Conjunctival Granulomatosis in Churg-Strauss Syndrome

Churg-Strauss syndrome (CSS) is a rare, potentially lethal systemic vasculitis characterized by necrotizing arteritis, eosinophil infiltration of the tissues, and extravascular granulomas. Ocular involvement includes orbital inflammation, scleritis or episcleritis, peripheral ulcerative keratitis, uveitis, anterior ischemic optic neuropathy, retinal artery and vein occlusions, choroidal ischemia, and oculomotor nerve palsies. Conjunctival involvement is rare and can present as nonspecific conjunctivitis, inflammatory thickening of the conjunctiva with or without amyloidosis, or conjunctival nodule with inflammation. We describe a patient with clinically inactive CSS who developed multiple bilateral conjunctival nodules.

Report of a Case. In June 2011, a 38-year-old woman had ocular irritation and foreign body sensation in both eyes (OD > OS) for 2 weeks. She denied ocular pain, diplopia, or a change in visual acuity. Since December 2010, she had attacks of asthma, recurrent sinusitis, episodes of upper gastric pain, and skin rash. A thorough workup revealed granulomas of the lung associated with tissue eosinophilia, peripheral eosinophilia, increased total IgE level, and positive findings for perinuclear antineutrophil cytoplasmic antibodies. Thus, the patient fulfilled

Figure 1. Yellowish nodular lesions in the bulbar conjunctiva of the right eye of a 38-year-old woman with treated Churg-Strauss syndrome.

Figure 2. Histologic findings. A, Conjunctival granuloma formation involving lymphocytes, macrophages, eosinophils, and a central necrosis (hematoxylin-eosin, original magnification ×10). B, Immunohistochemistry confirmed the presence of CD3⁺ T lymphocytes, CD68⁺ macrophages (not shown), and CD20⁺ B lymphocytes (not shown) (original magnification ×40). D, The conjunctival stroma also showed pronounced occlusive perivasculitis (asterisks), with the perivascular infiltrate mainly confined to arteries and consisting of CD3⁺ T lymphocytes, CD20⁺ B lymphocytes, histiocytes, and giant cells (immunohistochemistry, original magnification ×40).
granulomas composed of CD3+ T lymphocytes, CD20+ B lymphocytes, CD68+ macrophages, histiocytes, giant cells, and multiple eosinophils surrounding a center of tissue necrosis (Figure 2). The inflammation was particularly intense around vessels, especially arteries, and associated with vascular occlusion. The perivascular infiltrate mainly consisted of CD3+ T lymphocytes and CD20+ B lymphocytes as well as histiocytes and giant cells (Figure 2).

Gomori methenamine silver, periodic acid–Schiff, and Ziehl-Neelsen stains excluded fungi and mycobacteria. Histopathologic analysis of the conjunctival nodules confirmed the presence of multiple eosinophilic perivascular granulomas consistent with CSS.

Comment. Our patient with systemically inactive CSS developed multiple conjunctival eosinophilic granulomas. This clinical manifestation is extremely rare and, to our knowledge, has never been described in this form. Shields et al1 treated a 64-year-old man with CSS and progressive inflammatory thickening of the bulb and palpebral conjunctiva in 1 eye that resolved with systemic corticosteroids and cyclophosphamide. Margolis et al2 described a 30-year-old woman with CSS who had 1 well-demarcated conjunctival nodule with mild focal inflammation. Histologic analysis of both cases confirmed the presence of necrotizing eosinophilic granuloma, but a pathognomonic vasculitis was not observed.2,3 The findings in our patient with CSS are also distinct from conjunctival involvement in Wegener granulomatosis, where inflammation is mainly confined to the palpebral conjunctiva and tarsus followed by fibrovascular proliferation and scarring.6 Our case is special in some respects: multiple granulomas developed bilaterally in the clinically completely uninflamed conjunctiva of a patient with CSS under apparently successful systemic immunosuppressive therapy. Moreover, the excised conjunctival tissue showed a clear-cut vasculitis, which, according to Takanashi et al,2 is quite uncommon for the orbital inflammatory pseudotumor type of CSS where conjunctival lesions are included in contrast to the ischemic type. Signs of eosinophilic tissue infiltration and granuloma formation only rarely coincide with signs of vasculitis in ocular CSS.7 Thus, the development of conjunctival granulomas in successfully treated CSS and the demonstration of vasculitis on biopsy may be early signs of activation and should prompt a careful clinical follow-up.

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COMMENTS AND OPINIONS

Angle Involvement and Glaucoma in Patients With Biopsy-Proven Iris Melanoma: A Response

Khan et al4 reported the results of a multicenter international study to identify representative epidemiological, clinical, and pathological characteristics of melanoma of the iris. Their conclusion states, “This multicenter, Internet-based, international study successfully pooled data and extracted information on biopsy-proven melanoma of the iris.”1

In the third paragraph of their article, Khan et al reference 9 “reports of clinical and histopathologic prognostics and outcomes for treatment,”1 one of which was published in 1981 by Frederick A. Jakobiec, MD, DSc, and myself, entitled “Are Most Iris ‘Melanomas’ Really Nevi? A Clinicopathologic Study of 189 Lesions.”2 Khan et al state, “However, these studies had several limitations. First, they included many patients whose tumor diagnosis was not histologically confirmed.”1 I would like to point out that our study was a retrospective clino-