Case Report
Olgu Sunumu
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Case Report: Hydatid Disease as a Potential Cause of Leukocytoclastic Vasculitis in Endemic Regions

Olgu Sunumu: Endemik Bölgelerde Lökositoklastik Vaskülitin Potansiyel Nedeni Olarak Hidatik Hastalık

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ABSTRACT

Leukocytoclastic vasculitis (LCV) is a type of vasculitis that affects small vessels and is commonly associated with infections, malignancies, drugs, and autoimmune diseases. In this case, a 75-year-old female patient presented with clinical signs of LCV, and after ruling out common etiologies, hydatid disease (HD) emerged as a potential cause. This case highlights the importance of considering parasitic infections, particularly HD, in the differential diagnosis of LCV, especially in regions where these infections are endemic and in patients exposed to relevant environmental risk factors.
Keywords: Hydatid cyst, leukocytoclastic vasculitis, parasitic infections

ÖZ

Leukositoklastik vaskulitis (LCV), küçük damarları etkileyen ve genellikle enfeksiyonlar, maligniteler, ilaçlar ve otoimmün hastalıklarla ilişkilendirilen bir vaskulit türüdür. Bu vakada, LCV kliniği ile başvuran 75 yaşındaki kadın hastada, yaygın etiyolojik faktörler dışlandıktan sonra kist hidatik hastalığı olası bir neden olarak ortaya çıkmıştır. Bu vaka, LCV'nin ayırıcı tanısında, özellikle bu enfeksiyonların endemik olduğu bölgelerde ve çevresel risk faktörlerine maruz kalan hastalarda, paraziter enfeksiyonların, özellikle kist hidatik hastalığının göz önünde bulundurulmasının önemini vurgulamaktadır.
Anahtar Kelimeler: Kist hidatik, leukositoklastik vaskülitis, paraziter enfeksiyonlar

INTRODUCTION

Leukocytoclastic vasculitis (LCV) is a form of small-vessel vasculitis commonly manifested by palpable purpura, especially in the lower extremities. It has a broad differential diagnosis, with known associations to autoimmune diseases, infections, malignancies, and medications (1,2). Hydatid disease (HD), caused by *Echinococcus* tapeworms, is a zoonotic infection primarily affecting the liver and prevalent in certain endemic regions. While the link between HD and LCV has been scarcely documented (3), this case suggests the need to consider HD as a potential etiological factor for LCV, especially in regions where both conditions are endemic.

CASE REPORT

A 75-year-old woman presented with a 15-day history of palpable purpura on both lower extremities (Figure 1). She reported a similar episode four years ago, which resolved spontaneously. Her medical history was unremarkable, and she was not on any medications. The patient had a history of livestock farming and owned cats and dogs. No recent infections or significant health events were reported. Physical examination revealed palpable purpura on the lower extremities but no other significant findings. A skin biopsy confirmed the diagnosis of LCV, prompting further investigation of its underlying cause.

Autoimmune screening showed a weakly positive antinuclear antibody at a titer of 1:100 and positive anti-Ku antibodies. However, tests for rheumatoid factor, anti-cyclic citrullinated peptide, anti-double-

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stranded DNA (anti-dsDNA), and antineutrophil cytoplasmic antibodies (ANCA) were all negative. Urinalysis was normal, and C-reactive protein was elevated at 31.1 mg/L. These findings did not support a diagnosis of any specific connective tissue disorder.

Further investigations to identify potential infectious causes, including hepatitis, tuberculosis, and *Brucella*, were negative. A malignancy workup, including tumor markers, fecal occult blood test, mammogram, and imaging studies [computed tomography (CT) of the neck, pelvis, and thorax], also yielded negative results. However, an abdominal CT scan revealed a type 5 hydatid cyst in segments 7 and 8 of the liver (Figure 2). Indirect hemagglutination testing for echinococcal infection was positive with a titer of 1/2560. The patient was diagnosed with hydatid cyst disease, and consultations with infectious diseases and general surgery confirmed that no immediate treatment was necessary.

Given the patient's environmental exposure to livestock and pets, along with the absence of other underlying conditions and negative infectious, malignancy, and autoimmune screening, HD was considered a likely cause of her LCV. The patient was started on a short-term course of prednisone, resulting in significant improvement in the purpura.

