Received: 6 February 2024 | Revised: 23 March 2024 | Accepted: 29 March 2024

DOI: 10.1002/ccr3.8769

CASE REPORT

Clinical Case Reports Open Access WILEY

Leukocytoclastic vasculitis in a patient with ankylosing spondylitis: A case report

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Key Clinical Message

Although the concurrent occurrence of vasculitis with AS is uncommon, when patients diagnosed with AS exhibit symptoms including skin petechiae, purpura, abdominal discomfort, malaise, elevated ESR, and reduced complement levels, vigilant monitoring for vasculitis is advisable following the exclusion of secondary vasculitis triggers such as malignancies, infections, and pharmaceutical agents.

Abstract

The primary characteristic of ankylosing spondylitis (AS) involves inflammation occurring within the sacroiliac joint and the spine, leading to destruction and eventual ankylosis. A notably infrequent complication associated with AS is vasculitis, with limited reports linking AS to vasculitis. This case study documents a 48-year-old male, diagnosed with HLA-B27-positive AS for the past 15 years, who developed abdominal pain and skin lesions following the cessation of his medication on his own. Subsequent clinical evaluations identified leukocytoclastic vasculitis (LCV) related to AS after excluding all other potential causes of LCV, including drug-related sources, cancer, hepatitis B and C viruses, Henoch-Schönlein purpura (HSP), and IgA nephropathy.

KEYWORDS

ankylosing spondylitis, cutaneous vasculitis, leukocytoclastic vasculitis, small vessel vasculitis

1 | INTRODUCTION

Ankylosing spondylitis (AS), the predominant condition within the spectrum of spondyloarthropathies, is a chronic inflammatory disease impacting the spine, peripheral joints, and tendons. A defining characteristic of AS is the inflammation observed in the sacroiliac joints and spinal column, resulting in ankylosis.[1] According to a comprehensive systematic review and meta-analysis, the prevalence of ankylosing spondylitis (AS) fluctuate from 0.02% in Sub-Saharan Africa to 0.35% in communities located in the northern Arctic region.[2] AS extends beyond skeletal involvement, influencing non-skeletal organs such as the eyes, lungs, and kidneys. Vasculitis stands as an exceedingly uncommon complication of AS, potentially leading to severe outcomes including cardiovascular

ailments, ocular (iris) complications, and aneurysms.[3] Although rare, instances linking spondyloarthropathies to vasculitis have been documented. This article delves into a case where AS coincided with leukocytoclastic vasculitis (LCV).

2 | CASE HISTORY AND EXAMINATION

A 48-year-old male with a history of AS presented at the rheumatology outpatient department due to abdominal discomfort and dermal manifestations that began approximately 1 month prior. The diagnosis of AS was established based on the ASAS classification criteria, which included chronic inflammatory low back pain, evidence of bilateral sacroiliitis via magnetic resonance imaging (MRI), presence of HLA-B27 antigen, and a history of uveitis 15 years ago. For the past 6 years, his treatment regimen included 500 mg of sulfasalazine administered twice daily and 50 mg of indomethacin also taken twice daily, leading to a disease state of remission over the past 5 months. He was hospitalized in the rheumatology department for both diagnosis and management of suspected vasculitis. The patient had ceased his medications 4 months prior for reasons not disclosed. About a month before his hospital admission, he noticed skin lesions characterized by petechiae, palpable purpura, and erythematous patches across his lower limbs, buttocks, the upper section of his back, more mildly on both upper limbs, and on the surface of his ears (Figure 1A,B). He reported widespread, mild joint pain, akin to generalized arthralgia, affecting most body joints with progressively worsening intensity, epigastric pain, and a productive cough. Further assessments identified emphysematous changes and apical opacity in the lower lobe of the left lung. The patient's medical history included a right hip joint arthroplasty and a 20-year history of smoking one pack a year. An upper gastrointestinal endoscopy was performed due to the epigastric pain, revealing a gastric ulcer, which was subsequently managed with omeprazole. Laboratory tests indicated elevated Erythrocyte Sedimentation Rate (ESR) at 78 mm/hr and decreased C3 complement levels at 61 (normal range: 90–180).

