

Chronic Recurrent Annular Neutrophilic Dermatitis. An Entity?

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Two cases with recurrent annular lesions of similar clinical appearance and course are described. The histopathological investigations showed in both cases dense and diffuse infiltrates mainly of polymorphonuclear neutrophilic leucocytes in the mid dermis. No fever, leucocytosis or elevated ESR were observed. Systemic corticosteroid treatment controlled symptoms but recurrence after treatment was withdrawn occurred in both cases. It is discussed if we are dealing with a variant of Sweet's syndrome or a new entity.

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In 1964, Sweet described 8 cases of acute febrile neutrophilic dermatitis (1). Since then, several cases have been reported from different parts of the world (2). Major and minor criteria are proposed for Sweet's syndrome (3). When reviewing the literature we have noticed a few cases (4-7), histopathologically identified as Sweet's syndrome but clinically characterized by chronic recurrent annular solitary lesions with sharply raised borders localized to the face, usually without malaise and fever. We here describe 2 cases with more widespread, but clinically and histopathologically rather similar lesions and question if we are dealing with a special entity.

CASE REPORT

Case 1

A 51-year-old female, previously healthy and without any known rashes. The patient was admitted to the Department of Dermatology in Kristianstad in August 1983 with a 6-week history of two centrifugally progressing lesions in the scalp and forehead regions. She presented two tender plaques with a dusky red center and well defined raised 2-3 mm wide sharply demarcated borders. The size of the two lesions were 5×8 and 2×2 cm, respectively. No history of preceding fever, infection or trauma was obtained. Laboratory investigations including ESR, hemoglobin, WBC counts with differential counts, liver enzymes, serum protein electrophoresis, syphilis serology and urine analysis for albumin, glucose and red cells were all normal. Histopathological investigation showed normal epidermis, subepidermal oedema and in the mid corium a dense and diffuse infiltrate consisting of mainly

polymorphonuclear neutrophilic leucocytes and some lymphocytes and eosinophils. Leukocytoclasia, fibrinoid necrosis of the capillaries or extravasation of erythrocytes were not observed (Figs. 1 *a* and *b*). A course of tetracyclines followed by another course of V-penicillin was without effect. Three months after start of the rash Dapsone 100 mg daily was administered for two months with doubtful effect and finally withdrawn due to hemolytic anemia and dizziness. Approximately one month later the patient went into complete remission lasting for 3½ years, when she returned with 6 new 1×1 cm to 5×7 cm tender lesions located to the forehead, trunk and upper extremities (Figs. 2 *a* and *b*). Routine blood and urine analysis, C₃, C₄, C₂, C_{1q}, C_{1s} complement factors, ANA and Borrelia serology were all normal. A light microscopy examination resulted in the same findings as mentioned above. A direct immunofluorescence investigation showed deposits of fibrinogen in the vessels and C₃ granules at the basement membrane zone, but no IgG, IgM or IgA antibodies. A new course of Dapsone 100 mg daily for 6 weeks and then reduced to 50 mg daily, due to side effects, for 4 weeks showed no clinical effect. Then systemic corticosteroid prednisolone 30 mg daily was administered for a week resulting in disappearance of tenderness and infiltration, leaving dusky red macular patches. During the following month the dose of prednisolone was reduced to 10 mg daily before withdrawal. This dose controlled all symptoms besides the remaining macular discoloration. Within a week after discontinuation of oral prednisolone new typical lesions occurred in new places without reactivation of former lesions. Again oral prednisolone was given in the same dose as before, this time resulting in complete clearance within 2 weeks, but still leaving discoloration in the old lesions. A second time, approximately a week after cessation of prednisolone treatment, a new 2×2 cm lesion recurred in the scalp. This was left untreated and within a month a spontaneous remission was noticed, except for the discoloration. The following three months the patient remained in remission, but discoloration was still visible in a 9-month old lesion on the right upper arm. Until now the patient has had two recent bouts responding to prednisolone in moderate doses during a 2-3 week period.

Case 2

A 38-year-old female, who has lived in an institution for mentally retarded since the age of 8. She has an IQ of 30 without any hereditary or organic known causes, otherwise a normal somatic health. The patient was admitted to the Department of Dermatology in Malmö at the end of April 1987 with a 4-week history of a centrifugally growing lesion on the right side of the neck. Before admission she was treated with