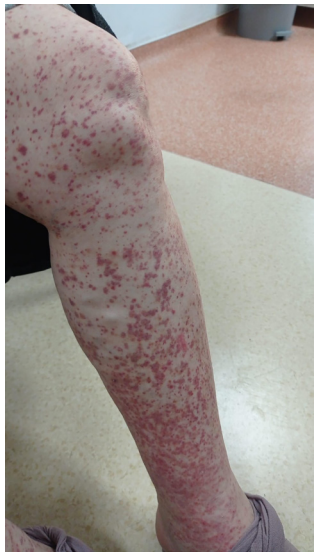

Figure 1. Palpable purpura on the left lower extremity

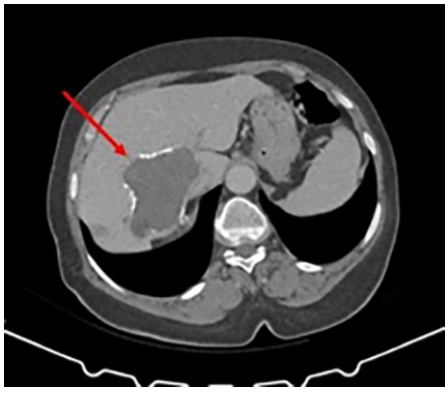

Figure 2. Abdominal CT scan showing a type 5 hydatid cyst measuring 79x73 mm, located in liver segments 7-8
CT: Computed tomography

DISCUSSION

LCV is a common type of cutaneous small vessel vasculitis (CSVV), manifested by palpable purpura, especially in the lower extremities (4). According to the nomenclature of vasculitis provided by the 2012 Revised International Chapel Hill Consensus Conference, CSVV is categorized as a single-organ vasculitis (5). The etiology of LCV is multifactorial, with several potential underlying causes (1). It can be seen in immune-mediated vasculitis (e.g., ANCA-associated vasculitis, Henoch-Schönlein purpura) (2). Additionally, LCV may arise as a complication of connective tissue disorders such as rheumatoid arthritis and systemic lupus erythematosus, Sjögren syndrome (6). It is also associated with malignancies and can be triggered by infectious agents (6). Finally, various medications (7), vaccines (8), and other factors have been recognized as potential contributors to the development of LCV.

HD remains a significant health issue in endemic regions, transmitted through the fecal-oral route, often from dogs or other canids (9). Although the liver is the most frequently affected organ, other organs such as the heart, lungs, spleen and brain, may also be involved (10,11). In many cases, hydatid cysts remain asymptomatic and are incidentally discovered during imaging studies, as seen in our patient (11,12).

Parasitic infections can act as environmental triggers for autoimmune diseases by disrupting immune balance. A striking example is a reported case of polymyalgia rheumatica and giant cell arteritis occurring alongside cystic echinococcosis, suggesting a potential causal link (13). Similarly, LCV may also emerge in the context of parasitic infections, though this association is extremely rare. In the literature, only one case report describes a 10-year-old patient who presented with LCV and necrotizing pneumonia, and was ultimately diagnosed with pulmonary HD (3). The report emphasizes that echinococcal infection should not be overlooked as a potential cause of LCV (3). In light of our patient's exposure to livestock and pets, the presence of a cyst on abdominal CT, serologic evaulation, and the negative findings from autoimmune, malignancy, and other infectious screenings, HD was considered a contributing factor to the development of LCV. Our case is notable as the second reported instance of LCV associated with HD, this time involving the liver.

Although direct evidence linking cystic HD with LCV remains limited, molecular interactions between the human host and the parasite suggest a possible connection. During the early stages of hydatid cyst formation, a cell-mediated immune response involving macrophages, neutrophils, and eosinophils is triggered. *Echinococcus granulosus* stimulates both TH1 and TH2 immune responses (14-16). After treatment or the natural death of the cyst, the TH2 response decreases rapidly, and the TH1 response becomes predominant (12,17). This immune reaction can persist even in the inactive stage, especially in individuals with a history of previous active infection. Additionally, residual immune complexes from prior infections may remain in circulation, potentially sensitizing the immune system and causing it to react to the cyst, even without active parasitic involvement (18).

CONCLUSION

This case emphasizes the importance of considering parasitic infections such as HD in the differential diagnosis of LCV, especially in endemic regions. While the direct link between

cystic HD and LCV remains under study, this case contributes to the growing body of evidence supporting HD as a potential cause of vasculitis. Further research is needed to better understand the underlying mechanisms.

