A cutaneous-only variant of rheumatoid vasculitis with symmetric flexural pyoderma gangrenosum-like lesions

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Rheumatoid vasculitis, an uncommon extraarticular manifestation of rheumatoid arthritis, usually develops in long-standing cases affecting small-to-medium-sized vessels. It is a poor prognostic marker when multiple systems are affected, skin and neurologic involvement being most frequent. Rheumatoid vasculitis is seen more commonly in seropositive and nodular rheumatoid arthritis patients who are male or smoke. Herein, we present the peculiar case of a 48-year-old female with rapid onset, progressive, multiple ulcers predominantly over the flexures. Differential diagnoses of pyoderma gangrenosum, pyoderma vegetans, pemphigus vegetans, and vasculitis were considered, and the patient was evaluated. On investigation, her rheumatoid factor titer was strongly positive; a skin biopsy revealed leukocytoclastic vasculitis. On clinical, serological, and histopathological correlation, a diagnosis of rheumatoid vasculitis was made, responding well to high-dose prednisolone. On follow-up at six months, her skin lesions had healed well with moderate scarring; however, she developed severe joint pain, warranting the initiation of disease-modifying antirheumatic drugs. To the best of our knowledge, this is a previously unreported clinical and morphological presentation of rheumatoid vasculitis.

Keywords: rheumatoid vasculitis, rheumatoid arthritis, cutaneous-only vasculitis, secondary vasculitis.

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INTRODUCTION

Rheumatoid vasculitis is a relatively uncommon disease manifestation in patients with rheumatoid arthritis [1]. The morphological pattern can include palpable purpura, nodulo-ulcerative lesions, livedo reticularis, erythema elevatum diutinum, and hemorrhagic bullae. Bannatyne first described vascular involvement in rheumatoid arthritis in 1898 [2]. Rheumatoid vasculitis may occur in association with pyoderma gangrenosum, though it can also present alone with ulcerated lesions [3,4]. Herein, we present a peculiar case of a 48-year-old female with multiple, rapid-onset, progressive ulcers predominantly over the flexures.

CASE PRESENTATION

A 48-year-old lady came to our emergency room with a one-week history of multiple, rapid-onset, progressive, somewhat painful, ulcerated lesions, predominantly over the flexures. She had a history of documented low-grade fever associated with these lesions. Mild arthralgia and morning stiffness in the shoulder, wrist, and small joints of the hands were noted on and off for the last three months. The symptoms were mild and sporadic, so the patient did not seek any specific treatment. She was also a known hypertensive and diabetic.

The systemic examination revealed no abnormalities. On mucocutaneous examination,

multiple, well-defined, circular-to-elongated ulcers with few smaller satellite lesions were present over the right axilla, bilateral inframammary folds, and bilateral groins (Figure 1 a-c). The margins were punched out, and the floor of the ulcers had adherent yellowish to brown-black crusts (Figure 1 a-c). Similar isolated lesions were seen over the abdomen and thighs. No regional lymphadenopathy was noted, and the mucosae were unremarkable.

Routine & relevant investigations, including a complete blood count (CBC), renal function test (RFT), liver function test (LFT), urine microscopy, chest X-ray (CXR) & electrocardiogram (ECG), were within normal limits. A screening anti-nuclear antibody (ANA) [ELISA] and an anti-neutrophilic cytoplasmic antibody (ANCA) profile were sent and returned negative. However, her rheumatoid arthritis (RA) factor (46.9 IU/ml) was strongly positive, and the erythrocyte sedimentation rate (ESR) was extremely elevated at 120 mm/hr. The pathergy test was negative. A skin biopsy, keeping differential diagnoses of pyoderma gangrenosum, pyoderma vegetans, pemphigus vegetans, and vasculitis, was done after informed consent.

The biopsy revealed epidermal necrosis and ulceration; the dermis had a dense neutrophilic infiltrate in and around the vessel wall. Endothelial cell swelling, leukocytoclasia, and extravasation of red blood cells (RBCs) were present, and fibrinoid necrosis was seen (Figure 2 a-b).

