

Case 5: Rheumatoid Neutrophilic Dermatitis

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CASE SUMMARY

History

A 17-year old female student complained of symmetrical erythematous eruption that occurred over her limbs and face for 10 days. The rash was blanchable, progressing from legs and thighs to upper limbs and face. The extensor surface was predominantly involved. Some lesions were reported to show central vesicles followed by crusting. Those over the right face became confluent to form a plaque. The lesions themselves were asymptomatic. The trunk, scalp and nails were spared. Onset of the rash was followed by bilateral ankle and knee arthritis and swinging fever. There was no history of preceding upper respiratory tract infection or new drug intake.

She had a history of juvenile rheumatoid arthritis with recurrent involvement of ankles and knees since 9 years of age. Her arthritis was managed by paediatricians with occasional non-steroid anti-inflammatory drugs. She also had short stature and delayed pubertal development. Induction of puberty by daily low dose ethinyl-oestradiol was given six months ago. Furthermore she had congenital ano-urethral fistula pending corrective operation.

Physical examination

Her general condition was satisfactory. Blanchable erythematous papules and nodules were distributed bilaterally over the extensor surfaces of her legs, thighs, upper limbs and face (Figure 1). Vesicles with crusting developed over some of the lesions. An erythematous non-tender plaque was present on her right face (Figure 2). Bilateral knee and ankle arthritis were present. The eyes and fundi were unremarkable.



Figure 1: Bilateral erythematous papules over both arms. Note vesicle formation and crusting on some lesions.



Figure 2: Erythematous plaque on right face in the acute stage

Differential diagnoses

The differential diagnoses included rheumatoid neutrophilic dermatitis, Sweet's syndrome and vasculitis.

Investigations

Two skin biopsies were performed showing similar features. A circumscribed collection of neutrophils with leucocytoclasia was found in the upper dermis, and perforating through the overlying epidermis to form a subcorneal pustular collection. Reactive fibrinous deposits were present in the vessels around the neutrophil collection. No frank vasculitis or ulceration was noted. The direct immunofluorescence test was negative. These features were consonant with the diagnosis of rheumatoid neutrophilic dermatitis. Bacterial and viral cultures from the vesicular fluid were both negative.

The patient also had leucocytosis ($12.4 \times 10^9/L$, more than 80% being polymorphs) and hypochromic microcytic anaemia. There was occult gastrointestinal bleeding probably due to gastric erosions which may be drug related. Her erythrocyte sedimentation rate was raised at 77 mm/hour.

Progress

The skin lesions began to regress 3 weeks after onset of rash. No more new lesions were noted 6 weeks from the onset of rash. Residual erythema and post-inflammatory pigmentation were present on old lesions.

A diagnosis of rheumatoid neutrophilic dermatitis was made in view of the clinical picture and the compatible histopathological findings.

Discussion

The pathologist, Dr. W. Y. Lam believes that neutrophilic dermatoses are a group of related conditions (Table 1) characterised by (1) a neutrophilic infiltrate, (2) a lack of micro-organisms on special stains and culture, and (3) clinical improvement on treatment with systemic steroids. Leucocytoclasia is variable in these disorders and there is generally no significant fibrinoid necrosis of vessel walls, but limited vascular damage often present. Erythema elevatum diutinum has prominent vessel damage and is best included as vasculitis and not neutrophilic dermatoses. In all cases, histopathology is not specific enough to rule out infection, cultures and special stains of tissue are essential to exclude a potential infectious aetiology.

Table 1: Neutrophilic dermatoses

- Sweet's acute febrile neutrophilic dermatoses
- Sweet's-like neutrophilic dermatoses in autoimmune disease, pregnancy, and lymphoproliferative disorders
- bowel-associated dermatosis-arthritis syndrome
- rheumatoid neutrophilic dermatosis
- acute generalised pustulosis / pustular bacterid
- pyoderma gangrenosum

REVIEW ON RHEUMATOID NEUTROPHILIC DERMATITIS

Rheumatoid neutrophilic dermatitis (RND) is a recently recognised, rare cutaneous manifestation of rheumatoid arthritis, first described by Ackerman in 1978.¹ About 14 cases have been reported in the English literature.²⁻⁹

Clinical features^{3, 8, 10}

There are symmetrical erythematous papules and plaques with occasional vesicles occurring over extensor surfaces of proximal limbs in patients with moderate to severe rheumatoid arthritis. Lesions are relatively asymptomatic. Crusted superficial ulcerations may occur. The duration of individual lesions varies from 1 to 3 weeks. Resolution occurs spontaneously or with improvement of the underlying rheumatoid arthritis. It usually heals without scarring but temporary hyperpigmentation may result.

