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ANNULAR LEUKOCYTOCLASTIC VASCULITIS: A NEW FEATURE OF IGA VASCULITIS

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ABSTRACT

Background: Annular erythema is a rare manifestation of leukocytoclastic vasculitis. It may be associated with various drugs, infections, malignancies, or systemic diseases.
Case description: A 36-year-old woman with no personal medical history presented with annular erythema with target lesions and petechial purpura. The patient had fever and joint arthralgia. A skin biopsy showed leukocytoclastic vasculitis with IgA deposits on direct immunofluorescence. The diagnosis of immunoglobulin A vasculitis with annular leukocytoclastic vasculitis was made. The patient showed global improvement with topical steroids without relapse.
Conclusion: An annular variant of leukocytoclastic vasculitis is a rare manifestation of immunoglobulin A vasculitis.

KEYWORDS

Annular erythema, Ig-A mediated vasculitis, leukocytoclastic vasculitis, vasculitis, topical corticosteroids

LEARNING POINTS

- Annular erythema may reveal an IgA vasculitis.
- Annular leukocytoclastic vasculitis is a rare manifestation of IgA vasculitis.
- This presentation is treated with topical steroids.

INTRODUCTION

Annular erythema is a group of chronic, annular and erythematous skin rashes that can occur at any time of life, from infancy to old age. It most commonly affects the thighs and legs, but can also occur on the face, trunk, and arms. The lesions rarely cause mild itching or stinging[1]. Sometimes it is associated with underlying diseases and conditions such as drug use, bacterial and viral infections, Graves' disease, cholestatic liver disease and food. But often no specific cause of annular erythema is found[2]. It can be confirmed by histopathology, which shows a perivascular lymphocytic infiltration, and rarely, annular leukocytoclastic vasculitis (ALV)[3]. Here, we report a case of ALV as the first clinical feature of immunoglobulin A vasculitis (IgAV).

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CASE DESCRIPTION

A 36-year-old woman with no personal medical history presented with a macular erythematous rash with raised and infiltrated edges, confluent in places forming a large annular dermatitis, non-itchy, non-painful and non-oozing, located on the forearms *(Fig. 1A)*. She also had a circular lesion in concentric rings (target lesions) located on the fingers *(Fig. 1B)*. The patient described the onset of the eruption as a small, raised pink-red spot that slowly enlarged and formed a ring shape, with the central area flattening and clearing. The rings enlarged progressively to reach a diameter of approximately 6 cm on the forearms and 3 cm on the fingers. Moreover, she developed petechial and necrotic purpura of the lower limbs *(Fig. 1C)*. She complained of arthralgia of the knees and ankles with long-lasting fever of one week. There were no additional mucosal lesions. She had no abdominal pain, blood in the stool, recent drug use or toxic exposure. Physical examination revealed a high temperature of 38 °C, heart rate of 80 beats per minute, blood pressure of 130/80 mmHg, a normal respiratory rate of 16/min and normal cardiorespiratory auscultation. Routine laboratory tests showed leukocytes at 10.56 g/l (normal range 4–10 g/l) with neutrophils at 6.76 g/l (normal range 1.5–7 g/l), haemoglobin at 12.2 g/l (normal range 11.5–15 g/l) and platelets at 312 g/l (normal range 150–450 g/l). The C-reactive protein level was 62 mg/l (normal range: <3 mg/l). Liver tests, serum creatinine and electrolytes were within the normal ranges. The 24-hour proteinuria test was negative. Serum electrophoresis and immunoglobulin (IgG, IgA and IgM) levels were normal. Hepatitis B virus, hepatitis C virus and HIV serologies were negative. Antinuclear antibodies, antineutrophil cytoplasmic antibodies, cryoglobulinemia, rheumatoid factors, and C3/C4 levels were normal. A computed tomography scan and cardiac echogram were normal. A skin biopsy showed perivascular infiltration of neutrophils, nuclear dust, extravasated red blood cells with endothelial swelling, and fibrinoid necrosis of the vessel wall. Direct immunofluorescence testing showed perivascular deposition of IgA *(Fig. 2)*. The diagnostic of ALV related to IgAV was made. Treatment consisted of topical corticosteroids and bed rest, with a favourable outcome. The skin lesions cleared rapidly, and joint tenderness, fever and biological abnormalities normalised within one week. Topical corticosteroids were continued for one month. At 12 months follow-up, the patient was asymptomatic and considered to be in complete remission.