On clinicopathological correlation, a provisional diagnosis of rheumatoid vasculitis was made. She was started on high-dose prednisolone (1 mg/kg/day), showing marked improvement, The steroid dose was gradually tapered over two months, and the lesions healed well with post-inflammatory hyperpigmentation and mild scarring (Figure 3).

On follow-up at six months, she developed severe joint pain restricting her movements, and a rheumatological consult was requested. She subsequently fulfilled the EULAR (European League Against Rheumatism) criteria for rheumatoid

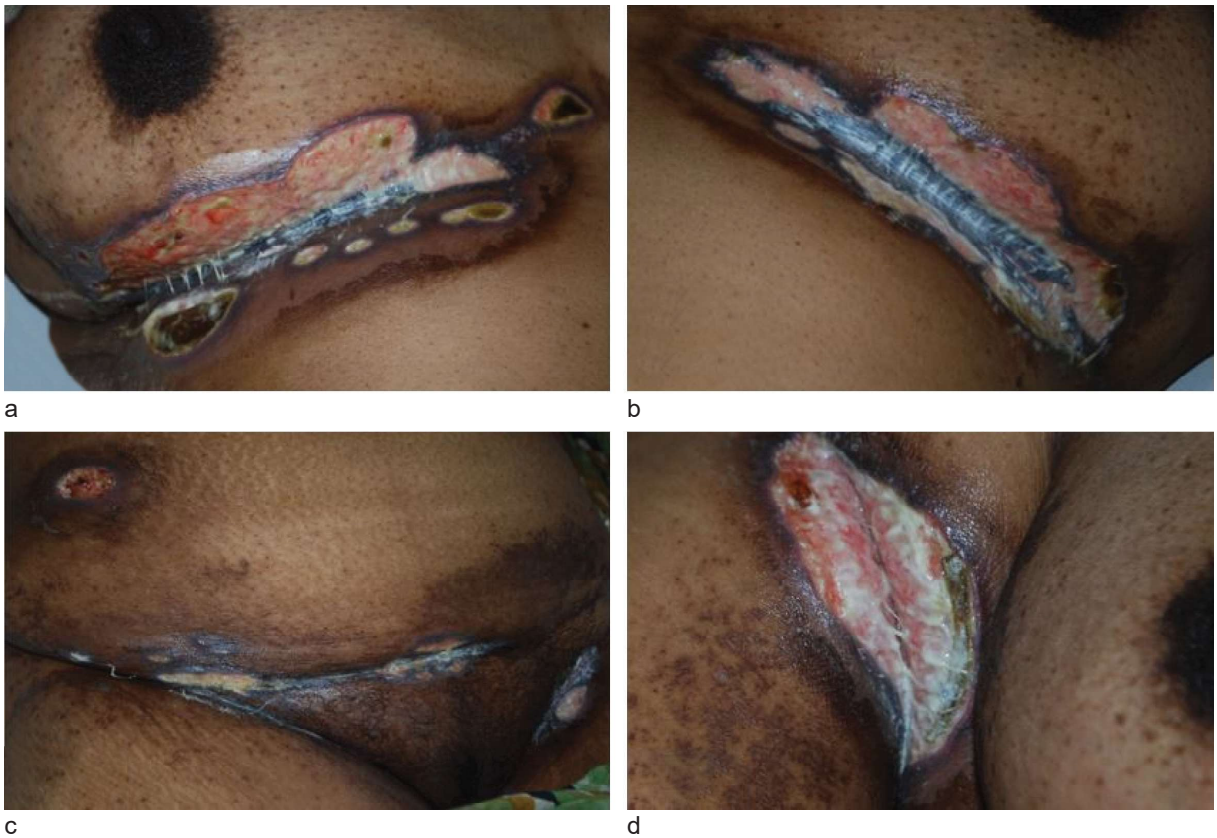
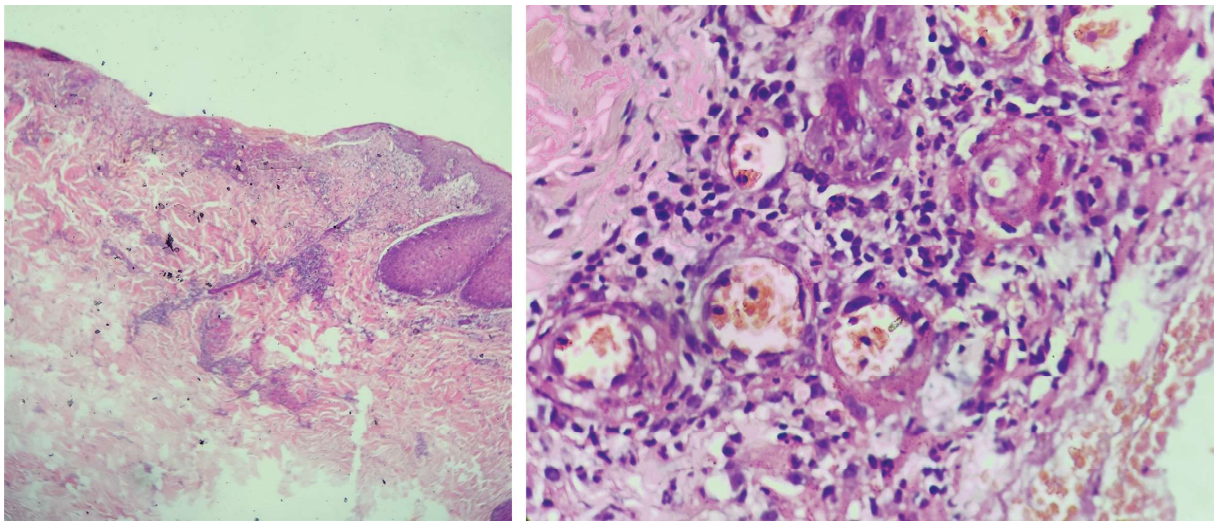