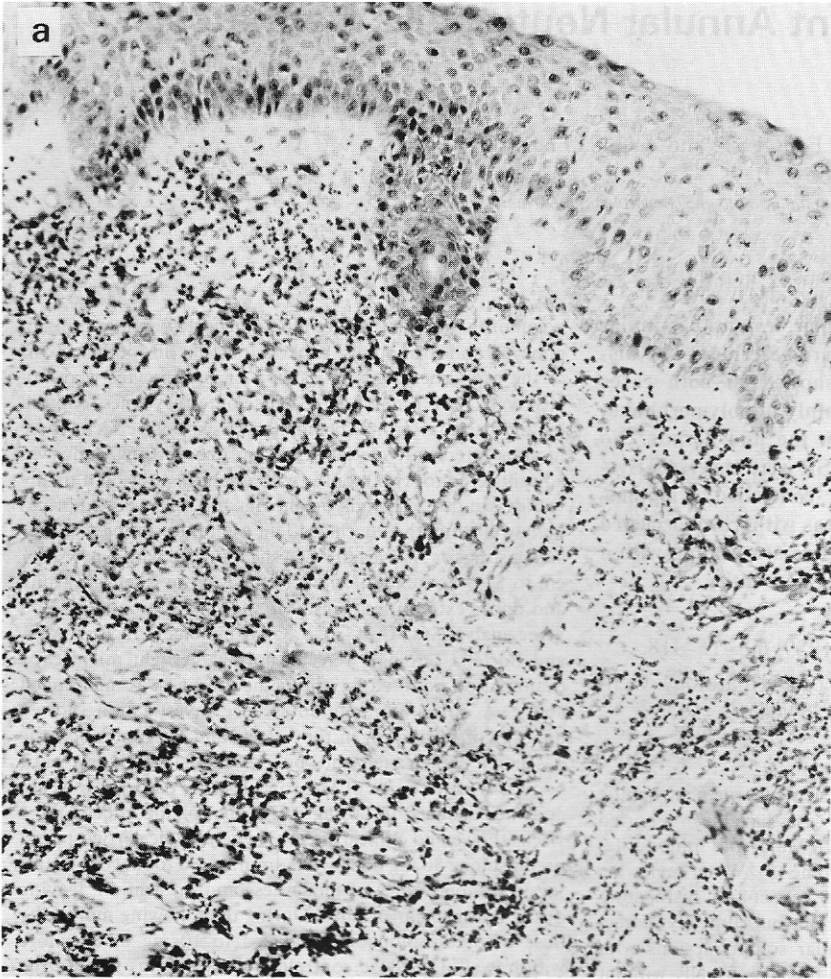


Fig. 1 a. Case 1. Subepidermal oedema and in the mid corium a diffuse infiltrate of mainly neutrophilic leukocytes ($\times 150$).

an econazole cream without any effect. At examination, she presented with an 8×6 cm dusky red annular lesion with a 2–3 mm wide raised sharply marked border. Due to the patient's mental state no history regarding tenderness was obtainable. No fever or deterioration of her general health was noticed. Laboratory investigations including ESR, hemoglobin, WBC counts with differential counts and liver enzymes were normal. Histopathological examination showed the same changes as in case 1. During the following 2 weeks 4 new annular lesions, 1–3 mm in size, appeared on the left cheek and neck, whereas the infiltrated annular border of the primary lesion on the right side of the neck had subsided, but still showed some infiltration and discolouration. Oral prednisolone 30–20 mg daily during 6 days resulted in disappearance of the infiltration but leaving discolouration in a 7-week

period. Then a new bout appeared which again was brought into remission during a 2½ month prednisolone medication starting with 30 mg daily down to 5 mg daily, when it was withdrawn. Three weeks later a new bout appeared, involving a 5×4 cm lesion on the right lower leg. A new prednisolone course starting with 30 mg daily was commenced. This time the dose was slowly reduced to 10 mg daily during a 6-week period, when the patient developed a new red nodular, 5 mm in size, lesion on the neck, with histopathological findings as described above. At this time the prednisolone dose was raised to 15 mg daily and then slowly reduced to 5 mg every second day during a 4-month period. No relapses were observed during that time but discolouration in former lesions was seen during the prednisolone treatments and disappeared 2½ months after the treatment was withdrawn.

