

# Chronic recurrent annular neutrophilic dermatosis revealing sarcoidosis



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## INTRODUCTION

Neutrophilic dermatoses are a clinically heterogeneous set of diseases characterized by the presence of a histologic neutrophilic infiltration. They are frequently associated with systemic diseases such as malignant hematologic diseases, inflammatory bowel diseases, and rheumatologic illnesses.<sup>1,2</sup> Chronic recurrent annular neutrophilic dermatosis (CRAND) is an exceptional form of neutrophilic dermatosis characterized by a typical clinical presentation and evolution, composed of annular and recurrent lesions, and characterized by the absence of deterioration of general health and hematologic abnormalities. We report a case of CRAND associated with sarcoidosis.

## CASE REPORT

A 42-year-old woman with no significant medical history presented with an annular plaque on the left forearm, with a centrifugal extension, which had been evolving since 1 month earlier. The physical examination revealed an erythematous lesion, bordered by a papular ring with central healing associated with a fine scale (Fig 1). The lesion was sensitive to palpation and mildly pruritic. The rest of the clinical examination result was normal.

The patient described 2 previous episodes with similar lesions located on the extensor surface of the forearms, the first episode having occurred 2 years earlier. The lesions resolved spontaneously within a 4- to 6-week period. The outbreaks were not associated with fever, nor were they preceded by drug intake.

### Abbreviation used:

CRAND: chronic recurrent annular neutrophilic dermatosis

Histopathologic examination showed papillary dermis edema. The superficial and medium dermis contained a dense inflammatory infiltration rich in neutrophils. The appearance was suggestive of Sweet syndrome. There was no evidence of vasculitis (Fig 2).

Blood testing did not reveal any leukocytosis nor an increase in C-reactive protein. Lyme disease serology result was negative. There were no clinical or paraclinical arguments for an underlying malignant hematologic malignancy or inflammatory rheumatism: complete blood cell count, rheumatoid factor, and the anticyclic citrullinated peptide level were normal. The upper and lower digestive endoscopic evaluation results were normal, as was the result of the gynecologic examination. Evaluation of HIV serology and antinuclear antibodies yielded negative results, and the thyroid function test result was normal. The full-body computed tomographic scan showed mediastinal and hilar lymphadenopathy, which were intensely hypermetabolic on the positron-emission tomographic scan, associated with 5 nonhyperbinding micronodules in the right lung. Histopathologic examination of a lymphadenopathy of the Baretz space showed nonnecrotizing epithelioid granulomatous inflammation with rare giant cells. The Ziehl-Neelsen stain result was negative. The histologic aspect was compatible

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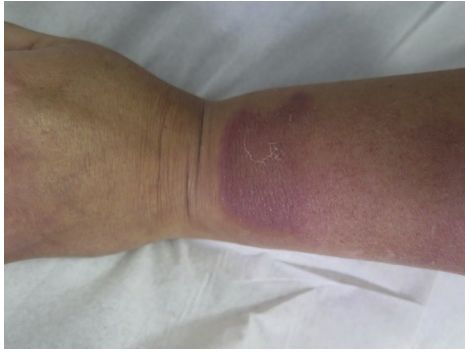
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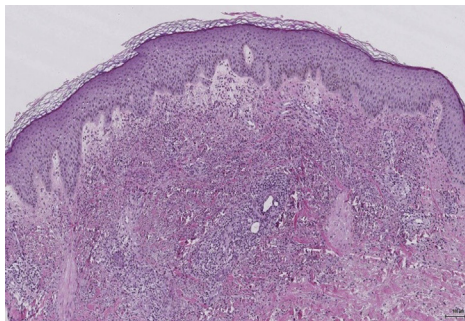
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**Fig 1.** Chronic recurrent annular neutrophilic dermatosis. Annular erythematous plaque of the left forearm, associated with fine scales.



**Fig 2.** Chronic recurrent annular neutrophilic dermatosis. Papillary dermis edema associated with a dense inflammatory infiltration rich in neutrophils in the superficial and medium dermis, without vasculitis, compatible with the diagnosis of Sweet syndrome. (Hematoxylin-eosin stain; original magnification:  $\times 100$ .)

with sarcoidosis. A diagnosis of CRAND revealing sarcoidosis was made.

The lesion of the forearm regressed rapidly within a few days with topical corticosteroids. No flare-ups occurred within a 3-year follow-up period.

In regard to the diagnosis of sarcoidosis, the angiotensin-converting enzyme level was within range, and the clinical and paraclinical examination did not show any other affected organ systems. Simple monitoring was proposed.

## DISCUSSION

CRAND is an exceptional form of neutrophilic dermatosis. Christensen et al<sup>3</sup> described this entity for the first time in 1989, and fewer than 10 cases have been reported (Table I). The clinical presentation is stereotypical and includes the occurrence of erythematous annular lesions with a centrifugal evolution and a central healing aspect associated with a fine scale and a papular border. It is a chronic disease with recurrent flare-ups during a few months to several years, with spontaneous regression of

lesions in most cases. The lesions are mostly located on the upper and lower limbs and on the face and neck.

