Case Report
Olgu Sunumu

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Unilateral orbital eosinophilic cellulitis (Wells syndrome)

Unilateral orbital eozinofilik selülit (Wells sendromu)

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Abstract

Eosinophilic cellulitis, also known as Wells syndrome, is a rare inflammatory skin disorder of unknown etiology. The disease has no known racial or sexual predisposition. It usually affects adults, although it can also be observed in children. Typically, it begins as a solitary burning or pruritic erythematous, edematous, and urticarial plaque, which usually has an acute cellulitis-like appearance. Nodules, blisters, or bullae may develop later in the initial lesion. The disease follows a pattern of spontaneous relapse and remission. The extremities and trunk are the most commonly involved localizations, but only a few cases of solely orbital involvement have been reported. Herein, we present a case of unilateral orbital eosinophilic cellulitis that remained undiagnosed for a long period.
Keywords: Eosinophilic, cellulitis, ptosis, Wells

Öz

Wells sendromu olarak da bilinen eozinofilik selülit, etiyolojisi bilinmeyen ve nadir görülen enflamatuvar bir deri hastalığıdır. Hastalığın bilinen bir ırk ve cinsiyet tercihi yoktur. Genellikle yetişkinleri etkilemekle birlikte çocuklarda da görülebilmektedir. Tipik olarak soliter, yanan veya kaşınan, eritematöz, ödemli ürtikeryal bir plak olarak başlar ve genellikle akut selülit benzeri bir görünüme sahiptir. Başlangıçtaki lezyonun ilerleyen dönemlerinde nodüller, vezikül veya büller ortaya çıkabilir. Hastalığın spontan nüks ve remisyonlardan oluşan bir klinik seyri vardır. En sık ekstremite ve gövde tutulumu görülür ve bugüne kadar oldukça az sayıda orbita tutulumlu olgu rapor edilmiştir. Burada uzun süredir tanı konulamayan tek taraflı orbita tutulumu ile seyreden bir eozinofilik selülit olgusunu sunuyoruz.
Anahtar Kelimeler: Eosinofilik, selülit, pitozis, Wells

Introduction

Eosinophilic cellulitis, also known as Wells syndrome (WS), is a rare inflammatory skin disorder of unknown etiology. It was first described by Wells in 1971 as a "recurrent granulomatous dermatitis with eosinophilia." In 1979, "eosinophilic cellulitis" was proposed by Wells and Smith[1]. The disease has no known racial or sexual predisposition. It usually affects adults, although it can also be observed in children. Typically, it begins as a solitary burning or pruritic

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erythematous, edematous, and urticarial plaque, which usually has an acute cellulitis-like appearance. Nodules, blisters, or bullae may develop later in the initial lesion. This then becomes the second stage, with progressive involution occurring over a period of two to eight weeks. Morphea-like residual skin atrophy and hyperpigmentation may be endpoint[2]. The initial plaque is often unresponsive to antimicrobial treatment and regresses spontaneously within weeks. The disease exhibits a pattern of spontaneous relapses and remissions[3]. The extremities and trunk are the most commonly involved localizations, but only a few cases of solely orbital involvement have been reported. Herein, we present a case of unilateral orbital eosinophilic cellulitis that remained undiagnosed for a long period.

Case Report

A seventy-year-old woman presented with relapsing and remitting swelling of the right upper and lower eyelids that had started four months earlier. She had been diagnosed with preseptal cellulitis and had received antibiotics. Several consultations with ophthalmology, infectious diseases, and rheumatology outpatient clinics preceded her referral. The lesion did not improve with antibiotic or antihistamine treatment. She was asked about comorbidities and the use of medications or herbal ingredients that could cause this condition. It was learned that she did not use anything else apart from the oral antihistamine used for chronic allergic rhinitis.

