

SHORT COMMUNICATION

Recurrent Finkelstein-Seidlmayer Disease in Four First-degree Relatives

Alessandro Ostini¹, Gian Paolo Ramelli¹, Carlo Mainetti², Mario G. Bianchetti^{1*} and Alessandra Ferrarini¹¹Department of Pediatrics, Ospedale San Giovanni, CH-6500 Bellinzona, and ²Department of Dermatology, Regional Hospital of Bellinzona, Bellinzona, Switzerland. *E-mail: mario.bianchetti@pediatrician.ch

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Acute cockade purpura and oedema of young children, also known as Finkelstein-Seidlmayer disease (FSD), is a rare and benign cutaneous small-vessel leukocytoclastic vasculitis syndrome. This condition, which mostly affects the extremities, face and ears, presents with non-pruriginous target-like lesions, often associated with tender, non-pitting oedema (1). It affects children aged 2–60 months in an approximate male-to-female ratio of 2:1, typically does not recur, and often follows a simple febrile illness or an active immunization (1). There are no reports of familial occurrence of FSD.

We report here the medical history of a 43-year-old woman and her 3 sons, currently aged 14, 12 and 9 years, who have been affected since birth by frequent and persisting relapses of FSD.

METHODS AND CASE REPORTS

The data and photographs for this report study were collected from the medical charts of the Departments of Pediatrics, University of Basel, Bern and Zurich, the Departments of Pediatrics and Dermatology of Southern Switzerland, and the family doctors. An interview with the patients was carried out (Table 1).

Mother: A 43-year-old Swiss woman had been treated by one of the authors for recurrent episodes of non-thrombocytopenic skin lesions occurring over her extremities, ears and face. Lesions included large, red-to-purpuric, non-pruriginous and tender plaques. History-taking revealed that some lesions probably developed at birth. The condition relapsed approximately every 2 months in early childhood and adolescence, always preceded by a simple airway illness or a diarrheal disease. Once she

reached adulthood, the recurrence rate decreased substantially to approximately once a year. The disorder also recurred during her 3 otherwise uneventful pregnancies. The exanthemata were not associated with arthralgia, abdominal pain, macroscopic stool bleeding or urine discoloration. Since stool guaiac testing for occult blood was slightly positive on no more than one occasion, no gastrointestinal endoscopy studies were performed.

During a recent interview, the patient reported that her grandmother had probably presented a similar recurrent skin disease in childhood. However, there was no family history of recurrent severe infections or bleeding disorders.

Son 1. The woman delivered a boy vaginally at term in the cephalic position, with a body weight of 2.73 kg and with no perinatal or neonatal disease indicators. Physical examination at birth disclosed purpuric swollen patches over his hands, a purpuric patch en cockade with a diameter of approximately 1 cm over the left ankle, and some facial petechiae. His stool was bright red and guaiac-positive. Although no test was performed to differentiate maternal from foetal blood, the diagnosis of maternal blood ingestion, the commonest cause of neonatal bloody stool, was made. Two days after birth, poor feeding and poor peripheral perfusion were reported. Blood cultures were found to be positive for *Escherichia coli* and coagulase-negative staphylococci. A suspected diagnosis of neonatal sepsis was made and parenteral antimicrobials were administered. The skin lesions progressively improved within 5 days and no further intestinal bleeding was observed. At the age of 2 months, non-pruriginous, tender, target-like plaques appeared over the right cheek and the right leg in the infant, who was doing well 3 days after the first vaccination. The lesions resolved within 3 days. Furthermore, often febrile (rectal temperature $\geq 39.5^{\circ}\text{C}$) recurrences developed at the age of 4, 6, 13, 14 and 19 months, mostly preceded by an airway or diarrheal disease. Recurrences were also noted at the age of 4 and 13 years (Fig. 1).



Fig. 1. (A) Haemorrhagic lesions on the extremities of son 1 at the age of 3 months. (B) Target-like lesion on the leg of son 1 at 15 months of age. (C) Hemorrhagic plaques at birth on the palmar side of the hands of son 1. (D) Annular oedematous thigh plaques in the mother at 34 years of age.