Histologically, CRAND is similar to Sweet syndrome.<sup>4</sup> However, it occurs in the absence of general symptoms (fever in particular) and without any blood abnormalities (such as leukocytosis and an increase in C-reactive protein level). Vignon-Pennamen et al<sup>5</sup> published a series of 9 patients with chronic recurrent lymphocytic Sweet syndrome associated with myelodysplastic syndromes. The authors explained that lymphocytic skin infiltrate with a clinical aspect of Sweet syndrome should lead to the research of atypical myeloid cells in skin infiltrate, blood, and bone marrow. In our patient, there were no atypical cell infiltrates on the skin biopsy and the complete blood cell count was normal. We did not perform a bone marrow aspiration.

CRAND is usually described as a neutrophilic dermatosis occurring without any underlying diseases<sup>3,4,6</sup>; however, Blaizot and Doutre<sup>7</sup> reported a case of CRAND associated with a monoclonal gammopathy IgG  $\kappa$  type, and Mir-Bonafé et al<sup>8</sup> described a case associated with rheumatoid arthritis.

In our patient, the clinical and histologic presentation was typical of CRAND, and the evaluations revealed sarcoidosis 2 years after the onset of the first lesion. There was no granuloma on the histologic examination of the cutaneous lesion, excluding a specific cutaneous localization of sarcoidosis or another granulomatosis.

Indeed, although palisaded neutrophilic and granulomatous dermatitis has already been described in association with sarcoidosis,<sup>9</sup> we excluded this diagnosis because of the absence of granulomas, degenerated collagen, or dermal fibrosis on the histologic examination of this lesion that had been evolving for 1 month.

We believe the occurrence of these 2 rare diseases was not coincidental, suggesting an underlying link; also, the association between neutrophilic dermatoses and sarcoidosis, although rare, has already been described.<sup>10</sup> Moreover, it is not uncommon for a systemic disease to be discovered a few years after the onset of a neutrophilic dermatosis. To date, no guidelines exist for the treatment of CRAND. In most cases, topical or general short-term corticotherapy has allowed rapid regression of the lesions without preventing their relapse.<sup>4</sup> In some very recurrent forms, dapsone and colchicine have been used effectively in the prevention of recurrences<sup>3,5-7</sup>; hydroxychloroquine seems ineffective in preventing them.<sup>4</sup>

**Table I.** Clinical, hematologic, and histopathologic features of patients with chronic recurrent annular neutrophilic dermatosis

Age, years, sex	Localization of lesions	No. of outbreaks and duration of evolution	Systemic/extracutaneous symptoms	Laboratory investigations	Histopathologic analysis	Associated diseases	References
51, woman	Scalp, forehead, trunk, upper extremities	5 to 6 outbreaks in $\approx$ 5 y	No fever	No leukocytosis or elevated ESR, normal ANA and Borrelia serology result	Dense and diffuse neutrophilic infiltrate in the mid dermis, subepidermal edema	No associated disease	Christensen et al <sup>3</sup>
38, woman	Neck, left cheek, right leg	5 outbreaks in $\approx$ 1 y	No fever, no deterioration of general health	No leukocytosis or elevated ESR, normal liver enzyme levels	Dense and diffuse neutrophilic infiltrate in the mid dermis, subepidermal edema	No associated disease	Christensen et al <sup>3</sup>
41, woman	Right forearm, neckline	4 outbreaks in 1 y of evolution	No extracutaneous symptoms	No leukocytosis, no elevation of CRP, negative Borrelia serology and ANA result	Dense neutrophilic infiltrate and dermal edema	None	Croci-Torti et al <sup>4</sup>
38, woman	Legs	2 outbreaks in a few months	No systemic or extracutaneous symptoms	No leukocytosis, no elevation of CRP level, negative ANA and Borrelia serology result	Dense neutrophilic infiltrate and dermal edema	None	Croci-Torti et al <sup>4</sup>
47, woman	Left forearm, right shoulder	3 outbreaks in $\approx$ 1 y of evolution	No systemic complaint, no fever	Normal hemogram result, elevated ESR (63 mm/h), negative ANA result	Papillary dermal edema and dense perivascular infiltrate mainly comprising neutrophils, mononuclear cells, and histiocytes. No vasculitis.	No underlying disease	Ghosh et al <sup>6</sup>
73, woman	Face, top of the back, shoulders, arms	Multiple flare-ups during 6 mo	No fever or systemic clinical signs	No leukocytosis, no inflammatory syndrome. Serum protein electrophoresis showed monoclonal gammopathy IgG $\kappa$ type.	Dense neutrophilic infiltrate of the dermis without vasculitis	Monoclonal gammopathy IgG $\kappa$ type	Blaizot and Doutre <sup>7</sup>
66, woman	Left ankle with a centrifugal extension toward the left hip	Unique clinical course	No fever, malaise, myalgia, or lymphadenopathy	Normal results for hemogram, biochemistry, CRP, and screening for autoantibodies	Neutrophilic infiltration in the mid and upper dermis, with no edema or vasculitis	Rheumatoid arthritis diagnosed 22 y earlier	Mir-Bonafé et al <sup>8</sup>
42, woman	Forearms	3 outbreaks in 2 y of evolution	No fever, no alteration of general health	No leukocytosis, no increase in CRP level, negative Borrelia serology result	Dense neutrophilic infiltrate and dermal edema. No vasculitis.	Sarcoidosis	Present case

ANA, Antinuclear antibody; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate.

CRAND is a rare neutrophilic dermatosis with a stereotypical clinical presentation. We report a case of CRAND associated with sarcoidosis. It seems therefore justified to carry out a complete evaluation to search for a systemic disease, as for any neutrophilic dermatosis.

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