Eosinophilic Cellulitis (Wells Syndrome) in a Pediatric Patient: A Case Report and Review of the Literature

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We report a 4-year-old boy who presented with multiple pruritic, annular, erythematous plaques on the lower extremities of 1 week’s duration. Histopathology of an affected area revealed a dense dermal infiltrate of eosinophils and flame figures without evidence of vasculitis. A diagnosis of eosinophilic cellulitis (EC), or Wells syndrome, was made. The patient had an excellent response to topical and systemic steroids following 1 week of treatment. This case appeared to be idiopathic, as there was no cause identified such as arthropod bites or tinea infection. The patient’s EC cleared and has not had a recurrence.

Eosinophilic cellulitis (EC), or Wells syndrome, is a cutaneous disease that typically presents abruptly with pruritic, erythematous, and edematous plaques that resemble urticaria or cellulitis. The lesions often resemble bacterial cellulitis but notably are cool to the touch. Both sexes are equally affected. Although it is more commonly seen in adults, pediatric cases have been reported in the literature. Eosinophilic cellulitis is thought to be precipitated by a number of triggering factors including drugs, arthropod bites, and viral infections.

Eosinophilic cellulitis has been described as typically having 2 phases. The first phase begins with prodromal symptoms of pain and burning followed by the development of plaques that expand centrifugally and may be associated with systemic symptoms.

The lesions then evolve into the second stage in 1 to 3 weeks with induration of the plaques. The lesions typically resolve in 4 to 8 weeks but often recur, occasionally leaving postinflammatory pigment changes. Peripheral blood eosinophilia is present in approximately 50% of cases.

Pathology of the involved lesions typically reveals a dermal infiltrate of eosinophils and histiocytes with scattered flame figures. Older lesions may contain multinucleated giant cells.

We describe a case of EC with no identifiable cause in a pediatric patient.

Case Report

A 4-year-old boy presented with pruritic plaques on the lower extremities of 1 week’s duration (Figure 1). The patient’s parent denied recent animal or plant contact, arthropod bites, or trauma.

Medical history was notable for visual impairment and mental retardation and negative for atopy. The patient was not taking any daily medications. Family history was unremarkable.

Physical examination revealed edematous, annular, erythematous plaques with pseudovesiculation on the lower extremities. A potassium hydroxide preparation from a lesion was negative. The remainder of the physical examination was unremarkable. The vital signs did not reveal any fever. A fungal culture was sent and a punch biopsy was performed. The patient was started on fluocinonide ointment 0.05% and ketoconazole cream. A complete blood cell count was within reference range.

Punch biopsy of a plaque on the lower extremity revealed a dense dermal infiltrate of eosinophils, histiocytes, and lymphocytes. Flame figures were present without evidence of vasculitis (Figure 2). Periodic acid–Schiff and Gomori methenamine-silver stains were negative for fungal organisms.

These findings established the diagnosis of EC and the patient was started on prednisolone syrup.
15 mg (1 mg/kg) daily. Fluocinonide ointment was continued and the ketoconazole cream was stopped. A follow-up visit after 1 week of treatment demonstrated marked improvement. Physical examination revealed hyperpigmented patches, smaller plaques on the lower extremities, and no new lesions. To date (2 years later), the patient has not had any recurrence.

**Comment**

Eosinophilic cellulitis is a rare condition with few cases reported in the literature. It was first described by Wells\(^1\) in 1971 in 4 patients who presented with recurrent localized swelling of the extremities characterized by acute EC in early lesions followed by granulomatous dermatitis.

The condition is hypothesized to be caused by a localized hypersensitivity reaction triggered by various stimuli that lead to eosinophil-induced cytotoxicity. Some of the reported triggering events that have occurred in children are arthropod bites\(^4,6\); bee stings\(^7\); thimerosal-containing vaccines\(^8,9\); and infections including varicella,\(^10\) molluscum contagiosum,\(^11,12\) Parvovirus B19,\(^13\) mumps, and erysipelas (treated with penicillin).\(^1\) Reported triggers in adults include adalimumab,\(^14\) arthropod\(^15,16\) and spider bites,\(^17\) parasitic infections such as toxocariasis\(^1\) and onchocerciasis,\(^18,19\) Churg-Strauss syndrome,\(^20\) and malignancy.\(^21\) Two congenital cases have been described in the literature, both thought to be caused by maternal exposure to danazol during pregnancy.\(^22,23\) An unknown genetic trigger also has been proposed with the report of 2 male siblings and their mother all having been diagnosed with EC.\(^2\) Furthermore, a dominant syndrome in a family presenting with EC, mental retardation, and abnormal body habitus has been described in the literature.\(^24\)

It has been observed that patients with EC have increased levels of serum IL-5 and eosinophil cationic protein. A close correlation between levels of these proteins and the clinical activity of the disease has been reported.\(^25\) The mechanism may be related to the biologic functions of each. IL-5 mobilizes eosinophils from the bone marrow, promotes homing of eosinophils, and enhances eosinophil degranulation and subsequent tissue destruction.\(^26\) Eosinophil cationic protein is a ribonuclease secreted by activated eosinophils and is toxic to bacteria and helminths as well as mammalian cells.\(^27\)

The most common clinical presentation described in children has been edematous erythematous plaques, whereas in adults the lesions more commonly appear granulomatous.\(^28\) Findings that have been found in association with EC include fever, lymphadenopathy, arthralgia,\(^2\) and anterior uveitis.\(^29\) Secondary infection also may occur as a complication.\(^11,30\)

**Figure 1.** Multiple erythematous, edematous, annular plaques (A) on the anterior legs with pseudovesiculation (B and C).
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Eosinophilic cellulitis should be included in the differential diagnosis of erythema annulare or presumed cellulitis when antibiotics are ineffective. A complete history should be obtained including recent infections, arthropod bites, bee stings, medications, and vaccinations; stool should be tested for ova and parasites for identification of a possible trigger; and a complete blood cell count with differential should be ordered. A biopsy should be performed for a definitive diagnosis. An extensive hematology/oncology workup is not warranted unless systemic features are present or the course is prolonged.26

REFERENCES


Figure 2. Lower-power photomicrograph of dermal inflammatory changes and multiple flame figures (A) (H&E, original magnification ×20). Higher-power view of dermal infiltrate of eosinophils, histiocytes, and lymphocytes, as well as flame figures (B) (H&E, original magnification ×40).

Adult patients with EC usually experience a rapid response to oral steroids. The recommended starting dosage for prednisone is 1 to 2 mg/kg daily and 5 mg every other day for persistent disease.31,32 Pediatric patients also respond well to systemic steroid therapy and/or medium- to high-potency topical steroids, depending on the extent of disease.26 It has been recommended that oral prednisolone or prednisone be administered at a dosage of 2 mg/kg daily for 5 to 7 days with a taper over 2 to 3 weeks.26 Evidence for the efficacy of lower-dose systemic steroid therapy 1 mg/kg daily is provided by the outcome of the case reported herein. There is a considerable range in the course of EC from rapid resolution, as in our case, to more recalcitrant disease characterized by exacerbations and remissions.33 The mean duration of disease for adults and children is 5 and 3 years, respectively.28
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