3 | METHODS (DIFFERENTIAL DIAGNOSIS, INVESTIGATIONS AND TREATMENT)

Owing to the prolonged use of indomethacin, its cessation by choice 4 months prior, and the emergence of petechiae and purpura 1 month thereafter, the possibility of indomethacin-associated LCV was not initially contemplated for the patient.[4] The diagnosis leaned towards vasculitis based on both clinical observations and paraclinical findings, which included symptoms of stomach ache, general malaise, headache, widespread myalgia, joint pain,

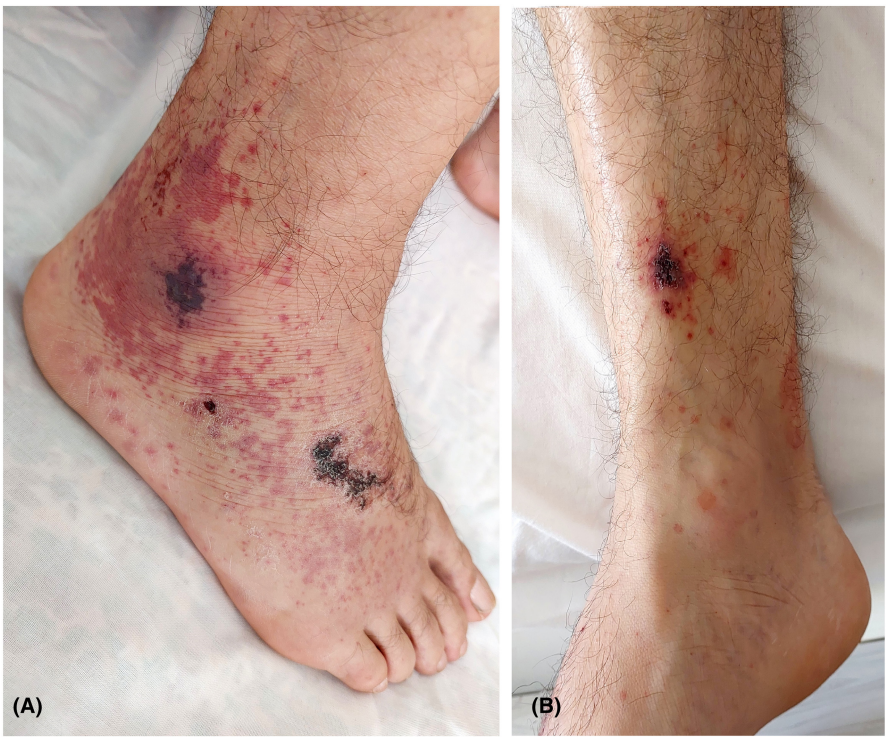

FIGURE 1 Skin lesion in favor of vasculitis associated with petechiae and purpura.

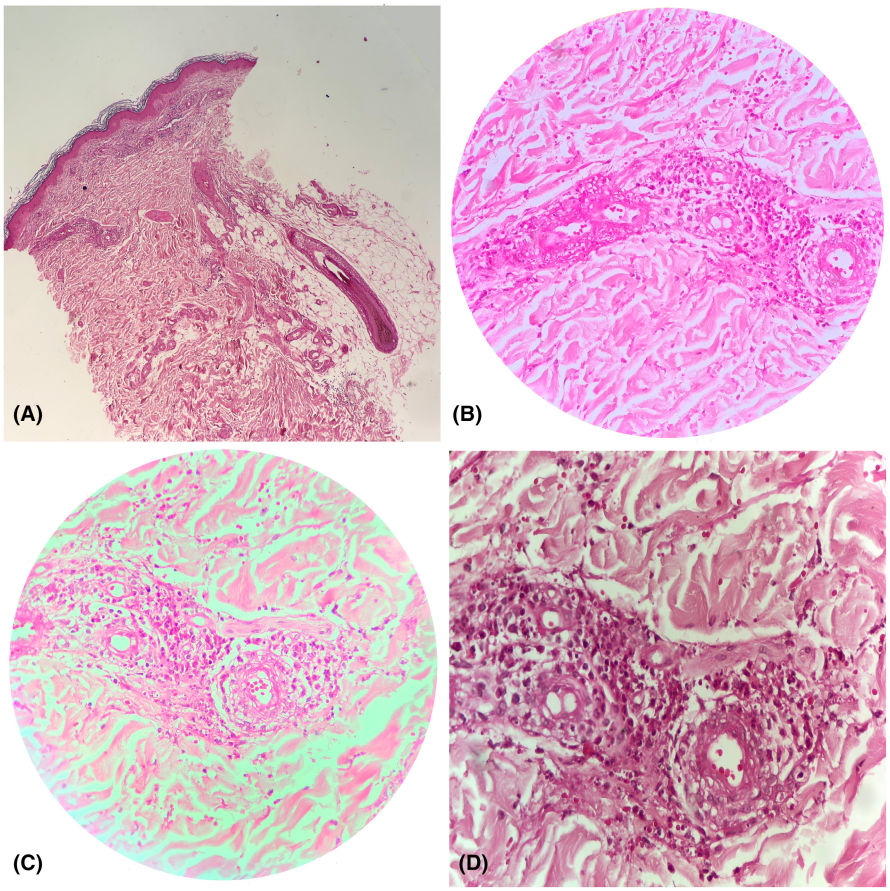

FIGURE 2 Skin punch biopsy shows vasculitis.

