

# Cancer associated vasculitis

Tuba Demirci Yıldırım<sup>10</sup>, Gökçe Kenar Artın<sup>10</sup>

<sup>1</sup> Dokuz Eylül University, Department of Internal Medicine, Division of Rheumatology, İzmir, Türkiye.

Corresponding Author: Tuba Demirci Yıldırım • E-mail: tuba.demirciyildirim@deu.edu.tr This manuscript was peer-reviewed by Prof. Dr. Umut Kalyoncu

### **Definition**

Cancer-associated vasculitides are classified as 'vasculitides of possible etiologies' according to the Chapel Hill Consensus Conference [1]. All hematological and solid organ neoplasms, clonal B cell lymphoproliferative diseases, and myelodysplastic syndromes (MDS) can cause vasculitides [1]. Used in terminology with probable etiology (e.g. 'ANCA vasculitis associated with gastric cancer') [1]. Paraneoplastic vasculitis terminology is also used in many studies [2]. When the case reports were examined, the most common vasculitis-associated solid tumor was renal cell cancer. In contrast, in the retrospective study of Podjisek et al., the most common was found in lung cancer [3].

## **Clinical Features**

Cancer-associated vasculitis may be a paraneoplastic syndrome, or it may be due to infections, drugs, and cryoglobulin accumulation. In cancer-associated vasculitides, leukocytoclastic vasculitis (LCV) was the most common (45%), followed by PAN (36%) [4]. Leukocytoclastic vasculitis is a histopathological definition of diffuse small vessel vasculitis that can be found in various types of vasculitis affecting the skin and internal organs [3,4].

### Diagnosis

It is important to make a good differential diagnosis of both vasculitides and malignancy and to confirm the diagnosis with tissue biopsies [5]. When ANCA-associated vasculitides is associated with hematological malignancies, worse prognosis,

more difficult treatment, increased risk of infection, and complications are seen.

# **Differential diagnosis**

The development of vasculitis during cancer may be due to many factors, such as infections, drugs, and cryoglobulin deposition (especially for hematological malignancies). These factors may not be present in approximately 60% of patients.

## **CASE PRESENTATION**

A 44-year-old male patient with no known disease history was admitted to the rheumatology service for further examination and treatment with complaints of rash on bilateral legs, joint pain, and weight loss. He also had rash, nausea, and night sweats for 3 months. Physical examination revealed 3 cm diffuse swelling around the right neck and palpable purpura in both lower extremities (Figure 1). In the laboratory tests, hemoglobin was 11.5 gr/dl (13.5-16.5), leucocytes 8.5  $\times 10^3$  /  $\mu L$  (3.91-10.9), platelets 195 x10<sup>3</sup> / μL (166-300), blood urine nitrogen 15 mg/dl (0-20), creatinine 0.8 mg/dl (0.4-1.2), erythrocyte sedimentation rate (ESR) 54 mm/ hour (0-20), C-reactive protein (CRP) level was 105.8 mg/dl (0-5), positive anti-nuclear antibodies (ANA) (1/320 fine speckled pattern), positive proteinase 3 (pr3-ANCA), respectively. His skin biopsy was reported as LCV, and immunofluorescence examination revealed IgA, IgG, and fibrinogen deposits. Cervical lymphadenopathy with 37x27 mm size, thick cortex, heterogeneous internal structure, and peripheral vascularity was detected

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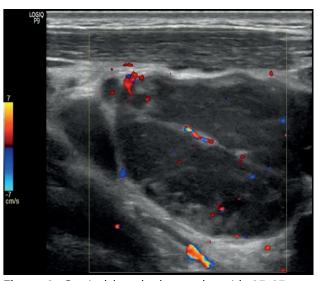
in cervical ultrasonography (Figure 2). The nasopharyngeal examination was performed, and a biopsy was performed from the lymphadenopathy present in the cervical. The biopsy examination result of cervical lymphadenopathy was reported as low-grade B-cell lymphoid neoplasia. Also, the bone marrow biopsy resulted in a transformation from chronic lymphocytic leukemia to Hodgkin lymphoma. As a result of F-18 fluorodeoxyglucose positron emission tomography (FDG- PET), the patient was diagnosed with early-stage Hodgkin lymphoma (ESHL). He was started on conventional chemotherapy for Hodgkin lymphoma.



Figure 1. Palpable purpura in both lower extremities.

### **Key messages**

- Clinicians rarely encounter malignancy in the etiology of cutaneous vasculitides.
- Whether this is an etiological cause or a paraneoplastic condition is still being discussed.
- most common underlying malignancy is usually hematological malignancy.
- The prognosis varies depending on the underlying neoplasia.



**Figure 2.** Cervical lymphadenopathy with 37x27 mm size, thick cortex, heterogeneous internal structure, and peripheral vascularity.

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