DISCUSSION

To the best of our knowledge, this is the first observation of ALV related to IgAV. As mentioned by Meissner et al.[4], ALV was first described by Degos and Guiliane. It is histologically defined by neutrophilic invasion and fibrinoid necrosis around small dermal blood vessels.

Cribier et al.[5] describe ALV as a distinct subtype that fulfils the following criteria: (1) multiple attacks over years with sudden onset and spontaneous resolution after 7–10 days; (2) annular purpuric patches with centrifugal extension; (3) extension to the limbs and trunk producing polycyclic lesions that clear, leaving mild haemosiderin deposition; (4) no extracutaneous symptoms and good general health; (5) histological changes of leukocytoclastic vasculitis with mild vascular changes and intense polymorphonuclear cell infiltration, and (6) complete clearance of all lesions with dapsone.

We performed a literature review of reported ALV. *Table*

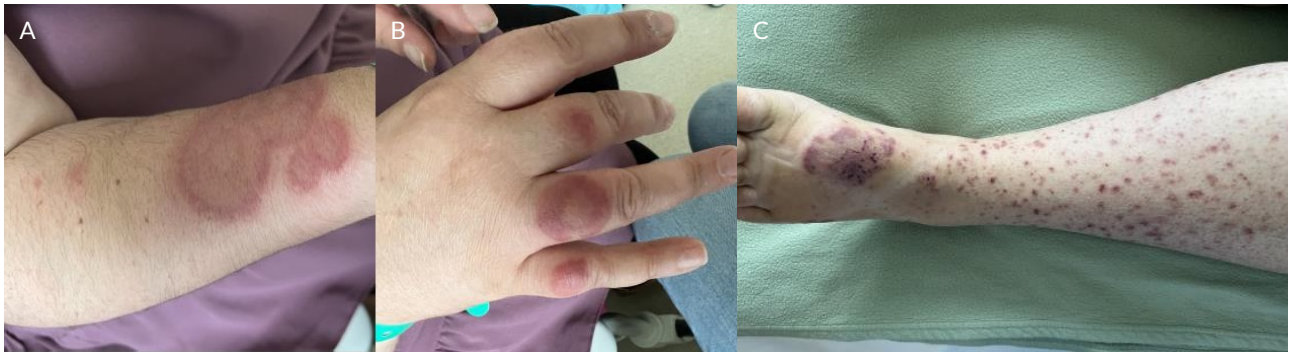

Figure 1. Skin lesions: A) annular lesions, B) target lesions and C) petechial purpura.

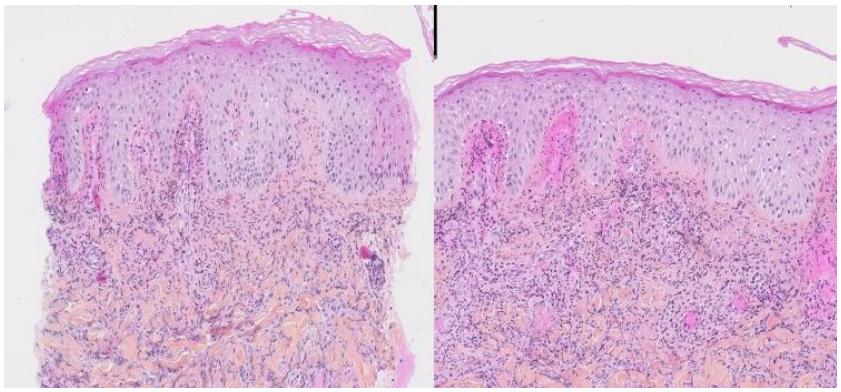

Figure 2. Leukocytoclastic vasculitis on the forearms: A) Haematoxylin and eosin magnification ×20; B) Haematoxylin and eosin magnification ×64.

