Volume 21 Number 6
June 2015

Case presentation

A case of self-healing juvenile cutaneous mucinosis

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Dermatology Online Journal 21 (6): 10

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Abstract

Importance: Self-healing juvenile cutaneous mucinosis is a very rare, self-limiting disease characterized by the abrupt onset of asymptomatic papules and nodules located primarily on the face and periarticular regions of a juvenile patient. There have been less than 20 cases reported since it was first described in 1973.

Observations: Most cases have been reported in children 15 years and younger. Herein we present a case affecting a 17-year-old. To our knowledge, this the oldest reported patient with this condition in the USA.

Conclusions and Relevance: Despite the rarity of this disease, it is important to keep SHJCM on the differential in pediatric patients presenting with proliferating papules and nodules. Knowledge of this entity may prevent unnecessary diagnostic testing and aggressive treatment in the pediatric population with this self-limited disease.

Keywords: Self-healing juvenile cutaneous mucinosis, SHJCM, pediatric nodules

Introduction

Self-Healing Juvenile Cutaneous Mucinosis (SHJCM) is a rare disease characterized by the abrupt onset of asymptomatic papules and nodules located primarily on the face and periarticular regions of a juvenile patient [1]. It was first described in 1973 by Colomb [2] and Bonerandi [3] in the French literature and since then, less than 15 case reports or small series have been reported [4]. As implied by the name, this disorder is characterized by the spontaneous resolution of lesions over weeks to months. Unlike other mucinoses, SHJCM is not associated with paraproteinemias or other systemic disorders. Herein we report this condition in a 17-year-old patient. To our knowledge, this is the oldest patient presenting with this condition in the United States.

Case synopsis

A 17-year-old boy with a history of atopic dermatitis presented to clinic with a slowly growing, slightly tender, skin-colored to erythematous nodule on his left inferior forehead. Except for mild erythema, the overlying skin was normal. The patient was otherwise in good health and denied any fevers or systemic symptoms. He denied any recent trauma or arthropod bites to the area. The nodule was initially suspected to be an inflamed epidermal inclusion cyst and was treated with doxycycline without improvement. The patient returned to clinic several months later with a new similar skin-colored subcutaneous nodule on his left superior forehead, adjacent to the original nodule (Figure 1). A punch biopsy was performed for further evaluation. (Figure 1).

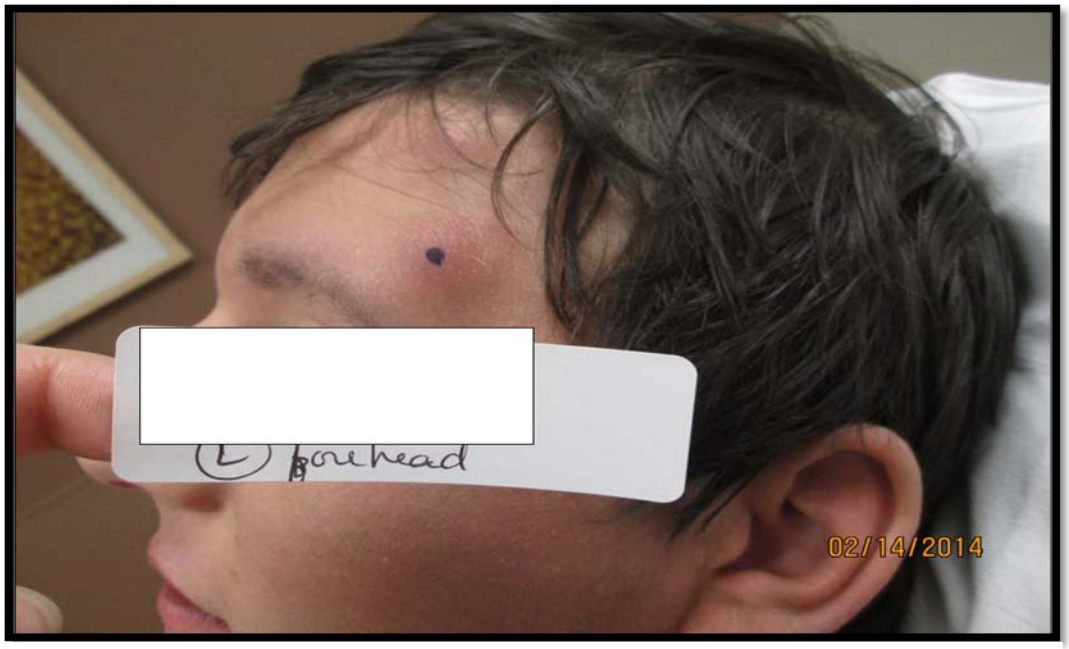
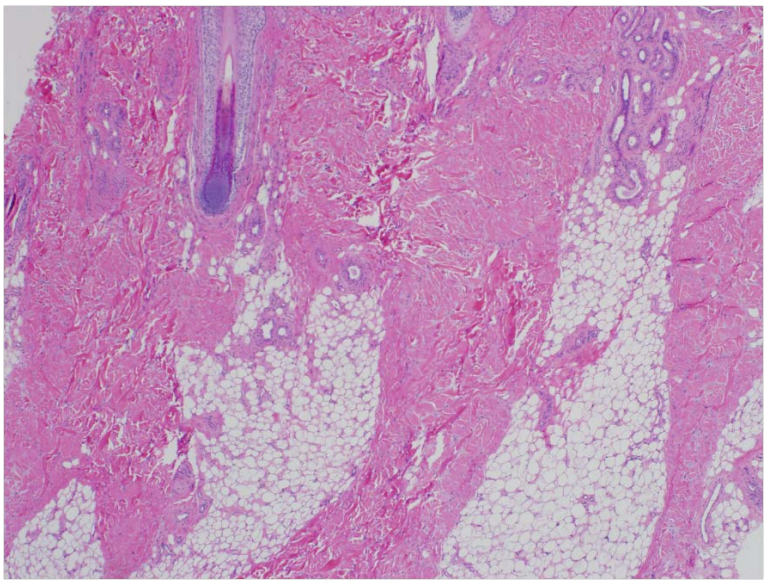


