

An Annular Variant of Nodular Vasculitis: A Case Report

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Received July 8, 2015; Accepted August 25, 2015; Published August 28, 2015

ABSTRACT

Panniculitis is classified histologically into distinct categories based on the location of inflammation in the subcutaneous tissue, and the presence of vasculitis. Nodular vasculitis (NV) is defined as predominantly lobular panniculitis with large vessel vasculitis. We present the case of a 73-year-old Japanese woman presenting with painful, annular, erythematous plaques on the trunk and extremities. Each erythematous plaque developed centrifugally and subsided with reticular pigmentation in a few months. Histological examination revealed lobular panniculitis and vasculitis, involving both the arteries and veins; thus, she was diagnosed with NV. The erythema continued to flare intermittently, and systemic administration of prednisolone alleviated the eruptions. We believe that the present case is unique in both annular morphology and distribution of eruptions.

Keywords: Nodular vasculitis, Annular variant, Cyclosporine

INTRODUCTION

Nodular vasculitis (NV) is defined as predominantly lobular panniculitis with large vessel vasculitis [1-4]. The indurated, painful, erythematous plaques of NV are typically found in the lower extremities. Here, we present a rare case of annularly developing erythema diagnosed histologically as NV, and the differential diagnosis

REPORT OF CASE

A 73-years old Japanese woman presented with a 10-month history of painful, annular, erythematous plaques on her trunk and extremities. Examination revealed tender, infiltrated erythema, appeared as painful infiltrated erythema and developed centrifugally and acquired an annular pattern (Figure 1A). The erythematous plaque improved as they expanded annularly without treatment for a few months (Figure 1B). On the annular erythema, we observed partial crusts but no ulcers. Inside the annular erythema, the plaques subsided with reticular pigmentation. The patient had a medical history of non-insulin-dependent diabetes mellitus, but she did not experience any fever, malaise, or arthralgia or other systemic symptom, since the eruptions had developed.

With the exception of an elevated C-reactive protein (CRP) level (4.39 mg/dL; normal range, 0–0.30 mg/dL),

results of laboratory investigations including pancreatic enzymes and liver functions, were normal. Autoantibodies, such as anti-nuclear antibody, anti-DNA antibody, proteinase 3-antineutrophil cytoplasmic antibody (PR3-ANCA), and myeloperoxidase ANCA (MPO-ANCA), were not detected.

Histological examination showed slight perivascular lymphocyte infiltration in the upper dermis, with and erythrocytes leaking through the dermis. In the fat lobules, dense infiltration consisting of lymphocytes and neutrophils was found (Figure 2A). At the center of the fat lobule, we detected vasculitis composed of neutrophils and lymphocytes, which involved both the arteries and veins.

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Citation: Higuchi T, Takano Y & Yoshida M (2015) An Annular Variant of Nodular Vasculitis: A Case Report. *Dermatol Clin Res*, 1(3).

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Non-steroidal anti-inflammatory drugs were ineffective in providing symptomatic relief, and systemic administration of prednisolone (20 mg/day) alleviated her pain, and the annular erythema flared intermittently. After discontinuing prednisolone, the erythema relapsed occasionally. During recurrence, dapsone therapy was attempted, but no effect was observed. However, cyclosporine administration helped improve the erythema.

DISCUSSION

Bazan first described erythema induratum (EI) as “erythema indurated scrofulosorum” to designate indurated erythema on the posterior lower legs [1]. Montgomery later proposed the term “nodular vasculitis” for this condition [2]. Currently, there appears to be a consensus for considering EI and NV as the same entity for the most common variant of mostly lobular panniculitis with vasculitis [3,4]. NV typically presents as purple-red erythema on the lower legs, and developed to be brownish subcutaneous nodule. Histologically, the inflammation is subcutaneous as lobular panniculitis with vasculitis [5]. There is some controversy regarding the type of vasculitis and the site of vessels in NV. In a histopathological analysis of 101 specimens with vasculitis in EI, Segura et al reported that vasculitis was evident in 91 cases [6]. They found that the vasculitis affected different vessels, such as small vessels at the center of the fat lobule as well as large septal arteries or veins. In our case, panniculitis was prominent in fat lobules and vasculitis was observed in both the arteries and veins. Therefore, we made histopathological diagnosis of NV, and ruled out polyarteritis nodosa cutanea (PNC), in which panniculitis is mostly septal and vasculitis is neutrophilic.

Our case is unique in that the patient’s erythema developed in an annular pattern and was found in atypical areas of the trunk or upper extremities. Thus, it is important to consider other differential diagnoses given the unusual presentation of NV in this case.

Erythema nodosum migrans is a subtype of erythema nodosum [7,8]. The erythema appears on lower legs, and develops centrifugally with central fading and healing and has a migratory pattern. Histological examination reveals a mostly septal panniculitis, similar to erythema nodosum, without the presence of ulcers or occurrence of subsidence after improvement. Differentiating between erythema nodosum and EI histologically can be difficult, but in our case, the panniculitis was lobular.

In lupus erythematosus profundus (LEP), the erythema is occurs in the other areas such as the lower limbs and develops into a deep subsidence of the affected area. The

panniculitis is both lobular and septal with mucin deposition, on DE-BMZ or wall vessels, deposition of immunoglobulin or complement is found [3]. In our case, there was no deposition, and autoimmunity was not detected serologically.

In antiphospholipid syndrome, reticular livedo or ulceration is found in the presence of thrombosis caused by antiphospholipid antibody or lupus anticoagulant. Histologically, thrombosis is prominent and vasculitis is generally missing through the dermis to subcutaneous tissue. In our case we observed reticular pigmentation after the improvement of erythema but not livedo, and antiphospholipid autoantibodies were not detected serologically.

The difference between PNC and the present case warrants a detailed discussion, as vasculitis is identified in subcutaneous tissue. Histological examination of PNC reveals mostly septal panniculitis and leukocytoclastic vasculitis [3]. Thus, the etiology of PNC is believed to be mainly vasculitic. In comparison, histological study of NV shows predominantly lobular panniculitis and the vasculitis is mostly granulomatous and subsequent to the affected area of panniculitis. In our case, a specimen consisting of early coin-sized erythema was found to have, septal panniculitis and no vasculitis. Therefore, we consider our case to be septal panniculitis and diagnosed the patient with NV.

Annular leukocytoclastic vasculitis (ALV) is a rare variant of leukocytoclastic vasculitis and develops annularly. Criber et al first defined ALV, reporting approximately 30 cases thus far [9-11]. The course and affected area of ALV is similar to our case. However, in ALV, dapsone should result in complete resolution of all lesions. Our case failed to respond to dapsone. Furthermore, the vasculitis in ALV is dermal leukocytoclastic. Hence, our case does not fulfill the diagnostic criteria of ALV.

Erythema elevatum diutinum is considered a chronic form of chronic leukocytoclastic vasculitis and can occasionally manifest with an annular pattern [12]. However our case did not show any fibrosis in the dermis and dapsone was not effective.

The mechanism of the annular form in NV for our case remains unknown. By reviewing the differential diagnosis, some conditions of panniculitis with or without vasculitis are suggested as an annular “variant” subtype. To the best of our knowledge, our case is the first case of an “annular variant” of NV. Furthermore in NV, plaques usually appear on the lower legs and atypical forms of NV are noted involving shins, thighs, or other sites. Our case is also unique with

regard to the affected area, such as the trunk, buttock, and thighs.

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