

Transient neonatal Behcet's disease

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SUMMARY A term neonate with a transient form of Behcet's disease is described. The clinical and immunological features are detailed. As healing of the severe ulceration in this condition gives rise to scarring and cosmetic deformity recognition is important. Therapeutic intervention with corticosteroids is recommended after diagnosis.

Behcet's disease is rare in the United Kingdom, though it is common, up to one in 5000 of the population, in Mediterranean countries and Japan.¹ The aetiology is unknown, but the case for it being an immune complex mediated disorder is strong.² To date there have been two reports of a transient neonatal form of Behcet's disease in infants of affected mothers.^{3 4} We describe a third case and the immunological abnormalities associated with it.

Case report

A 2800 g girl was delivered at term to a 33 year old mother with one normal 7 year old child from the same marriage. She had an eight year history of Behcet's disease, characterised by severe recurrent oral ulceration, colitis, arthritis, conjunctivitis, pustulonecrotic skin lesions, thrombophlebitis, and a positive pathergy reaction. The disease was kept in remission with oral prednisolone 7.5 mg daily, the dose being doubled with any major flare up of

disease activity. She remained on prednisolone throughout the pregnancy.

At birth the infant was well but had a few small, periungual, pustulonecrotic lesions. At the age of 8 days she developed multiple mouth ulcers, which steadily progressed to severe destructive ulceration of the lips, buccal mucosa, and tongue, together with pustulonecrotic skin lesions, mainly on the hands and feet, and periungual ulceration. She had a positive pathergy reaction at the sites of venepunc-



Fig. 1 Pustulonecrotic, periungual, and skin lesions on the infant's right hand at the age of 12 days.

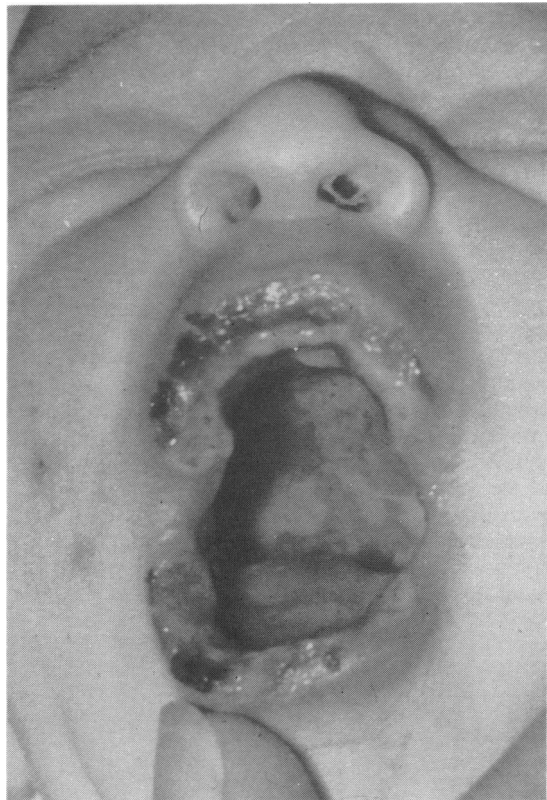


Fig. 2 Mouth and face at the age of 19 days, showing the changing pattern of ulceration on the palate and tongue, increasing severity of ulceration of the lips, but healing of the lesion on the right cheek. A further pustular lesion is developing on the right cheek, below and medial to the original lesion.

ture. The lesions were identical to those her mother suffered in relapse (Fig. 1).

Investigation showed her to have a neutrophil leucocytosis of $19 \times 10^9/l$ in a total white cell count of $29 \times 10^9/l$. Bacterial and viral infection screens yielded negative results. Electron microscopy of pustule fluid produced negative results for virus particles, and paired viral titres showed no rise to any of the common viral pathogens, including herpes simplex. IgG and IgM concentrations were raised at 10.5 and 1.3 g/l, respectively. C3, C4, and Clq were normal, but IgG immune complexes were present and the total haemolytic complement was markedly reduced at 8%. Though non-specific, in the absence of evidence of infection, these findings suggest a disease secondary to an immunological disorder.

The infant was initially treated with broad range antibiotics and intravenous acyclovir, neither of which seemed to have any effect on the clinical progress of her disease. New lesions continued to occur during the first month of her life (Fig. 2), but none occurred after the age of 5 weeks and all healed by 8 weeks. Extensive depressed scarring of the lips and small depressed scars elsewhere remain.

Discussion

This is the third reported case of neonatal Behcet's disease and the first in paediatric publications. It is also the first case in which immunological abnormalities are described. Fam *et al* described their case 13 years in retrospect from the presence of small pitted scars and a suggestive history.³ Thivolet *et al* described the clinical features of an affected neonate.⁴ From these three reports several important points emerge. Neonatal Behcet's disease requires recognition as a transplacentally acquired disease in infants, like transient neonatal lupus erythematosus⁵ and other immunological diseases. It is probably caused by the transplacental passage of immune complexes or autoantibodies. It causes severe destructive mucosal and skin lesions, which

heal with scarring. It resolves spontaneously after the first six to eight weeks of life.

The scarring resulting from the destructive mucosal and skin lesions causes serious cosmetic deformity. Early diagnosis of this condition is desirable, both to prevent unduly prolonged administration of unnecessary treatment and also to allow appropriate treatment to be given in the early stages to limit ulceration. The least toxic, effective treatment in Behcet's disease is prednisolone. Though thalidomide is reported as being effective for the mucocutaneous lesions in Behcet's disease⁶ it has not been used in neonates and is associated with side effects, such as peripheral neuropathy. Cytotoxic drugs and cyclosporin A are clearly inappropriate. Early use of prednisolone may prevent, or limit the extent of, subsequent scarring and therefore ought to be employed once disseminated herpetic infection is excluded by the developing clinical picture and electron microscopic findings.

We thank Dr K Bardan for his help with the maternal history and for the clinical photograph of the mother and Dr A M Ward for his help with the immunological assays and interpretation.

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Received 24 March 1986