Figure 1. Two subcutaneous erythematous nodules on patient's left forehead.



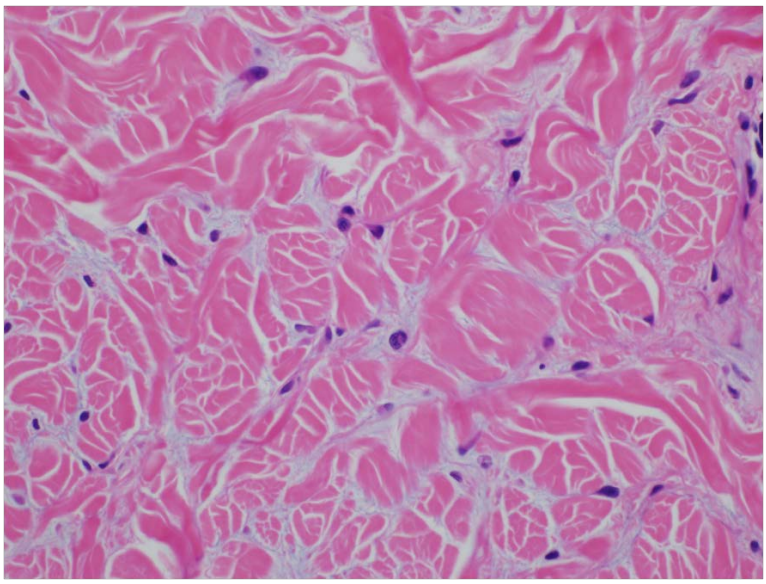


Figure 2. Low power (4x) view of punch biopsy specimen from left forehead **Figure 3.** High power (20x) view of punch biopsy specimen from left forehead

Histopathologic features of the biopsy revealed a diffuse deposition of mucin throughout the majority of the dermis and widening of the septae. There was an increase in collagen deposition and a marked proliferation of irregularly arranged fibroblasts (Figure 2). At higher power, the mucin was noted to separate the collagen bundles in the dermis. A sparse lymphocytic perivascular infiltrate was present. In addition, conspicuous ganglion-like mononuclear cells were also noted (Figure 3). An alcian blue stain was used to demonstrate the significant increased presence of mucin in the specimen.

Our patient had no associated symptoms. However, a baseline lab evaluation was performed to rule out underlying diseases that may accompany mucinoses. A complete blood count, liver function tests, renal function panel, thyroid stimulating hormone, creatinine kinase, aldolase, and serum and urine protein electrophoresis were all within normal limits. He presented for follow up in January 2015. He reported that the nodules initially slowly increased in size for approximately 4 months prior to stabilizing. On the follow up exam, the lesions, although still present, had begun to partially involute as compared to his first visit in October 2013. The continued presence of these lesions after 18 months does not contradict the diagnosis of SHJCM because similar cases have persisted for as long as two to three years [2, 5, 6].

