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CASE REPORT

Purpura fulminans: Functional results in two paediatric patients after suffering multiple amputations☆

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KEYWORDS
Purpura fulminans;
Meningococcal infection;
Amputation;
Paediatric patients;
Functional results

Abstract Purpura fulminans (PF) is an unusual haemorrhagic process that is usually associated with meningococcal sepsis and other infectious processes. It usually affects neonates and young children, and starts with a benign infection that progresses to a high fever, purpura ecchymosis, disseminated intravascular coagulopathy, necrosis and gangrene. The treatment of these children usually requires making difficult decisions, since the surgeon and the families must come to terms with the possibility of following an aggressive line of treatment that could lead to multiple mutilating sequelae, or follow palliative treatment. In this study, we review the clinical presentation, treatment and results of two cases of PF treated in our hospital between the years 2002 and 2005. The children presented in this study had a good long-term functional result and an acceptable quality of life, despite being subjected to multiple amputations.

PALABRAS CLAVE
Púrpura fulminante;
Infección meningocócica;
Amputación;
Pacientes pediátricos;
Resultado funcional

Púrpura fulminante: resultado funcional en 2 pacientes pediátricos tras sufrir múltiples amputaciones

Resumen La púrpura fulminante (PF) es un proceso hemorrágico inusual, que se asocia habitualmente a la sepsis meningocócica y a otros procesos infecciosos. Suele afectar a neonatos y a niños pequeños, y comienza como una infección benigna que progresa presentando fiebre alta, equimosis purpúrica, coagulopatía intravascular diseminada, necrosis y gangrena. El tratamiento de estos niños suele requerir la toma de decisiones difíciles, ya que el cirujano y los familiares deben plantearse la posibilidad de continuar con una línea de tratamiento agresiva que puede dejar múltiples secuelas mutilantes o seguir un tratamiento paliativo. En este estudio revisamos la presentación clínica, tratamiento y resultados funcionales de 2 casos de PF tratados en nuestro centro entre los años 2002 y 2005. Los niños presentados en este estudio tuvieron un buen resultado funcional a largo plazo y una calidad de vida aceptable a pesar de haber sido sometidos a múltiples amputaciones.

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Introduction

Purpura fulminans (PF) or gangrenous purpura is a rare haemorrhagic process commonly associated with meningococcal sepsis and other infectious processes.[1] It usually affects newborns and young children and begins as a benign infectious process which presents high fever, purpuric ecchymosis, hypotension, disseminated intravascular coagulation, necrosis and gangrene.[2]

Patients who suffer an episode of PF associated with severe peripheral ischaemia and gangrene of limbs or digits usually require multiple amputations. The treatment of these children often requires making difficult decisions, since the surgeon and family members must consider the possibility of continuing an aggressive treatment which could leave multiple crippling sequelae or following a palliative treatment.[3]

In this study we reviewed the clinical presentation, treatment and long-term functional results of 2 cases of PF treated at our institution between 2002 and 2005.

Case 1

The patient was a 3-year-old girl referred from another centre, who presented a symmetrical haematoma and peripheral erythema on both lower limbs of 4 weeks duration, associated with fever and asthenia. The patient had no history of interest, weighed 2.0 kg after a normal pregnancy and had successfully received appropriate immunisations for her age. Clinical examination highlighted the presence of a putrid odour and necrosis in both feet with a well-defined sock pattern (Fig. 1A). Femoral pulses remained intact. Blood cultures were obtained and empirical intravenous antibiotic therapy was started. Doppler ultrasound of both lower limbs showed a low resistance index in both popliteal arteries without a retrograde component, suggesting bilateral distal stenosis. Two days after admission she underwent transtibial amputation of both legs. Subsequently, the patient was admitted to the paediatric intensive care unit for 2 days. Initial blood cultures and serological markers were negative. Early rehabilitation was started and postoperative recovery was satisfactory. The patient began walking with prosthetics in both legs and was discharged at 4 weeks after surgery (Fig. 1B and C).

At 3 years from the first intervention, the patient was treated again for the reconstruction of amputation stumps. Three months after this second surgery, the patient developed an ulcer on the right stump which required a third operation for bone remodelling and wound closure. Rehabilitation began immediately after surgery. During subsequent reviews, the patient reported moderate dependence for usual activities, could walk and run freely and could get up from the ground without help. Subsequently, the patient was lost from our records since she was of foreign origin (Fig. 1).

Case 2

The patient was a 5-year-old girl who was admitted to our hospital due to fever, vomiting and drowsiness with 12 h duration. She had no history of interest and had been adequately vaccinated. Physical examination noted the presence of hypotension and disseminated purpura, predominantly in the distal left leg, right foot, left hand and right hand fingers.

