British Journal of Plastic Surgery (2003), **56**

doi:10.1016/S0007-1226(03)00023-7

Idiopathic purpura fulminans and varicella gangrenosa of both hands, toes and integument in a child

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SUMMARY. Chicken pox is a common childhood illness and, though a vaccine is readily available, it is not routinely included in the vaccination schedule of most countries owing to its mild clinical nature. However, varicella gangrenosa is a rare complication of this disease, infrequently reported in the literature. We report the case of a child who developed purpura fulminans in the convalescent phase of chicken pox and subsequently presented with peripheral gangrene of both hands and the toes of the right foot, and skin gangrene. To our knowledge, bilateral gangrene of the hands has rarely been reported, and we present this case to highlight the serious nature of complications following varicella infection. © 2003 Published by Elsevier Science Ltd on behalf of The British Association of Plastic Surgeons

Keywords: varicella gangrenosa, purpura fulminans, protein S, disseminated intravascular coagulation, amputation.

'Varicella gangrenosa', a term first used by Hutchison in 1881 to describe the gangrenous skin lesions that may follow chicken pox infection, is an extremely rare complication, thought to occur in 0.05–0.16% of patients.[1,2] Though commonly described in children, and seen more frequently in the lower than the upper extremity, it can also affect the foetus following an in utero varicella infection.[2] Thomas et al described a case of periorbital varicella gangrenosa necessitating orbital exenteration in an unsensitised adult.[3] We report the case of a child who developed gangrene of both hands, the toes of the right foot and the integument, and discuss the management.

Case report

A 6-year-old male was transferred from a paediatric unit to the Al-Razi Orthopaedic Hospital. Ten days after the onset of chicken pox, he had developed an ecchymotic area on the leg, which spread rapidly to involve both upper and lower limbs and the buttocks. On admission to the paediatric unit, the following investigations were found to be abnormal: haemoglobin 7.2 g dl^{-1}; white blood corpuscles $14.4 \times 10^9\ l^{-1}$; platelets $732 \times 10^9\ l^{-1}$; alanine aminotransferase 1514 U l^{-1}; aspartate aminotransferase 2614 U ml^{-1}; creatine kinase 1191 U l^{-1} and antistreptolysin O titre 200 IU ml^{-1}. Subsequently, he deteriorated further and developed disseminated intravascular coagulation (DIC). While both protein C and protein S activity were reduced, lupus anticoagulant was not detected. The relevant haematological data is shown in Table 1. All the major vessels of both upper and lower limbs were patent on Doppler scanning. During this period the child received heparin (started at 20 U $kg^{-1}\ h^{-1}$ to maintain a patient time that is two to three times the control time of the activated partial thromboplastin time (APTT) test), immunoglobulin (1 g kg^{-1}), fresh frozen plasma (40 ml $kg^{-1}\ day^{-1}$) and oral prednisolone (2 mg kg^{-1} - day^{-1}), in addition to cloxacillin, cefotaxime sodium and acyclovir. During the following days, the purpura regressed, but the child was left with gangrene of both hands and four toes

Table 1 Haematological data (several days after admission)

Parameter	Normal value	Observed value
haemoglobin	11.5–14.5 g dl^{-1}	4.9 g dl^{-1}
white blood corpuscles	5.0–$12.0 \times 10^9\ l^{-1}$	$23 \times 10^9\ l^{-1}$
platelets	140–$440 \times 10^9\ l^{-1}$	$124 \times 10^9\ l^{-1}$
prothrombin time	10–14 s	20 s
activated partial thromboplastin time	25–39 s	44 s
fibrinogen	1.5–4.0 g l^{-1}	1.4 g l^{-1}
Fibrin degradation products	$<$500 ng ml^{-1}	2000 ng ml^{-1}
factor II (prothrombin)	60–160%	112%
factor VIII-C	50–150%	235%
protein C activity	70–140%	49%
protein S activity	60–140%	13%
antithrombin III activity	84–123%	48%