Son 2. The woman's second child, a boy, was born vaginally in the 35th gestational week, with a body weight of 2.76 kg and without perinatal or neonatal disease indicators. Oedematous painful skin lesions with central petechiae expanding centrifugally to assume a polycyclic configuration on both hands, feet, on the back and on the penis were noted at birth. The child was non-toxic in appearance and the skin lesions resolved spontaneously 4 days later. A first relapse was observed at the age of 10 months after an upper respiratory illness. The skin lesions persisted for approximately 3 weeks, probably because the airway disease was followed by a diarrheal one. Two relapses were noted at the age of 2 and 12 years, characterized by extremely painful plaques.

Son 3. The third son was born, like his brothers, without perinatal or neonatal disease indicators. Physical examination at birth disclosed painful oedematous target-like lesions on his hands, feet, cheeks, ears and extremities. The rash resolved spontaneously a few days later. A first relapse occurred at the age of 4 months, a few days after his second vaccination. By the age of 8 years, approximately 6 further relapses had occurred.

Laboratory investigations. Laboratory testing failed to disclose thrombocytopenia, coagulation disorders (by prothrombin time and partial thromboplastin time), immunoglobulin (Ig)M rheumatoid factor antibodies, anti-neutrophil or anti-nuclear autoantibodies and pathologically reduced C3 and C4 complement levels (this investigation was not performed in son 3). Furthermore, urinalysis did not reveal proteinuria or haematuria, either during or between attacks. A skin biopsy, performed exclusively in son 1, revealed arterioles infiltrated with neutrophils whose nuclei were partly fragmented and pyknotic (no immunofluorescence study was performed).

DISCUSSION

We describe here the familial occurrence of a condition with an apparent dominant or, less likely, X-linked inheritance. The condition is characterized by non-pruriginous target-like lesions, sometimes associated with tender, non-pitting oedema; absent or not significant visceral involvement; recent history of a simple febrile illness or active immunization; onset at birth, tendency to recur (following a simple infection or, less frequently, an active immunization) during infancy, early childhood, school age, adulthood and pregnancy; normal screening test for autoimmunity; and a skin biopsy showing leukocytoclastic vasculitis. The condition satisfies the diagnostic criteria recommended for FSD (1). The patients described in this report are exceptional because familiarity, onset at birth, recurrence and relapses persisting into adolescence, adulthood and pregnancy have not been reported previously for this annular vasculitis (1).

Table I. *Clinical characteristics of Finkelstein-Seidlmayer disease in the 4 patients*

Characteristics	Mother	Son 1	Son 2	Son 3
Current age, years	43	14	12	9
Age at onset	Birth(?)	Birth	Birth	Birth
Frequency of relapses, /year				
≤24 months of age	Frequent	≈5	≈1	≈2
>2 years of age	≈6 ^a	≤1	≤1	≤1

^aSince adulthood ≈1/year (including her 3 pregnancies).

The diagnosis of FSD has also been reported in a 21-year-old woman, with a purple rash and swelling affecting the arms, face, and ear, which spontaneously resolved and did not recur (2). Furthermore, recurrent FSD has been observed in an infant with Wiskott–Aldrich syndrome, an X-linked immunodeficiency characterized by thrombocytopenia, eczema, recurrent infections and increased risk of autoimmune disorders and malignancies (3). Drug-induced cutaneous vasculitides presenting with annular lesions resembling FSD have also been observed occasionally (4).

In the past FSD has often been considered a variant of Henoch–Schönlein purpura (5). This hypothesis is supported by a report documenting the concurrent appearance of FSD and Henoch–Schönlein purpura in a female infant and her brother (6). Cases also exist of children who have findings overlapping between the 2 vasculitides (7). On the other hand, there are enough clinical and prognostic differences to consider FSD a distinct entity, including the different skin lesions, and especially the failure to detect depositions of immunoglobulin A in most biopsy specimens (1, 8). Finally, thus far, family clusters have been exclusively reported for Henoch–Schönlein purpura (9). As a consequence, it is currently assumed that FSD and typical Henoch–Schönlein purpura are similar, but different, vasculitides (10).

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