Figure 1. Multiple ulcers with punched-out margins; floor covered with yellowish necrotic slough and brown-black crusts in the (a) right axilla, (b) right infra-mammary fold, and (c) right groin



a b

Figure 2. Skin biopsy (hematoxylin & eosin stain) showing epidermal ulceration and necrosis. The dermis shows infiltration of the vessel wall by neutrophils with fibrinoid necrosis, leukocytoclasia, endothelial cell swelling, and extravasation of erythrocytes; (a) 100×; (b) 400×

arthritis (score = 8) and was started on a disease-modifying anti-rheumatoid drug (DMARD). Hence, a final diagnosis of rheumatoid vasculitis was made, where the vasculitic lesions preceded the development of frank rheumatoid arthritis.

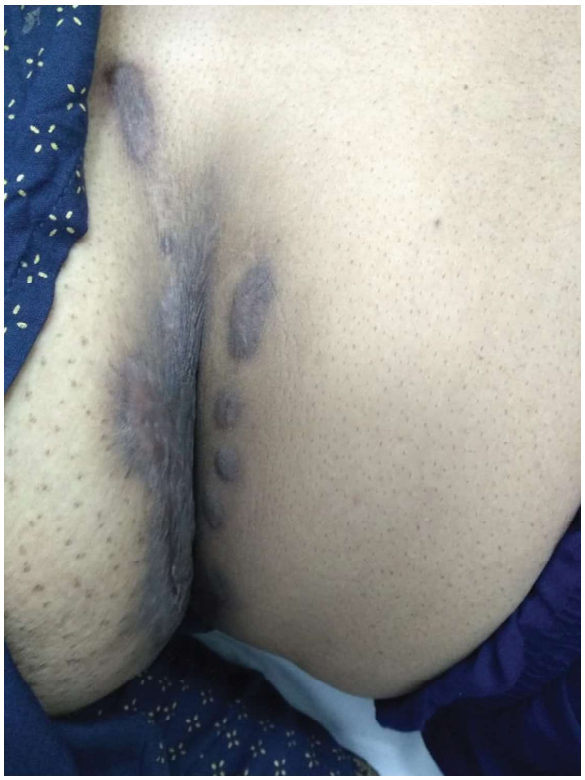


Figure 3. Healed lesions with post-inflammatory hyperpigmentation and scarring over the right inframammary region at the six-month follow-up