Histopathology^{8, 10}

Dense perivascular and interstitial neutrophilic infiltrates occur in upper and mid-dermis, extending into the subcutaneous fat. Leucocytoclasia may or may not be present, but overt leucocytoclastic vasculitis is absent. Papillary dermal neutrophilic microabscesses may be present. Spongiotic intraepidermal vesicles or subepidermal vesicle formation are occasionally seen.

Diagnosis

Diagnosis is made on clinical grounds, with the support of compatible histological features. The most difficult differential diagnosis is Sweet's syndrome. In

Sweet's syndrome the lesions are typically tender, oedematous plaques whereas those of rheumatoid neutrophilic dermatitis are relatively asymptomatic. Rheumatoid neutrophilic dermatitis generally lacks the extracutaneous involvement seen in Sweet's syndrome and frequently the inflammatory infiltrates are not as dense and extensive or associated with prominent dermal oedema as in Sweet's syndrome.¹⁰ On the other hand, some investigators consider rheumatoid neutrophilic dermatitis as a variant of Sweet's syndrome.^{2, 8, 10} Erythema elevatum diutinum can be distinguished clinically by its chronic course, and the presence of intense leucocytoclastic vasculitis histologically. The need to rule out a potential infectious aetiology has already been stressed.

Pathogenesis

The pathogenesis is not well understood. In addition to rheumatoid neutrophilic dermatitis, several other skin conditions have been reported to occur with rheumatoid arthritis; all exhibiting neutrophilic reaction in the absence of vasculitis (e.g. pyoderma gangrenosum, Sweet's syndrome).⁷ In rheumatoid arthritis, activated fibroblast-like cells in the synovium produce soluble inflammatory mediators that induce cartilage and bone changes. The same process may occur in the skin, where cutaneous fibroblasts generate neutrophil chemoattractants. The recruited neutrophils may be responsible for the inflammatory reaction in this disorder.¹¹ Processes that may play a role include immune complex activation, cell adhesion and migration, and cytokine release.⁸

Management

There have only been a few publications on specific therapy. Difficulty arises because many of the reported patients have also received second line anti-arthritic agents, and rheumatoid neutrophilic dermatitis often resolves spontaneously with improvement of underlying rheumatoid arthritis.^{7, 8}

Dapsone has been tried.⁶ Other possible drugs that may be used include topical or systemic steroid,⁵ and hydroxychloroquine,³ etc.

Conclusion

Rheumatoid neutrophilic dermatitis is a rare cutaneous manifestation of rheumatoid arthritis and its tendency towards spontaneous resolution needs to be considered in its management.

Learning points:

In suspected neutrophilic dermatoses, histopathology is not specific enough to rule out an infection. Cultures and special stains are essential to exclude a potential infectious aetiology.

References

1. Ackerman AB. Histologic diagnosis of inflammatory skin diseases: a method by pattern analysis. Philadelphia: Lea & Febiger; 1978; 449-451.
2. Scherbenske JM, Benson PM, Lupton GP, Samlaska CP. Rheumatoid neutrophilic dermatitis. Arch Dermatol 1989; 125:1105-1108.
3. Sánchez JL, Cruz A. Rheumatoid neutrophilic dermatitis. J Am Acad Dermatol 1990; 22:922-925.
4. Lowe L, Kornfeld B, Clayton J, Golitz LE. Rheumatoid neutrophilic dermatitis. J Cutan Pathol 1992; 19:48-53.
5. Yamamoto T, Ohkubo H, Nishioka K. Rheumatoid neutrophilic dermatitis. Int J Dermatol 1994; 33(11):798-800.
6. Harkaway K, Elenitas R, Margolis D. Erythematous papules in a patient with rheumatoid arthritis. Arch Dermatol 1995; 131:1195-1200.
7. Hassab-el-Naby HMM, Alsaleh QA, Khalifa MA. Rheumatoid neutrophilic dermatitis. Int J Dermatol 1996; 35(3):207-208.
8. Mashek HA, Pham CT, Helm TN. Rheumatoid neutrophilic dermatitis. Arch Dermatol 1997; 133:757-760.
9. Hirota TK, Keough GC, David-Bajar K, McCollough ML. Rheumatoid neutrophilic dermatitis. Cutis 1997; 60:203-205.
10. Templeton S-F, Jorizzo JL, Solomon AR. Neutrophilic dermatoses. In: Arndt KA, Robinson JK, Wintroub BU, LeBoit PE, eds. Cutaneous medicine and surgery. Philadelphia: WB Saunders, 1996; 372-378.
11. Lovell CR, Maddison PF. Rheumatoid arthritis and the skin. In: Champion RH, Rye RJ, eds. Recent advances in dermatology. Edinburgh: Churchill Livingstone, 1992; 1-16.