Varicella gangrenosa

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SUMMARY A four year old girl presented with varicella gangrenosa, and haematological investigations showed a disseminated intravascular coagulation. The child subsequently developed a unilateral deep venous thrombosis. She was treated with oral steroids and intravenous heparin and made a full recovery.

Gangrene of skin and deeper tissues is an unusual complication of varicella. The term varicella gangrenosa has been applied to various conditions. In 1881, Hutchinson¹ coined the term varicella gangrenosa and referred to Stokes' earlier description in 1807 of gangrenous changes after chickenpox. In 1977, John described three distinct varieties: (a) moist, infective gangrene; (b) dry gangrene, secondary to arterial thrombosis; (c) purpura fulminans—that is, gangrene associated with a disseminated intravascular coagulation.² The child we studied presented with purpura fulminans and subsequently developed a unilateral deep venous thrombosis.

Case report

A 4 year old girl developed chickenpox and initially had a relatively mild clinical progress. On the seventh day of the illness her parents noted bruising of the child's legs that was distributed over her

Figure Child on the 10th day of her illness.

thighs and calves. The areas affected became swollen, painful, and exquisitely tender. The child was admitted to the children's unit on the 10th day.

On admission an examination showed a child whose weight and height were both on the 55th centile for her age. The patient was in no distress and had normal vital signs. Her skin showed evidence of healing chickenpox lesions. Both limbs were swollen, with large ecchymotic areas and surrounding erythema over the anterior and lateral aspects of thighs and calves (figure). The lesions were painful and extremely tender and covered 9% of her total body surface. Lower limb pulses were normal. Examination was otherwise normal. Initial haematology results showed a haemoglobin of 115 g/l, white cell count of $6.6 \times 10^9/l$, platelet count of $95 \times 10^9/l$, prothrombin ratio of 1.6 (normal 1.0) cephalin-kaolin time of 48 seconds (normal range 35–45 seconds) and fibrinogen degradation products were raised at 40 mg/l (normal range <10 mg/l). All the blood cultures during the patient's stay in hospital were negative.

Treatment was started with intravenous phenoxymethylpenicillin, as prophylaxis against infection, and oral steroids. Over the next few days the ecchymotic areas became gangrenous with subsequent formation of bullae. On day 17 she complained of a painful right calf. Clinical impression of a deep venous thrombosis was confirmed on venography. Full anticoagulation treatment was started with intravenous heparin for 10 days. A further 10 weeks of anticoagulation treatment was completed with oral warfarin. Steroid doses were decreased and stopped on day 24; the phenoxymethylpenicillin was also stopped. During her subsequent hospital stay a thick eschar formed over the lesions. On separation there was evidence of re-epithelialisation at the edges with central scar tissue formation.

Discussion

Chickenpox, caused by *Herpesvirus varicella*, is in general a benign condition. Many complications, however, may occur: sepsis resulting in septicaemia; osteomyelitis; pneumonia; postinfectious encephalitis, and occasionally myelitis and polyradiculitis. Other complications are pneumonitis with nodular infiltration seen on x ray film of the chest, myocardi-

tis and appendicitis; and haemorrhagic chickenpox where bleeding occurs into the vesicles. Varicella gangrenosa is a rare complication: Barenberg and Lewis reported one case in 2000 cases of varicella seen in their hospital over 14 years. Wishik and Bullowa saw four cases in 2534 patients with varicella.

Our patient presented with the purpura fulminans variety of varicella gangrenosa. This variety consists of gangrenous skin lesions with a disseminated intravascular coagulation. Purpura fulminans was first described by Guelliot in 1884 and Henock in 1887. Hjort's criteria³ for the diagnosis of purpura fulminans include: (1) It is a disease of children. (2) It is preceded by a benign disorder—usually an infection. (3) After a variable time lag bleeding into the skin occurs and spreads rapidly. (4) These lesions become gangrenous with formation of bullae. (5) Coagulation abnormalities compatible with disseminated intravascular coagulation may be found. (6) Thrombosis of venules and capillaries with a prevascular inflammatory infiltrate is found on investigation of the histology. This process is thought to represent a human equivalent of the generalised Shwartzman reaction of laboratory animals. There is no apparent relation between the gangrene and the severity of the varicella eruption. Indeed, as in our case, the mildness of the eruption has been noted.⁴ In Spicer's literature review of 100 cases, varicella was the third most common preceding illness, 19% had an entire extremity amputated, and the overall mortality was 35%.⁵ Our patient, who was affected extensively in both lower limbs, made a full recovery with minimal scar tissue formation.

Because of the rarity of this condition no definitive treatment can be recommended. Treatment in the acute phase is largely supportive. Heparin treatment has been used with some good results. The dose and duration of treatment vary widely, but most reports suggest a high dose over at least three weeks. Controlling the coagulopathy does not affect the ecchymosis or the incidence of gangrene and there are many reports of unfavourable experiences.⁶ Steroids have been used in many cases with varied results. Our patient responded well to steroids and heparin without resort to skin grafting or amputation.

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