The patient was admitted to the paediatric intensive care unit to start empirical intravenous antibiotic therapy and fluid therapy for expansion. CSF samples were normal and blood cultures were positive for *Neisseria meningitidis* (*N. meningitidis*) of undetermined serotype. On the third day after admission, the patient suffered acute renal failure, which improved after 6 days with haemofiltration. After this, she developed disseminated intravascular coagulopathy (DIC), presenting anaemia and thrombocytopenia and requiring treatment with blood derivatives. The ischaemic regions had an unfavourable evolution despite conservative treatment (Fig. 2A and B). At 20 days after admission and after the demarcation of necrotic areas, we performed amputation of the right foot, distal 1/3 of the left forearm, transtibial of the left leg and all the distal phalanges of the right hand. Within 10 days of the first operation we had to conduct a transtibial reamputation of the right leg due to a poor outcome of the surgical wound. At 6 weeks of the initial operation we performed reamputation of the second and fourth fingers. Rehabilitation was initiated early and the patient began walking with prosthetic lower limbs made to measure.

At 6 years of the review, the amputation stumps were in good condition and the patient reported a relatively good quality of life (Fig. 2C and D). She was able to put on the prostheses without help and could walk and run independently. The mobility of the shoulders, elbows, hips and knees was good, although she rejected the use of the left upper limb prosthesis. The patient showed moderate dependence for usual activities, both in the Barthel scale[4] (total score of 85%) and in the locomotor index scale (37/42).[5] According to the Russek classification, the patient developed a partial restoration of normal activities (with restrictions only for certain activities including dancing, sports, etc.) (Fig. 2).[6]

Discussion

PF was first described by Guelliot in 1884.[1] It is associated with infections caused by *N. meningitidis*, *Capnocytophaga canimorsus*, and other gram-negative pathogens, as well as deficiencies of protein C or protein S. In some cases it is not possible to identify the cause.[7]

N. meningitidis is a gram-negative diplococcus which only affects humans. It can be found regularly in the nasal mucosa, pharynx, intestinal tract and vagina. Acute meningococcal infections start with unspecific clinical symptoms and have a torpid evolution, causing fever, hypotension and coagulation disorders. In their most severe form they can cause septic shock, peripheral ischaemia, PF, necrosis and gangrene.[1] These processes are triggered by the release into the circulation of large amounts of lipopolysaccharide (LPS) endotoxin from the pathogen. This endotoxin activates macrophages, T cells and endothelial cells, which in turn cause the release of large amounts of prostaglandins and cytokines and lead to the activation of the coagulation cascade.[2]

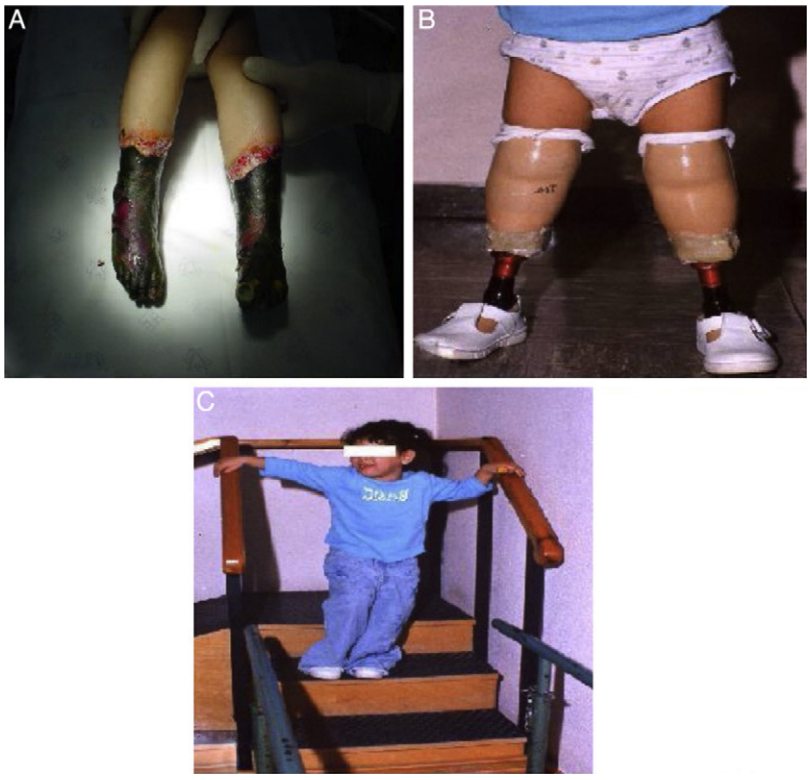

Figure 1 (A) Necrosis of both feet with a well-defined sock pattern. (B) The patient with walking prostheses in both feet. (C) The patient in rehabilitation at 2 weeks after surgery.