Dermatological examination revealed solid edema, erythema, and ptosis in the right periorbital region (Figure 1). Although the patient did not respond to systemic antibiotics, preseptal cellulitis and granulomatous diseases, such as sarcoidosis, cutaneous tuberculosis, and atypical mycobacterial infections, were established as differential diagnoses. Contact dermatitis was deemed unlikely because of the unilateral presentation and granulomatous nature of the lesion, as well as the absence of any known ocular exposure. The healthcare team requested a complete blood count, kidney and liver function tests, IgG, IgE, angiotensin-converting enzyme (ACE) level, sedimentation, C-reactive protein (CRP), and QuantiFERON tests. The autoantibody and QuantiFERON test results were negative. Thoracic computed tomography revealed no hilar or mediastinal lymphadenopathy. The parenchymal tissue was also clear. Therefore, cutaneous tuberculosis and sarcoidosis were excluded. She also had negative blood culture results from her previous infectious disease consultations. The ACE level was 71 U/L (8-52), the sedimentation rate was 42 mm/hr (1-15), and the CRP level was 25 mg/L (0-5). No eosinophilia was detected in the hemogram. Magnetic resonance imaging revealed no mass or tumor in the retro-orbital region; however, swelling and contrast enhancement of the right preseptal region and lacrimal gland were observed. Therefore, a biopsy was performed on the right palpebra and lacrimal glands. Histopathological examination of the palpebral sample revealed severe eosinophil-rich inflammation around the vessels, occasionally infiltrating the veins, and fibrin in a few vessel walls. In the right lacrimal gland, we observed fibroadipose tissue with eosinophil-rich inflammatory infiltrate that caused shedding of the endothelium of the vessel walls (Figure 2). She was diagnosed with eosinophilic cellulitis based on clinical and histopathological findings. The patient was evaluated for potential underlying systemic conditions and malignancies. No associated diseases were observed. Systemic methylprednisolone treatment was initiated at a dose of 0.5 mg/kg, and a dramatic response was observed (Figure 3). Once the disease was controlled, the methylprednisolone dosage was tapered to 2 mg/day. However, attempts to further reduce the dosage to 2 mg every other day resulted in relapse. The patient was under our care for 14 months, and the disease remained well-controlled on a maintenance dose of 2 mg/day methylprednisolone for the past eight months. Regular monitoring of the bone mineral density revealed no significant changes. Aside from a 2 kg weight gain, no other corticosteroid-related adverse effects were observed.

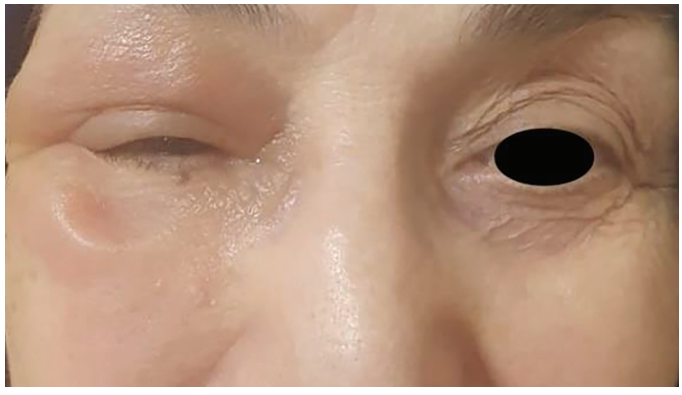

Figure 1. Solid edema, erythema, and ptosis in the right periorbital region

Discussion

Eosinophilic cellulitis is a rare inflammatory dermatosis with an unclear etiology. Many cases appear idiopathic, whereas others suggest a triggering event or may be associated with an underlying disease. These underlying disorders include hematological and non-hematological malignancies such as renal cell carcinoma, colon carcinoma, and nasopharyngeal carcinoma. Churg-Strauss syndrome, ulcerative colitis, and hypereosinophilic syndrome have also been associated with eosinophilic cellulitis[4].

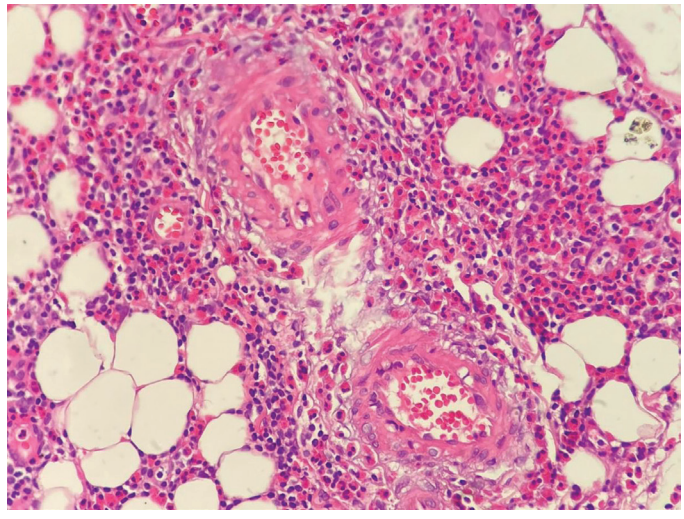

Figure 2. Mass-forming infiltration consisting of eosinophils that concentrate around the vessels and infiltrate the vessel wall (x400 HE)
HE: Hematoxylin-eozin

The pathogenesis of WS is unknown. Mitchell et al.[5] hypothesized that allergic hypersensitivity may be involved in the pathogenesis. Urticaria, peripheral eosinophilia, and triggers such as drugs and insect bites indicate a possible allergic phenomenon[6]. España et al.[7] presented a patient with WS in whom they found a close correlation between clinical activity, eosinophils in the blood and bone marrow, eosinophil cation protein, and interleukin (IL)-5 levels in the peripheral blood and tissues. They highlighted the crucial role of IL-5.