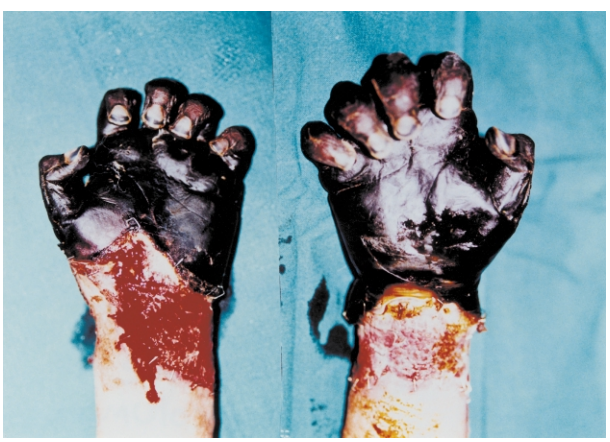

Figure 1—Varicella gangrenosa affecting the right and left hands in a 6-year-old boy.

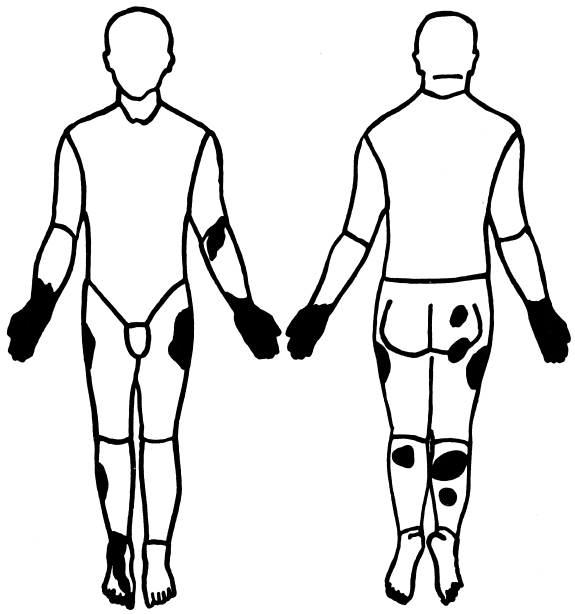

Figure 2—The areas affected by varicella gangrenosa.

of the right foot, and multiple cutaneous necrotic patches on both lower limbs, the right buttock and the left elbow (Figs 1 and 2). His routine investigations and coagulation profile returned to normal and, in addition to bilateral hand amputation, he underwent multiple debridements of the necrotic skin patches, which were covered by split-skin grafting.

Discussion

Although purpura fulminans was first described by Guelliot in 1884, the idiopathic type was first recognised as an entity only in 1964.[4] Idiopathic purpura fulminans usually follows the initiating febrile illness and manifests with rapidly progressive purpura, which may lead to skin necrosis, gangrene of limbs or digits and major organ dysfunction.[5] Deficiency of protein S is thought to be central to the pathogenesis of idiopathic purpura fulminans, and DIC is believed to be the major pathophysiological mechanism responsible for peripheral gangrene.[5,6] While the medical management has been exhaustively discussed elsewhere,[4,5] heparin, fresh frozen plasma, replacement of deficient blood components and supportive therapy forms the mainstay of treatment. Recombinant tissue plasminogen activator, immunoglobulins, glucocorticoids, prostacyclin, ketanserin, topical nitroglycerine, dextran, hyperbaric oxygen, regional anaesthesia and leeches have all been used with variable results.[4,5,7] In general, a conservative approach, excising gangrenous areas after they have demarcated from purpuric and indeterminate zones, will help to limit the tissues excised. However, in the presence of infection early aggressive surgical debridement may help to prevent invasive wound sepsis. When compartment syndrome is suspected in patients with tense limbs and distal ischaemia, early fasciotomy should be considered.[4] If there is established gangrene, conservative amputations may be unavoidable. In our patient multiple debridements were necessary before the wounds were fit for grafting, and the extremity gangrene ultimately led to bilateral hand amputation. The sequelae of varicella infection can be devastating, as in our patient, and, besides aggressively treating the antecedent purpura fulminans and DIC that are often associated with this complication, vaccination against varicella may be helpful in reducing the morbidity.

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Paper received 22 May 2002.
Accepted 30 December 2002.