MEDDOCS Open Access Publisher

ISSN: 2639-9237

Journal of Case Reports and Medical Images

Open Access | Case Report

Mucinous Mayhem: A Case of Self-Healing Juvenile Cutaneous Mucinosis in a Filipino Boy

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Received: June 08, 2023

Accepted: June 23, 2023

Published Online: June 30, 2023

Journal: Journal of Case Reports and Medical Images

Publisher: MedDocs Publishers LLC

Online edition: http://meddocsonline.org/

Keywords: Self-healing juvenile cutaneous mucinosis; Cutaneous mucinosis; Mucin.

Abstract

Self-Healing Juvenile Cutaneous Mucinosis (SHJCM) is a rare, self-limiting disease with an unknown etiogenesis. Pediatric patients present with a variety of skin lesions that spontaneously resolve in a matter of weeks to months. We present a case of a 14-year-old Filipino boy with a 2-month history of recurrent well-defined, nontender nodules with surrounding halo-like ecchymoses on the trunk and extremities that spontaneously resolve in 10-14 days. There were no previous reports of trauma over the affected areas. No other systemic symptoms were noted in the patient. Patient's co-morbidities include allergic rhinitis and atopic dermatitis but is otherwise well. Patient tested negative for Anti-Nuclear Antibody (ANA). Histopathological findings revealed mucin deposition in the reticular dermis up to the subcutaneous tissue which stained positive with Alcian blue. This is the first reported case of SHJCM in a Filipino boy. We report this case together with a brief overview and update on the current literature regarding SHJCM.

Introduction

Self-Healing Juvenile Cutaneous Mucinosis (SHJCM) is a rare form of cutaneous mucinosis in the pediatric population. It presents as an acute eruption of lesions with varied morphologies, ranging from papules to nodules, that spontaneously regress, without the need for medical or surgical intervention. Currently, only a few cases have been reported and are available in the literature. This sudden eruption of lesions may alarm and cause unwanted distress to the patients' parents or caretakers. Clinical and histopathologic examination are of equal importance in diagnosing this disease. In this article, we present a case of SHJCM presenting as recurrent nodules in a Filipino boy, as well as a brief review of the available literature on SHJCM. To the authors' knowledge, this is the first reported case of SHJCM in a Filipino patient. Familiarity with this disease is essentials for clinicians to avoid unnecessary and invasive testing in affected patients, and in reassuring our patients of its benign, self-limiting nature.

Case Presentation

We present a case of DA, a 14-year-old Filipino boy with a 2-month history of recurrent nodules on the trunk and extremities, without any history of trauma nor injury prior to the appearance of the lesions. He was previously diagnosed with allergic rhinitis and atopic dermatitis but not on any maintenance nor controller medications. The family history is also unremarkable. The nodules initially appeared as two well-defined, slightly tense, round, smooth, nontender, nonpruritic, movable 0.5 x 0.5 cm. nodules with surrounding ecchymoses on the anteromedial aspect of the right knee. The patient did not have any other associated symptoms such as fever, malaise, dyspnea, chest pain, abdominal pain and joint pain. The lesions gradually improved and eventually resolved completely with no complications over the course of 10-14 days without any treatment instituted. One week prior to being seen at the clinic, a recurrence of another lesion of the same quality appeared over the left forearm (Figure 1). Upon consultation, general physical ex-

Cite this article: Juan Paolo DSV, John Ruben CV, Amanda Christine FE, Eileen Liesl AC. Mucinous Mayhem: A Case of Self-Healing Juvenile Cutaneous Mucinosis in a Filipino Boy. J Case Rep Clin Images. 2023: 6(1): 1145.

amination was unremarkable. On dermatologic examination, there was a solitary, well-defined, firm, nontender nodule measuring 0.5 x 0.4 cm. with surrounding ecchymosis on the left forearm. During this time, the initial impression of the clinician was traumatic panniculitis versus lupus panniculitis. A complete blood cell count was done which showed values within normal range. Anti-Nuclear Antibody (ANA) testing revealed a negative result. Histopathologic examination via 6-mm punch biopsy (Figure 2) revealed sparse perivascular and interstitial infiltrates of lymphohistiocytes, and mucin deposition in the dermis that extend to the subcutis in a septo-lobular pattern. Within these dense mucin collections are an increased number fibroblasts, as well as scattered, large, gangliocyte-like cells. Alcian blue staining confirmed this deposition to be mucin. These findings were consistent with self-healing juvenile cutaneous mucinosis.

