&E 400×; focally necrotic keratinocytes, spongiotic vesicles, vascular thrombosis and red blood cell extravasation.