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LETTER TO THE EDITOR EDİTÖRE MEKTUP

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Kaposi Sarcoma: Another Cause of Secondary Lymphedema

Kaposi Sarkomu: Sekonder Lenfödemin Diğer Bir Nedeni

Selda ÇİFTCİ İNCEOĞLU[a], Aylin AYYILDIZ[b], Banu KURAN[a]

[a]Health Sciences University, Faculty of Medicine, Şişli Hamidiye Etfal Training and Research Hospital, Department of Physical Medicine and Rehabilitation, İstanbul, Türkiye
[b]İstanbul Başakşehir Çam and Sakura City Hospital, Clinic of Physical Medicine and Rehabilitation, İstanbul, Türkiye

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Dear Editor,

Lymphedema due to Kaposi sarcoma (KS) is a clinical condition that should be included in the differential diagnosis of secondary lymphedema. A case referred to our clinic from dermatology due to lymphedema and previously evaluated for KS due to skin lesions drew our attention to this issue. There is limited information about lymphedema in publications on KS from our country. Therefore, we prepared this letter to emphasize that Kaposi sarcoma should be included in the differential diagnosis of secondary lymphedema and to share this perspective with our colleagues.

KS is a sarcoma caused by Human Herpes Virus-8 (HHV-8), and can present with various subtypes, some of which may be associated with lymphedema. It is classified into 4 groups: classic, endemic, epidemic (associated with Human Immunodeficiency Virus) and iatrogenic type (typically occurring after transplantation).[1] HHV-8 has a tropism for lymphatic endothelial vessels, causing multicentric lymphatic endothelial proliferation and obstruction.[2] Furthermore, the progression of lesions may lead to external obstruction of lymphatic vessels. Lymphedema resulting from lymphatic vascular obstruction usually affects the lower extremities and may develop before, simultaneously with, or after the skin lesion of KS.[1]

Skin lesions on the extremities with lymphedema are crucial in the differential diagnosis. Skin lesions of KS usually appear as purple or brown macules, papules, or hyperkeratotic plaques. They are usually seen on the lower extremities and especially on the feet.[1] The sudden appearance of purplish lesions on a lymphedematous extremity may indicate angiosarcoma, which is a rare but highly aggressive malignancy.[3] In addition to angiosarcoma, melanoma, KS, basal cell carcinoma, or squamous cell carcinoma may develop in the chronic

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Correspondence: Selda ÇİFTCİ İNCEOĞLU
Health Sciences University, Faculty of Medicine, Şişli Hamidiye Etfal Training and Research Hospital, Department of Physical Medicine and Rehabilitation, İstanbul, Türkiye
E-mail: seldavd@gmail.com

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lymphedematous extremity.[4] In other words, KS can be both a cause and a consequence of lymphedema. Telangiectasis, hyperpigmentation, lipodermatosclerosis, and ulcers may also develop due to chronic venous insufficiency and may present as phlebolymphedema.[5] A skin biopsy is essential for diagnosis, as it directly influences the treatment approach. While complete decongestive therapy (CDT) is applied in cases of lymphedema secondary to KS or chronic venous insufficiency, CDT is not recommended for Stewart-Traves Syndrome which requires surgical treatment.[1,3,5]

In our case, histopathological examination did not confirm KS; instead, the patient was diagnosed with secondary lymphedema due to a history of trauma and recurrent infections. CDT was initiated, resulting in improved peripheral measurements and symptomatic relief. The patient is currently being followed up with a compression garment. We believe that the experience gained from this KS-related lymphedema case will contribute to our colleagues' understanding of the differential diagnosis in similar cases.

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Authorship Contributions

All authors contributed equally while this study preparing.

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