DISCUSSION

Rheumatoid vasculitis is seen in approximately 1–5% of patients with rheumatoid arthritis [5-7]. Due to the morphology and rapid onset, our first impression was an atypical pyoderma gangrenosum, even though the lesions were only slightly painful, with minimal tenderness on palpation. However, biopsy findings did not corroborate this. Since pyoderma gangrenosum is a diagnosis of exclusion, we considered other possibilities. Wegener's granulomatosis, a small vessel vasculitis, has been reported to present with such pyoderma gangrenosum-like lesions in the flexures [8]. However, the ANCA profile was negative, and no granulomas were seen.

Thus, on clinical, serological, and histopathological grounds, we reached a diagnosis of cutaneous-only rheumatoid vasculitis. To the best of our knowledge, such symmetric flexural pyoderma gangrenosum-like lesions have not been previously reported in rheumatoid vasculitis.

Rheumatoid vasculitis usually presents after a mean rheumatoid arthritis disease duration of 13 years, with anecdotal reports of vasculitis preceding or occurring early in rheumatoid arthritis (Table 1). Rheumatoid vasculitis preceding or occurring in

Table 1. Rheumatoid vasculitis (RV) presentations preceding or in early stages of rheumatoid arthritis (RA)

No.	Author, year	Age (yrs.) /sex	Features of RV	Associated arthritic features	Temporal relation of RV with RA
1.	Parker *et al.*, 2007 [9]	63/M	1. Mononeuritis multiplex 2. Acute abdomen: abdominal pain, nausea, and vomiting due to small bowel perforation	Symmetrical inflammatory polyarthritis of small joints	RV 7 months after RA diagnosis
2.	Sandhu *et al.*, 2013 [10]	74/M	Acute abdomen: periumbilical & right upper quadrant pain; recurrent emesis due to cholecystitis	Symmetrical inflammatory polyarthritis of small joints	RV 1 week after RA diagnosis
3.	Tourin *et al.*, 2013 [11]	61/M	Acute pulmonitis: pleuritic chest pain & dry cough	Symmetric inflammatory polyarthritis of small joints	RV 2 months prior to RA diagnosis
4.	Lee *et al.*, 2017 [12]	72/F	Hepatic artery vasculitis: elevated liver enzymes only, asymptomatic	Symmetric inflammatory polyarthritis of small joints	RV 2 months after RA diagnosis
5.	Sacks *et al.*, 2017 [13]	38/M	Mononeuritis multiplex	Symmetrical inflammatory polyarthritis of small joints	RV 6 years prior to RA diagnosis
6.	Anwar *et al.*, 2019 [14]	44/M	1. Nodulo-ulcerative cutaneous vasculitis 2. Pericarditis	Symmetrical inflammatory polyarthritis of small joints	Concurrent diagnosis of both.
	Our case	48/F	Cutaneous-only variant with multiple, symmetric, flexural pyoderma gangrenosum-like lesions	Symmetrical inflammatory polyarthritis of small joints	RV 6 months prior to RA diagnosis

early rheumatoid arthritis has been seen as an acute pulmonary syndrome due to vasculitis and even as an acute abdomen in the form of cholecystitis, hepatic artery involvement, and acute bowel perforation [9-12]. Since rheumatoid vasculitis is usually a systemic vasculitis, an unusual feature in our case was the cutaneous-only pattern.

CONCLUSION

We reported an unprecedented presentation of cutaneous-only rheumatoid vasculitis preceding overt rheumatoid arthritis. Histopathology plays a crucial role in diagnosis; since rheumatological diseases evolve with time, following up on such patients may improve our diagnostic ability and therapeutic interventions.

Conflict of interest: None declared.

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