and the presence of disseminated petechiae and palpable purpura on the limbs, accompanied by a decrease in C3 levels and an increase in ESR. A diagnostic skin punch biopsy from the affected areas revealed the presence of inflammatory cellular infiltration and fibrinoid necrosis within the walls of dermal vessels (Figure 2), leading to the diagnosis of LCV in an AS patient, supported by clinical, laboratory, and pathological evidence. Various etiologies, including drugs, infections, dietary factors, autoimmune disorders, connective tissue diseases, and malignancies, have been proposed as contributors to the development of LCV. Subsequent evaluation, including ANA, RF, ANCA antibodies, complement levels, HIV, hepatitis B and C serologies, serum IgA, IgG, and cryoglobulins, were all within normal limits. Screenings for cancer also yielded negative results. Although arthralgia can manifest in AS, it was attributed to LCV in this patient due to the remission of AS for the past 5 months and the rarity of small joint arthralgia in AS cases. Thus, the patient's arthralgia, along with skin involvement, was more indicative of constitutional symptoms of LCV than AS manifestations. Treatment commenced with a high dosage of glucocorticoids for three consecutive days, supplemented with cyclophosphamide. Subsequently, the patient exhibited clinical amelioration and was discharged a few days later.

4 | CONCLUSION AND RESULTS (OUTCOME AND FOLLOW-UP)

Given the infrequent co-occurrence of vasculitis with AS, when AS patients exhibit symptoms including skin petechiae and purpura, abdominal discomfort, general malaise, elevated ESR, and reduced complement levels, it is imperative to consider the possibility of concurrent vasculitis. This vigilance ensures that patients are not precluded from receiving prompt and appropriate treatment through early diagnosis.

5 | DISCUSSION

The association between AS and vasculitis has been seldom reported in the literature. In 2019, Chao Ye and Wenyuan Li[5] described a case of cutaneous vasculitis in a 22-year-old male patient positive for HLA-B27 and diagnosed with AS. Additionally, Mario Luiz Marques Piubelli and colleagues[6] detailed a case involving fatal necrotizing Candida esophagitis in a patient with concurrent LCV and AS. Reports by Senol Kobak et al. and Machet L et al.[7,8] also documented cases of AS coexisting with LCV in individuals aged 26 and 31 years, respectively. Furthermore,

Pàmies A et al.[9] described a case where LCV was associated with the use of golimumab in an AS patient.

To date, there have been five documented instances of LCV in patients diagnosed with AS, with the case presented in our study marking the sixth. Our case involves a 48-year-old male who has been managing HLA-B27-positive AS for 15 years. He presented with abdominal pain and skin lesions following the self-discontinuation of his medications. Subsequent investigations led to the diagnosis of LCV in association with AS. It is hoped that future comprehensive studies will shed light on the connection between spondyloarthropathies and small-vessel vasculitis, thereby refining treatment protocols. The cases reported thus far suggest a potential link between AS and the development of vasculitis, particularly in the presence of skin petechiae and purpura.

AUTHOR CONTRIBUTIONS

Alireza Khabbazi: Supervision; validation; writing – review and editing. **Sepideh Tahsini Tekantapeh:** Conceptualization; data curation; formal analysis; methodology; project administration; resources; writing – original draft. **Cyrus Asadzadeh:** Writing – review and editing. **Amir Vahedi:** Data curation; writing – original draft.

FUNDING INFORMATION

We did not receive funding for this article.

CONFLICT OF INTEREST STATEMENT

We have no conflicts of interest to declare.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are openly available in [repository] at http://doi.org/[doi], reference number [reference number].

ETHICS STATEMENT

The research followed the tenets of the Declaration of Helsinki.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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How to cite this article: Khabbazi A, Tekantapeh ST, Asadzadeh C, Vahedi A. Leukocytoclastic vasculitis in a patient with ankylosing spondylitis: A case report. *Clin Case Rep*. 2024;12:e8769. doi:10.1002/ccr3.8769