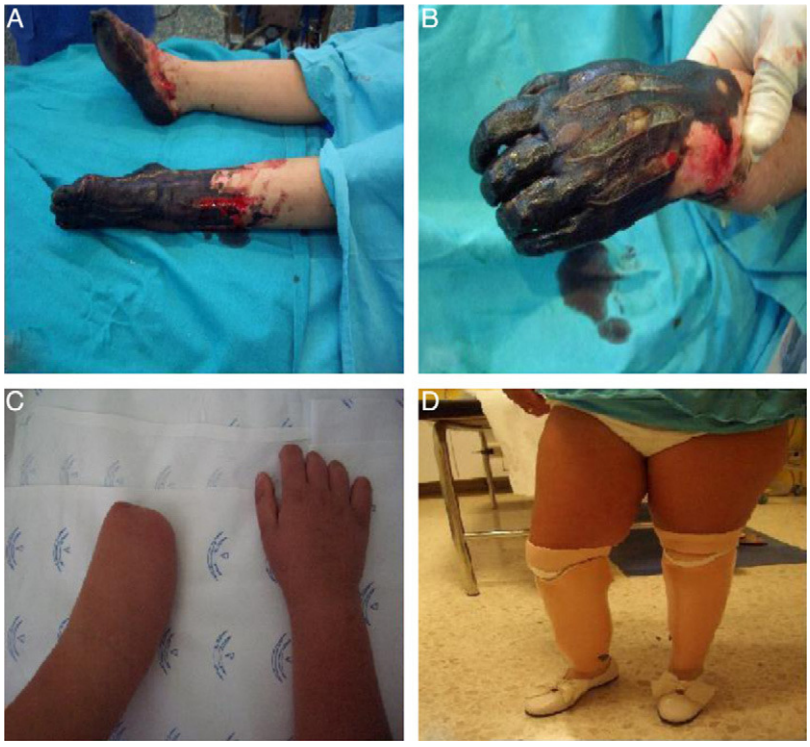

Figure 2 (A and B) Necrosis of both feet and left hand. (C) Amputation stumps of the left hand and right hand fingers at 3 years after surgery. (D) The patient with walking prostheses on both legs at 3 years after surgery.

Protein C deficiency associated with meningococcal infection contributes to the pathogenic process that triggers thrombonecrotic lesions of the skin and other organs and probably plays an important role in the systemic inflammatory response. Low levels of proteins C and S, as well as antithrombin III, have been observed in paediatric patients diagnosed with meningococcal infection who presented PF.[8] The meningococcal endotoxin has been described as a cause of intravascular coagulopathy (IVC).[7]

The conservative management of PF includes the use of antibiotics, means for volumetric expansion and oxygen. Good results have been reported with the use of antithrombin and protein C substitution therapies, correlating with improvement of biological markers. Surgical treatment includes debridement, scarectomies, fasciotomies and amputations.[7,9] A conservative management of the lesions is recommended during the acute phase of the disease, as long as no active local purulence is observed. Debridement or amputation should be performed when the viable tissue margins are well defined. Coverage of wounds with allografts may be necessary; the final coverage should be performed when local perfusion is recovered. It may be necessary to conduct subsequent surgical revisions to improve the quality of life of these children.[10]

During the postoperative period, patients may develop complications at the level of the surgical stump, such as skin necrosis, sensitivity alterations, contractures, surgical wound dehiscence, growth defects and delayed healing of the stump. Since the 2 cases presented were patients in growing age, our most important criterion in the planning of surgical treatment was to be extremely conservative when setting the amputation margins. This attitude increases the risk of immediate complications in the stumps. If this occurs, there is always the possibility of performing a second operation if necessary.

Affected patients require a long, multidisciplinary rehabilitation. There are few studies assessing the long-term quality of life of these patients. Knowledge of long-term functional results may facilitate therapeutic decision-making by the physician and family. In our study, both cases showed a good level of mobility, a moderate degree of dependence despite undergoing multiple amputations and an acceptable quality of life.

PF is a pathological process that can leave crippling sequelae. The children presented in this study had a good long-term functional outcome and acceptable quality of living despite having undergone multiple amputations.

Level of evidence

Level of evidence IV.

Ethical responsibilities

Protection of people and animals. The authors declare that this investigation did not require experiments on humans or animals.

Confidentiality of data. The authors declare that they have followed the protocols of their workplace on the publication of patient data and that all patients included in the study received sufficient information and gave their written informed consent to participate in the study.

Right to privacy and informed consent. The authors have obtained informed consent from patients and/or subjects referred to in the article. This document is held by the corresponding author.

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