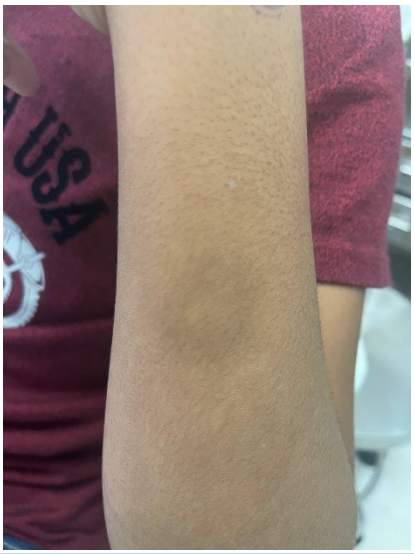

Figure 1: Left forearm of DA. 0.5 x 0.4 cm. solitary, well-defined, nontender, moveable nodule with surrounding ecchymosis on the left forearm.

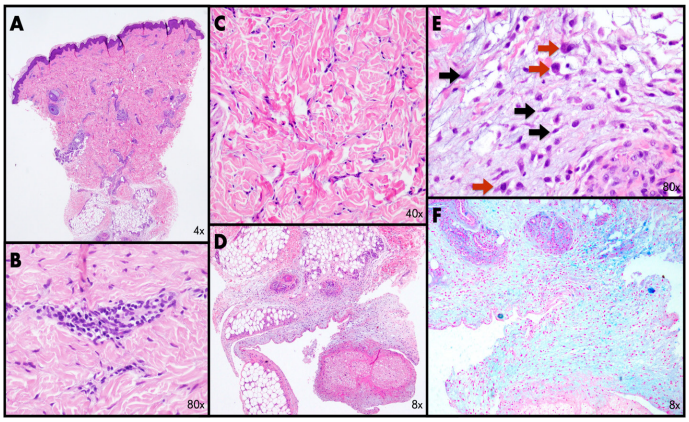

Figure 2: Histopathologic examination of left forearm nodule. **A** and **B)** Sparse perivascular infiltrate composed of lymphocytes. **C)** Interstitial mucin deposition in the dermis. **D)** Septo-lobular subcutaneous inflammation and mucin deposition. **E)** Presence of multiple spindle-shaped fibroblasts (black arrow) as well as large gangliocyte-like cells (red arrow). **F)** Alcian blue stain highlighting the dermal and subcutaneous mucin deposition.

Two weeks after the initial consult, there was complete resolution of the lesion on the left forearm without any noted sequelae nor complications. A new nodule on the right shoulder appeared two months after the initial consult upon teledermatology follow-up, but the lesion also spontaneously resolved without complications after two weeks without any treatment instituted.

Discussion

Self-healing juvenile cutaneous mucinosis is a rare, self-limiting disorder of the pediatric population that is characterized by the abrupt appearance of papules, nodules or plaques on different parts of the body, particularly on the face and surrounding joints [1,2]. According to available literature, patients afflicted with this condition range from 13 months to 15 years of age [3,4]. The first reported case of SHJCM was described in 1973 by Colomb, Racouchot and Vittori, in a previously healthy 13-year-old female, who presented with nodules on the scalp, hands, elbows and knees [1,4,5]. To date, there are only a handful of cases diagnosed to have SHJCM and reported in the literature.

Cutaneous lesions vary in presentation and may include any of the following [1,2,4,6,7]:

- Non-tender, ivory papules on the head, neck, trunk or periarticular regions as well as similar lesions arranged linearly on the chest and abdomen.
- Subcutaneous nodules on the head and periarticular regions.
- And hard, non-pitting periorbital and zygomatic edema.

The periarticular regions affected are usually the knees, elbows and hand joints. This acute eruption of proliferative lesions is most often not associated with other inflammatory signs and symptoms [7]. But there have been previous reports of patients presenting with fever, arthralgia [6], edema of the joints and hands, muscle fatigue and temporary hypertension [1].

Correlation of both the clinical picture as well as the histopathologic examination is of particular importance in this disease to distinguish it from other diseases that may present as proliferative nodules [8]. An incisional biopsy is the preferred manner of sample acquisition to be able to completely assess dermal, subcutaneous and fascial planes. Histologic findings may vary depending on the characteristic lesion sampled; however, all lesions will present with the following constant features:

- dermal mucin deposition that may extend to the subcutis and fascia, highlighted with an Alcian blue stain (pH 2.5).
- Increased number of spindle-shaped fibroblasts, and
- identification of peculiarly large gangliocyte-like mononuclear cells.

