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Case Report Open Access

Juvenile Spring Eruption in 4 Male Family Members over 3 Generations: A Case Report

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Abstract

Juvenile Spring Eruption (JSE) is a unique localised photodermatoses characterised by the development of a papulovesicular rash on the helices of the ears of young boys. This condition typically occurs after sun exposure in the springtime and is considered a variant of polymorphic light eruption. It receives scant attention in the dermatology literature and is likely to be underreported. In addition, the heritability of JSE has yet to be established. Here, we describe the case of a ten-year-old patient with JSE who has three male family members with the same condition, spanning three generations.

Keywords: Juvenile spring eruption; Family history; Heritability

Introduction

Juvenile Spring Eruption (JSE) is characterized by a papulovesicular rash on the light-exposed helix of the ears. It typically affects boys and young men and occurs after time spent outdoors in the early spring. It is considered a localized variant of polymorphic light eruption (PMLE) [1]. The condition is infrequently reported within the literature, but is believed to be more prevalent than this would suggest [1-3]. Most authors report outbreaks in the form of small epidemics, however, a positive family history is rarely mentioned [2,4]. Here, we describe the case of a patient with JSE who has three male family members with the same condition. Those affected are on the paternal side of the family and span three generations.

Case Report

A 10-year-old boy with no significant family history presented to his general practitioner with an itchy rash on his ears. This occurred one to two hours after he had been playing outside at school in early springtime. He had developed the same rash several times in the past in similar circumstances. On examination, the patient had a papulovesicular rash on the helixes of both ears with surrounding erythema. Symptoms resolved without residual scarring within one week. Laboratory investigations including full blood count, renal and liver function were normal. Anti-nuclear antibodies, anticardiolipin antibodies and serum electrophoresis were undertaken to exclude chilblain lupus erythematosus and perniosis, as these conditions may present with similar clinical findings [5].

The results for all three tests were normal. A clinical diagnosis of JSE was made based upon the presentation of an abrupt onset papulovesicular rash on the helices following sun exposure. On further enquiry, it was discovered that an identical eruption had occurred in 3 other family members, namely the patient's father, his uncle and his grandfather. All affected family members were on the paternal side. The patient's father still suffered from the rash on occasion, although less frequently than in younger years. The eruption would occur in the same circumstances; early springtime following a period of outdoor activity in the sun. No female family members reported symptoms.

Discussion

JSE is characterised by the onset of papulovesicular lesions on the ear helices after sun exposure in the early spring. It primarily affects young males between the ages of 5 and 12, although cases in young adulthood have also been reported [1-2,4]. It is considered a variant of polymorphic light eruption, although provocative phototesting is usually negative. PMLE is triggered by the UVB waveband in sunlight (290-320 nm). It is hypothesised that the pathophysiology of PMLE involves UVB induced modification of DNA. This leads to the formation of new antigens and a subsequent cell mediated autoimmune response. Similar autoimmune mechanisms may underlie JSE [6]. In JSE, lesions are limited to the ears and may evolve into blisters and crusts after 24-48 hours [1-4,7]. It is a self-limiting disorder, resolving in 2-3 weeks with minimal or no scarring [1,6]. Short hair and protuberant ears are commonly associated. Recurrences can be seen but the condition does not tend to persist into late adulthood [4]. The true incidence of JSE is unknown. The condition spontaneously resolves and therefore is likely to be under-reported and more prevalent than previously thought [1].

The majority of cases of JSE occur in the form of small epidemics [2,8,9]. The first reported outbreak in the UK took place in 1954: 121 out of 150 children in a camp in Surrey developed pruritic lesions affecting their ears after playing outside in the sun during the springtime. No infectious agent was identified and the condition was subsequently attributed to a combination of sunlight and cold weather [9,10]. There are subsequent reports of JSE erupting in cohorts of military soldiers with short haircuts following outdoor exercises [4,11]. Several authors have highlighted the possible role of cold weather in addition to sunlight in the pathophysiology of JSE, given that outbreaks tend to occur on cool spring mornings as opposed to warm summer days. In contrast to polymorphic light eruption, provocative photo-testing is frequently negative, which suggests that the etiology of

JSE differs and other factors such as genetics may play a role. As the JSE variant receives scant attention in dermatological texts, its heritability has yet to be established [12,13]. We reviewed the literature for reports of a positive family history of JSE. Berth-Jones et al. describe three cases of juvenile spring eruption occurring in a father and two sons [3]. Both Roelof van Ewijk and Warin report cases of JSE occurring amongst brothers [14,15] In one instance the rash resolved in one of the brothers after he grew his hair. In a larger case series of 18 patients with JSE, 13 reported a vague family history of the condition. All of those affected were male [13].

Provocative photo-testing and a skin biopsy was not carried out on our patient as diagnosis was made on clinical grounds. According to previous reports, histology appearances of JSE mimic those of PMLE, namely a mild perivascular lymphohistiocytic infiltrate in the papillary and reticular dermis [13].

In our family, the involved members were males on the paternal side of the family. They report being affected to a similar degree of severity and would experience recurrent episodes of a blistering rash on the ears during the springtime, throughout boyhood and into their early twenties. The patient's father still occasionally suffers from the rash, especially in sunny springs. Interestingly, the patient's maternal half-brother has remained unaffected despite participating in outdoor activities at the same time of year. As for the patient, symptoms resolved without residual scarring within a week with just sun avoidance advice.

Conclusion

Although environmental and genetic factors have been proposed to play a role in the pathogenesis of JSE, the only well-established contributing factor has been exposure to ultraviolet radiation [1]. There are a handful of cases in the literature that have anecdotally demonstrated the occurrence of JSE in two or more members of the same family [3,4,13,15,16]. In this case, we are able to strengthen the hypothesis of a genetic component to JSE.

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