WS is diagnosed based on a combination of clinical, laboratory, and histopathological findings. Approximately 50% of patients experience peripheral blood eosinophilia during active disease[3]. Histopathological findings vary depending on lesion age. In the acute phase, dermal edema and eosinophil-predominant inflammatory infiltrates were observed in the dermis of the skin. Flame figures appear in the subacute phase when degranulated eosinophils, leukocytes, and dermal histiocytes surround the dermal collagen bundles. During the resolution stage, eosinophils disappear, revealing palisaded phagocytic histiocytes surrounding the flame figures. Although flame figures are very supportive of the diagnosis, they are not pathognomonic and can also be observed in other dermatoses with hypereosinophilia[8].

Heelan et al.[9] proposed a diagnostic criterion for WS, although it requires validation in larger patient groups. According to this algorithm, two major and one minor criteria are required for diagnosis. The major criteria consist of typical clinical presentation, including any of the previously reported variants (plaque-type, annular-granuloma-like, urticaria-like, papulovesicular, bullous, papulonodular, fixed-drug eruption-like), relapsing and remitting course, exclusion of systemic disease, and typical histology of eosinophilic infiltrates without signs of vasculitis. Minor criteria include flame figures, granulomatous changes in histology, peripheral eosinophilia not persistent and not greater than 1500/μL, and detectable triggering factors.

WS can manifest on any skin surface and presents with various clinical symptoms. While lesions are typically found on the extremities, facial involvement is uncommon. Two cases of WS with ipsilateral eye involvement have been reported in the literature: an 81-year-old woman with ipsilateral erythematous eye edema[10] and a 56-year-old woman with left eyelid swelling and ptosis[11]. Similar to our experience, these cases were initially misdiagnosed as infectious diseases. However, the lack of response to treatment and recurring symptoms prompted biopsies, which ultimately confirmed the diagnosis of WS.

If a triggering factor that causes the disease could be detected, then treating this condition would also treat WS. If no precipitating factors are identified, the first-line treatment is topical/systemic corticosteroids. Most patients respond rapidly to systemic corticosteroid therapy. The highest rate of resolution with oral steroids was approximately 2 mg/kg/day for 1-2 weeks, followed by tapering over 2-3 weeks. If there is no response to steroid treatment or if there is a contraindication for steroids, dapsone or cyclosporine may be the treatment of choice[12].

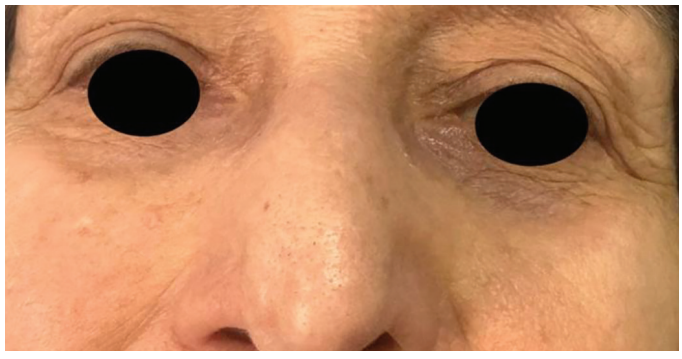

Figure 3. Improved condition of the patient after treatment

In our case, histopathology led us to a diagnosis of hypereosinophilic dermatoses. The patient's long-standing complaints and the absence of drug use could have caused the lesion and helped rule out a fixed drug eruption. The absence of systemic findings, lack of peripheral eosinophilia, and negative results for autoantibodies that did not meet the conditions for vasculitis ultimately ruled out both Churg-Strauss syndrome and hypereosinophilic syndrome.

In conclusion, establishing a diagnosis of WS requires exclusion of other diseases, multidisciplinary work, and a high degree of suspicion for WS. As seen in the current case, it may not always be present in typical locations, may mimic other diseases, and there may be no eosinophilia in the peripheral blood or flame figures in histopathology. When a patient presents with unilateral orbital edema, it may be helpful to consider WS, as with other differential diagnoses. A timely diagnosis can prevent potential complications, such as increased intraocular pressure and orbital compartment syndrome, which may lead to permanent vision loss. Additionally, owing to their relationship with malignancies, patients should be followed up for a long duration.

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: İ.N.D.T., Ç.T., Desing: İ.N.D.T., Ç.T., Data Collection or Processing: İ.N.D.T., Ç.T., Ç.Ö., Y.S., Ö.E., Analysis os Interpretation: İ.N.D.T., Ç.T., Literature Search: İ.N.D.T., Ç.T., Writing: İ.N.D.T.

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

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