*Ethics

Informed Consent: Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Footnotes

*Authorship Contributions

Concept: G.K., F.N.A.B., Z.B.K., Design: G.K., F.N.A.B., Z.B.K., Data Collection or Processing: G.K., F.N.A.B., Z.B.K., Analysis or Interpretation: G.K., F.N.A.B., Z.B.K., Literature Search: G.K., F.N.A.B., Z.B.K.,

Writing: G.K.

Conflict of Interest: No conflict of interest was declared by the authors.

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REFERENCES

1. Fraticelli P, Benfaremo D, Gabrielli A. Diagnosis and management of leukocytoclastic vasculitis. Intern Emerg Med. 2021; 16: 831-41.
2. Khetan P, Sethuraman G, Khaitan BK, Sharma VK, Gupta R, Dinda AK, et al. An aetiological & clinicopathological study on cutaneous vasculitis. Indian J Med Res. 2012; 135: 107-13.
3. Isik S, Sozmen SC, Guzeloglu E, Ozturk T, Asilsoy S. Pulmonary hydatid cyst disease mimicking necrotizing pneumonia in a child with leukocytoclastic vasculitis. Turk Pediatri Ars. 2018; 53: 117-9.
4. Caproni M, Verdelli A. An update on the nomenclature for cutaneous vasculitis. Curr Opin Rheumatol. 2019; 31: 46-52.
5. Sunderkotter CH, Zelger B, Chen KR, Requena L, Piette W, Carlson JA, et al. Nomenclature of cutaneous vasculitis: dermatologic addendum to the 2012 Revised International Chapel Hill Consensus Conference nomenclature of vasculitides. Arthritis Rheumatol. 2018; 70: 171-84.
6. Alpsoy E. Cutaneous vasculitis; an algorithmic approach to diagnosis. Front Med (Lausanne). 2022; 9: 1012554.
7. Mullick FG, McAllister HA, Wagner BM, Fenoglio JJ. Drug related vasculitis. Clinicopathologic correlations in 30 patients. Hum Pathol. 1979; 10: 313-25.
8. Corra A, Verdelli A, Mariotti EB, Ruffo di Calabria V, Quintarelli L, Aimo C, et al. Cutaneous vasculitis: lessons from COVID-19 and COVID-19 vaccination. Front Med (Lausanne). 2022; 9: 1013846.
9. Wen H, Vuitton L, Tuxun T, Li J, Vuitton DA, Zhang W, et al. Echinococcosis: advances in the 21st century. Clin Microbiol Rev. 2019; 32: e00075-18.
10. Sahin S, Kaya B. Evaluation of Hydatid Cyst Cases: A Single-center retrospective study. Turkiye Parazitol Derg. 2025;48: 222-7.
11. Govindasamy A, Bhattarai PR, John J. Liver cystic echinococcosis: a parasitic review. Ther Adv Infect Dis. 2023; 10: 20499361231171478.
12. Nunnari G, Pinzone MR, Gruttadauria S, Celesia BM, Madeddu G, Malaguarnera G, et al. Hepatic echinococcosis: clinical and therapeutic aspects. World J Gastroenterol. 2012; 18: 1448-58.
13. Bektan Kanat B, Ulugerger Avci G, Icli TB, Doventas A. Concomitant polymyalgia rheumatica and giant Cell Arteritis Associated with Cystic echinococcosis: a rare geriatric case. Acta Parasitol. 2024; 69: 1085-9.
14. Rigano R, Profumo E, Siracusano A. New perspectives in the immunology of *Echinococcus granulosus* infection. Parassitologia. 1997; 39: 275-7.
15. Rigano R, Profumo E, Teggi A, Siracusano A. Production of IL-5 and IL-6 by peripheral blood mononuclear cells (PBMC) from patients with *Echinococcus granulosus* infection. Clin Exp Immunol. 1996; 105: 456-9.
16. Zhang W, Li J, McManus DP. Concepts in immunology and diagnosis of hydatid disease. Clin Microbiol Rev. 2003; 16: 18-36.
17. Rigano R, Profumo E, Ioppolo S, Notargiacomo S, Ortona E, Teggi A, et al. Immunological markers indicating the effectiveness of pharmacological treatment in human hydatid disease. Clin Exp Immunol. 1995; 102: 281-5.
18. Aimulajiang K, Guo B. Echinococcosis immune response, immunopathogenesis and immune evasion from the human host. Li J, Wang W, Mehlhorn H, (editors). Echinococcus: control and elimination of echinococcosis with a focus on China and Europe. Springer; 2024. p.109-128.