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# A case of erythema elevatum diutinum associated with immunoglobulin A vasculitis

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Dear Editors,

Erythema elevatum diutinum (EED) is a rare and chronic cutaneous leukocytoclastic vasculitis [1]. The cause of EED still remains unknown, but immune complex deposition in dermal blood vessels is thought to be associated with the onset of the disease [1]. Here, we report a rare case of EED associated with immunoglobulin A (IgA) vasculitis.

A 72-year-old man presented with arthralgia and palpable purpura on his lower extremities (Figure 1A). There was no history of any antecedent infections, but he had a history of IgA nephropathy and rheumatoid arthritis (RA), which had been treated with oral prednisolone 2 mg/day for 2 months. Laboratory tests showed elevated serum levels of white blood cell count (10,700/μL [normal, 3,300–8,600/μL]), C-reactive protein (3.16 mg/dL [normal, 0.00–0.14 mg/dL]), creatinine (4.01 mg/dL [normal, 0.65–1.07 mg/dL]), and IgA (532 mg/dL [normal, 93–393 mg/dL]). A urinary test showed hematuria and proteinuria. Histopathological findings from palpable purpura on the left lower extremity showed inflammatory infiltration in the upper dermis (Figure 1B). Infiltration of neutrophils and lymphocytes with nuclear dusts of neutrophils was observed around the small vessels in the upper dermis (Figure 1C). Direct immunofluorescence did not detect any immunoglobulin or complement deposition. A diagnosis of IgA vasculitis was made based on the EULAR/PRINTO/PRES classification criteria. The dose of oral prednisolone was increased to 5 mg/day and his symptoms improved for 10 days. However, 1 month later, crusty red–to-violet nodules appeared on his fingers and elbow (Figure 1D). Histopathological findings from the nodule on the left finger showed prominent edema and dense inflammatory infiltration in the dermis (Figure 1E). Infiltration of neutrophils, lymphocytes, eosinophils, and histiocytes with nuclear dusts of neutrophils was observed around the vessels in the dermis (Figure 1F). A diagnosis of EED was made. Although diaminodiphenyl sulfone 100 mg/day was temporarily initiated, the treatment was discontinued due to anemia. The dose of oral prednisolone was increased to 15 mg/day and gradually improved his skin lesions.

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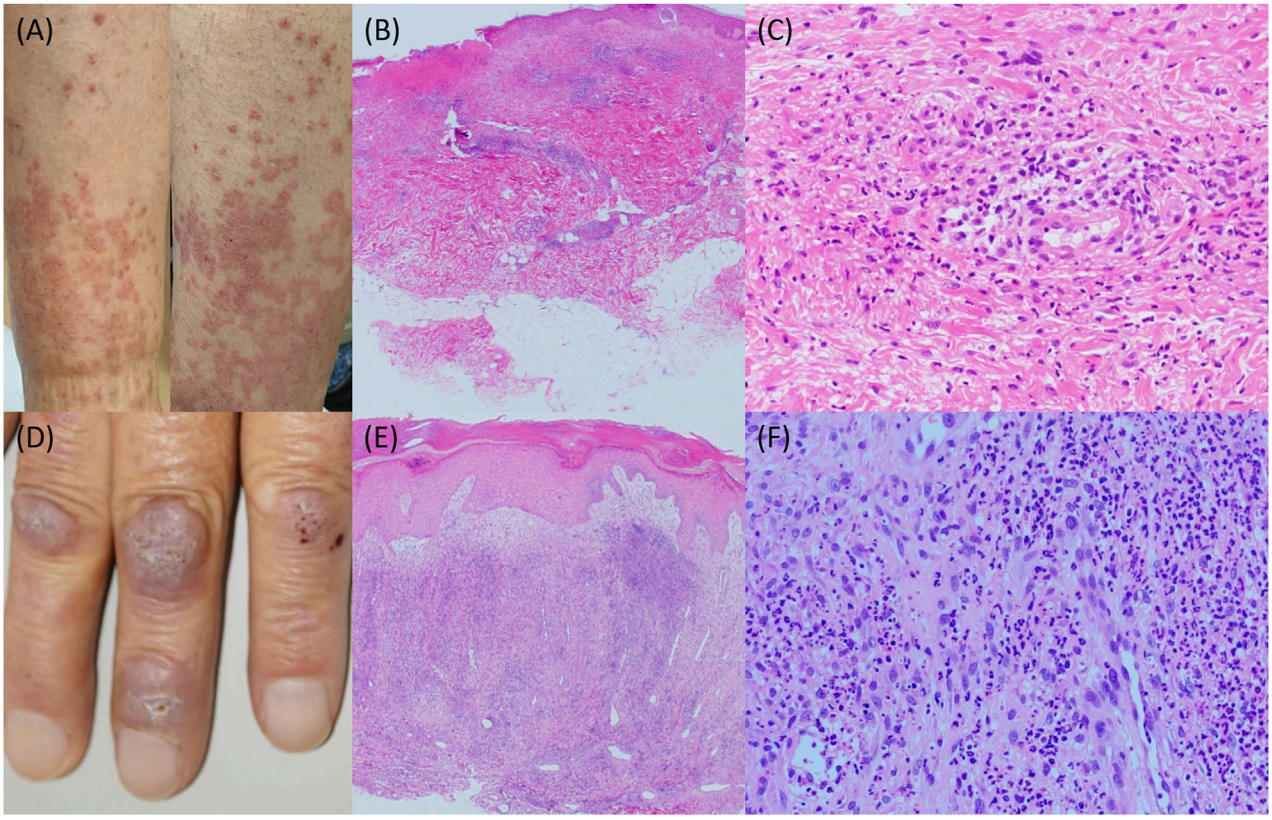