	Gender	Age (years)	Skin manifestation	Mucosal lesions	IFD	Systemic manifestation	Final diagnosis	Treatment	Outcome	Follow-up	PIH
Noussari et al., 2000[7]	Woman	76	erythematous purpuric papules, enlarged into annular plaques	No	IgG, IgM, and C3 deposits	Weight loss	MGUS	No	Improvement	ND	ND
Nakajima et al., 2000[8]	Woman	79	large, confluent purpuric violaceous erythema with annular configuration small palpable purpura	No	Fibrinogen deposits	nausea and diarrhea	Sjogren's syndrome	No	Spontaneously improvement	ND	Yes
Chiu et al, 2004[9]	Man	71	pruritic polycyclic purpura with centrifugal extension	No	-	Renal involvement	Drug: chlorzoxazone	Topical steroids Oral prednisone	Improvement	ND	Yes
Meissner et al., 2007[6]	Man	68	multiple annular-shaped purpuric lesions	No	-	No	chronic HBV infection with mixed cryoglobulinaemia	Dapsone 200mg/ day	Improvement	ND	ND
Yasukawa et al., 2008[10]	Man	51	several double annular pupura up to 10 cm in diameter	No	IgM deposits	No	T-cell lymphoma with polyclonal immunoglobulin G	Topical steroids	Improvement	1 year	ND
Meissner et al., 2008[11]	Woman	82	rapidly progressing annular-shaped purpuric lesion with bullous transformation	No	-	No	generic of amlodipinebesylat (Povidone K30, Povidone K90 and lactose.)	Methylprednisolone 1 mg/kg weigh	Improvement	ND	ND
Hong et al., 2011[6]	Woman	66	painful confluent annular erythema	No	-	No	ulcerative colitis	dapsone aceclofenac levocetirizine fexofenadine	Improvement	ND	No
Chanprapah et al., 2013[12]	Man	62	erythematous-to-purplish, non-blanchable macules and papules with an annular arrangement	No	-	No	Drug: anti-tuberculosis medication	No	Improvement	ND	Yes
Matsuzaki et al., 2016[13]	Woman	79	petechiae and erythematous papules, with a linear ringlike arrangement	No	IgA and C3 deposits	No	idiopathic mixed cryoglobulinemia	Prednisone 30mg/ day	Improvement than relapse	ND	no
Gambichler et al., 2020[14]	Man	28	well-defined annular purpuric lesions	Palatal petechiae	C3, C4, and fibrinogen deposits	Episcleritis	PR3-ANCA-positive Active ulcerative colitis	Prednisolone Mesalazine	Improvement	ND	ND
Çetinarslan et al., 2020[15]	Man	57	Multiple annular midatrophic erythematous plaques	No	-	No	Acute myeloblastic leukemia	Topical starost Cytosine arabinoside C	Improvement	8 months	ND
Our case	Woman	36	Large annular dermatitis and petechial purpura	No	IgA deposits	Arthromyalgia	IgA vasculitis	Topical steroids	Improvement	1 year	Yes

Abbrevations: ND, not determined; MGUS, monoclonal gammopathy of unknown significance; PIH, post inflammatory hyperpigmentation; HBV, hepatitis B virus.

Table 1. Etiological specter of ALV: Literature review of annular leukocytoclastic vasculitis since 2000.

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1 summarises the epidemiological, clinical presentation, aetiological spectrum, and management of ALV. In our case, the patient improved rapidly with topical steroids without relapse. In previous cases, dapsone seems to have been effective as well[4,6]. Typically, patients with IgAV present with palpable skin purpura predominantly on the lower limbs. It can extend to the upper extremities and on the trunk. Some patients showed necrotic lesions, ulcerations or bullae.

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