Aside from these key histopathologic features, a sparse perivascular lymphohistiocytic infiltrate may also be noted. Histopathologic examination of papules show dermal mucin deposition with extension to the subcutis. Nodules, on the other hand, still present with dermal mucin deposition but are more conspicuous within the subcutis and may sometimes extend to the fascial plane. The closest histologic differential for this disease is proliferative fasciitis, which also presents with mucin deposition, fibroblast proliferation and large, gangliocyte-like cells [6,7]. It must be noted that the presence of mucin in the dermis is consistently seen in SHJCM, while the mucin deposition in proliferative fasciitis involves deeper areas, within the subcutaneous and fascial planes [6]. Deposition of mucin in the dermis is the most typical finding that is seen in the histopathologic analysis of any lesion of SHJCM, allowing differentiation from proliferative fasciitis which tends to affect deeper structures in

the subcutis and fascia [4]. Clinically, lesions of proliferative fasciitis tend to be a single nodule while in SHJCM, it is typical to see more than one lesion affecting the patient [7]. Proliferative fasciitis affects mostly adults, with a third of patients having a history of trauma, while in SHJCM, patients are of the pediatric population without a previous history of trauma in the affected areas [6,7].

Despite increasing numbers of cases reported, the pathogenesis of SHJCM has remained elusive [5,7,8]. A proposed mechanism involves chronic antigenic stimulation from an uncertain trigger [2,4,5], probably from an infectious or inflammatory source, that is believed to induce fibroblast proliferation and mucin deposition. As was mentioned in previous case reports, these antigenic stimulants may be in the form of infections (Upper respiratory tract infection, *Bartonella* sp.) [3], drugs (chemotherapeutic drugs) or even tumors (nephroblastoma) [7]. Due to the periarticular location of the nodules, some also believe that mucin deposition may be a response to an underlying joint inflammation [8]. Deranged ancillary laboratory examinations may also be noted in some patients such as increased Erythrocyte Sedimentation Rate (ESR), lymphocytosis, increased aldolase and positive antibodies against *Bartonella* species [3,4].

It is important to note that compared to other mucinoses in the adult population, SHJCM has not been found to be associated with other systemic disorders such as paraproteinemia, bone marrow plasmacytosis and thyroid disease [1,4,6,7]; this may be attributed to the lack of data from long-term follow-up of patients with SHJCM. In a case series by Luschinger et al, one patient had progressive development of erosive polyarthritis as well as sclerodactyly and transitioned to having fibroblastic rheumatism while another patient developed an autoinflammatory rheumatologic disease [8]. Currently there is no strong evidence to support a significant association between SHJCM and rheumatologic conditions as current data are lacking [1,7].

Despite its worrisome presentation, as a rule, lesions of SHJCM resolve spontaneously [5]. According to the available literature, time to resolution of the disease vary from several weeks to as long as 2.5 years [3]. It is recommended that a semi-annual follow-up for the first two years, followed by an annual follow-up for two more years after, be advised for patients diagnosed with SHJCM [8]. Bothersome or permanent sequalae and complications are not expected in the course of this disease. Early recognition, patient education and reassurance remain to be the cornerstone of management in SHJCM as this disease is self-limiting. Knowledge of this disorder allows us to reassure our patients and their parents or watchers of its self-resolving nature, and for clinicians to avoid unnecessary testing and aggressive therapy in these pediatric patients.

Conclusion

Self-healing juvenile cutaneous mucinosis is an uncommon, self-limiting pediatric disorder characterized by the appearance of asymptomatic papules, plaques and nodules that resolve without permanent sequelae. A good history and physical examination coupled with histopathologic testing enables us to accurately diagnose this disease. We must always place it in our differential diagnoses when presented with patients having similar lesions. Identifying this disease allows us to reassure our patients of its benign prognosis. Reports with long-term follow-up of patients are currently lacking; therefore, it is advisable to have at least an annual examination of these patients to further understand and recognize permanent complications that may arise from this disease.

Funding

This study was self-funded by the authors.

Conflict of interest

Drs. Villena, Valeza, Esquivel, Cubillan declare no conflict of interest.

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