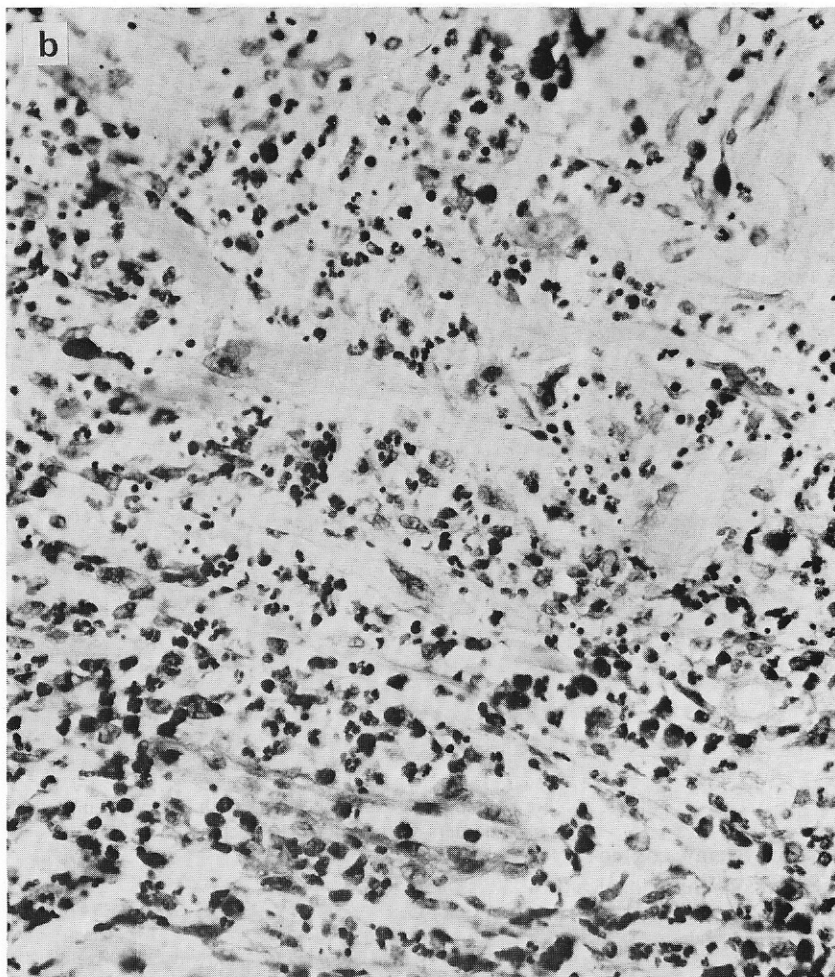


Fig. 1 b. Case 1. Higher magnification showing the diffuse infiltrate of neutrophilic leukocytes ($\times 370$).

DISCUSSION

We describe two cases with similar clinical lesions and course of the disease. Also histopathological examinations, laboratory investigations and response to systemic corticosteroid treatment were the same in the two cases. We suggest that the annular centrifugally spreading lesion with a sharply defined raised margin and the tendency to leave a dusky red discolouration should serve as typical recognizable features of the present condition. Furthermore, the chronic course without any sign of systemic illness separates the present cases from most cases described as Sweet's syndrome.

Whittle et al. (4) have reported two cases, which might be the same condition as here reported. However, we never observed herpetiform vesicles, pustulation or leucocytosis and the lesions were not solitary in our cases. Also Sweet himself, in 1968 (5) and later on in 1979 (6), has reported two cases, in which there were the annular abrupt borders. However, in one of the cases (6) severe malaise and fever were present. Borrie (7) reported a chronic, clinically similar solitary lesion in a woman without leucocytosis and fever.

Systemic corticosteroids were effective in our cases but recurrence after the treatment was withdrawn

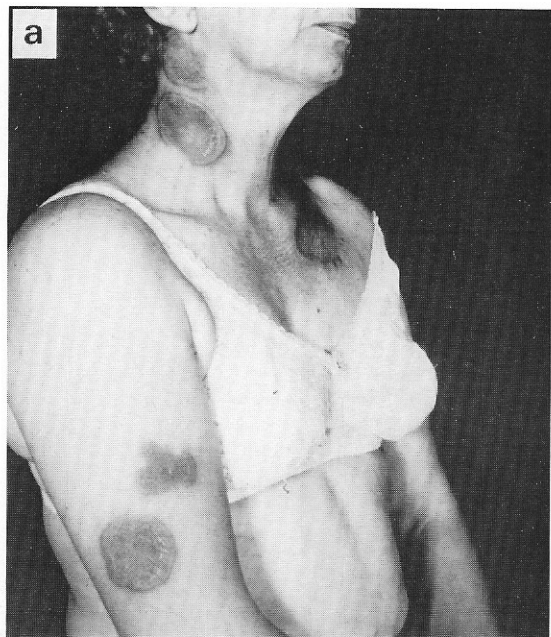


Fig. 2 a. Case 1. Annular lesions on neck and arms.



Fig. 2 b. Case 1. Annular lesions with sharply demarcated raised borders on the side of the neck.

occurred in both cases. Response to systemic corticosteroids is reported in one of the cases of Whittle et al. (4), the two cases of Sweet (5, 6) and in the case of Borrie (7), but relapses, as in our cases, are only reported by Sweet (6). In one of our cases, Dapsone was tried without effect. Spontaneous remission for 3 1/2 years was observed in one of our cases and is also reported by Whittle et al. (4). However, in cases with annular lesions with histopathologically dense infiltrates of neutrophils in the dermis moderate doses of systemic corticosteroid should be administered to relieve tenderness and the often cosmetically disfiguring lesion.

We suggest, due to characteristic annular lesions with abrupt borders, chronic course, histopathological findings and absence of abnormal blood tests, that the two cases reported here represent an entity. These are probably previously reported as Sweet's syndrome or as a variant of this.

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