FIGURE 1
**(A)** Palpable purpura on the lower extremities. **(B)** Histopathology showing inflammatory infiltration in the upper dermis (hematoxylin and eosin staining; ×20). **(C)** Histopathology showing leukocytoclastic vasculitis (hematoxylin and eosin staining; ×200). **(D)** Crusty red-to-violet nodules on the fingers. **(E)** Histopathology showing prominent edema and dense inflammatory infiltration in the dermis (hematoxylin and eosin staining; ×20). **(F)** Histopathology showing leukocytoclastic vasculitis (hematoxylin and eosin staining; ×200).

The diagnosis of EED is made by the characteristic clinical findings and histopathological findings [2]. Although the cause of EED is not fully understood, EED has been reported in association with various systemic diseases, including hematologic diseases, infections, and autoimmune diseases [1]. It is possible that the onset of EED was associated with RA. When EED develops in patients with RA, the disease severity of EED is associated with high serum levels of rheumatoid factor [3, 4]. In our case, serum levels of rheumatoid factor were high but not elevated during the clinical course. Thus, it is speculated that the onset of EED was associated with IgA vasculitis. IgA vasculitis shares many pathogenic similarities with IgA nephropathy [5]. In our case, the disease severity of IgA nephropathy was well controlled with normal serum IgA level. However, serum IgA level had been elevated after the onset of IgA vasculitis. Aberrant IgA and IgA complexes were associated with the pathogenesis of not only IgA vasculitis but also EED. To the best of our knowledge, there have been no case reports of EED associated with IgA vasculitis. Further case accumulation is needed to clarify the underlying pathomechanisms of EED.

## Data availability statement

The original contributions presented in the study are included in the article/supplementary material, further inquiries can be directed to the corresponding author.

## Author contributions

CY and KK wrote the first draft of the manuscript. All authors contributed to the article and approved the submitted version.

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## Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

## Generative AI statement

The author(s) declare that no Generative AI was used in the creation of this manuscript.

## References

1. Newburger J, Schmieder GJ. *Erythema elevatum diutinum*. Treasure Island (FL): StatPearls (2024).

2. Gibson LE, el-Azhary RA. Erythema elevatum diutinum. *Clin Dermatol* (2000) 18(3):295–9. doi:10.1016/s0738-081x(99)00120-0

3. Bittencourt MS, Serruya T, Loureiro LO, De Souza ADC, Neri CC, Moutinho ATM, et al. Erythema elevatum diutinum in a patient with rheumatoid arthritis. *Dermatol Online J* (2023) 29(5). doi:10.5070/D329562408

4. Nakajima H, Ikeda M, Yamamoto Y, Kodama H. Erythema elevatum diutinum complicated by rheumatoid arthritis. *J Dermatol* (1999) 26(7):452–6. doi:10.1111/j.1346-8138.1999.tb02025.x

5. Song Y, Huang X, Yu G, Qiao J, Cheng J, Wu J, et al. Pathogenesis of IgA Vasculitis: An Up-To-Date Review. *Front Immunol* (2021) 12:771619. doi:10.3389/fimmu.2021.771619