Discussion

Self-healing juvenile cutaneous mucinosis is a very uncommon disease characterized by the rapid onset of asymptomatic papules and nodules involving the face, periarticular regions, abdomen, and thighs [1]. It is a pediatric illness with a reported age range from one to fifteen years [4]. However, there is one case described in a 26-year old patient from Spain [5]. In most cases, lesions resolve spontaneously in weeks to months, with three cases persisting for 2.5 years [2, 5, 6]. Although SHJCM is usually not associated with inflammatory symptoms, there are cases presenting with fever, weakness, arthralgias, and myalgias [3, 4, 7, 8]. To our knowledge, SHJCM has been reported in the literature less than 20 times since its original description in 1973.

The etiology of SHJCM is not understood. A current hypothesis includes abnormal fibroblast and mucin production secondary to a reactive or reparative response related to chronic antigenic stimulation, such as viral infection or inflammation [5, 6]. Interestingly, there have been cases reported in association with positive *Bartonella* titers [1], upper respiratory tract infections [4, 7, 8], and chemotherapy treatment for nephroblastoma [6]. However, there is no definitively established association.

Clinically, three types of cutaneous lesions of SHJCM have been described: (1) nontender, ivory-white, grouped papules commonly arising on the head, neck, trunk, and periarticular areas; (2) deep subcutaneous nodules that favor the face and periarticular regions; and (3) hard periorbital and zygomatic edema [7]. Our patient presented with the second type with deep subcutaneous nodules that were limited to the face. It is interesting to note that the nodular type may be the only presenting feature, which can make the diagnosis of this rare entity even more difficult as it can be confused with other pathologies [4]. Self-healing juvenile cutaneous mucinosis usually lacks inflammatory symptoms but there have been reported cases of painful polyarthritis and swelling of the knees, elbows, and hands [7].

Histologic examination and description of both the dermal papules and subcutaneous nodules have been well described [1, 4]. However, most cases have reported on biopsies taken from the papules and not the subcutaneous nodules [1]. The characteristic histopathologic features of the papules include edema of the papillary and reticular dermis, separation of collagen bundles, slight increase in fibroblasts, a mild perivascular infiltrate, and alcian blue staining at pH 2.5 (hyaluronic acid) but not pH 0.5 [5, 7, 9, 10]. The deeper nodules have been less often described but tend to have mucinous areas in the mid and deep reticular dermis, arborizing thin-walled vessels, and prominent plump to spindle-shaped fibroblasts [8]. Nodules may also contain large epithelioid ganglion-like mononuclear cells embedded in a myxoid stroma [4, 11]. Immunohistochemical stains are negative for cytokeratin, epithelial membrane antigen, muscle actin and CD34 [11].

The clinical appearance and histologic findings of the nodules alone without papular involvement may raise suspicion for proliferative fasciitis. This is because subcutaneous and fascial histopathology in SHJCM is virtually indistinguishable from proliferative fasciitis [1]. Ganglion-like giant cells and connective tissue mucin is prevalent in both diseases. As implied by Nagaraj et al., the pathologist should not be distracted by the dominant fasciitis changes in the deep dermis but should focus their attention on the less prevalent collection of mucin in the dermis to aid with the diagnosis [1]. In contrast with SHJCM, proliferative fasciitis usually presents on the extremities of adults as an asymptomatic, firm, palpable 1 to 5 cm subcutaneous nodule with a history of trauma [12]. Thus, confusion between SHJCM and proliferative fasciitis may be especially true in the older pediatric population in which a history of trauma is more likely to be elucidated.

In contrast to adult mucinoses, SHJCM has not been associated with systemic disorders such as a paraproteinemia, bone marrow plasmacytosis, or thyroid disease. There are also no proven associations with autoimmune disease, although there are two reports of patients with increased aldolase levels suggesting the possibility of an association with juvenile dermatomyositis [5, 8]. Of note, there has been one case reported in association with nephroblastoma and another reported with carpal tunnel syndrome, but the significance of these associations is unclear and may be coincidental [6].

In conclusion, we presented a case of self-healing juvenile cutaneous mucinosis in a 17-year-old patient with only facial subcutaneous nodules. We reviewed the three common dermatologic findings and the associated histological findings. Additionally, we compared the similarities and differences with proliferative fasciitis. Key features to aid in the distinction between SHJCM and proliferative fasciitis include the lack of trauma in SHJCM and the deposition of mucin in the dermis. Despite the rarity of this disease, it is important to consider SHJCM in pediatric patients presenting with proliferating papules and nodules. Knowledge of this entity may indeed prevent unnecessary diagnostic testing and aggressive treatment in pediatric patients with this self-limited disease.

Acknowledgements: Cheri Adgerson, M.D.

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