Self-Healing Juvenile Cutaneous Mucinosis: A Case Report in the Middle East

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Conflict of interest: None declared

Patient: Male, 12
Final Diagnosis: Self-healing juvenile cutaneous mucinosis
Symptoms: Facial edema • nodular skin lesion
Medication: —
Clinical Procedure: None
Specialty: Pediatrics and Neonatology

Objective: Rare disease

Background: Self-healing juvenile cutaneous mucinosis (SHJCM) is a rarely diagnosed disease worldwide, with less than 20 reported cases in the literature. It is characterized by a rather benign course in juvenile patients with nodular and mucinous skin eruption and edema.

Case Report: A 12-year-old male patient previously healthy presented to the pediatrics clinic with a 1-week history of bilateral palmer pruritus and plantar tenderness upon walking, preceded by eruption of erythematous patch on his neck. The disease course evolved to include facial edema, erythema, nodular skin eruptions with a completely negative initial workup. The patient was labelled as a juvenile idiopathic arthritis patient and doomed to be a candidate for corticosteroid therapy. Upon further workup, a skin biopsy was taken and SHJCM was diagnosed. Complete resolution of symptoms was witnessed on symptomatic treatment after 5 months of diagnosis.

Conclusions: To our knowledge, this is the second case of SHJCM reported in the Middle East and the first to be reported in Lebanon. It is also the first case reported to have the longest follow-up period; 10 years of follow-up with no new findings or relapse. SHJCM is a rare disease whereby awareness of its features and presentation may help in diagnosing it and preventing unnecessary testing and aggressive treatment for a rather benign disease.

MeSH Keywords: Child • Mucinoses • Rheumatoid Nodule

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Background

Self-healing juvenile cutaneous mucinosis (SHJCM) is a rarely diagnosed disease worldwide, with less than 20 reported cases in the literature. Typically, the disease affects young individuals and manifests as multiple, transient papular and nodular lesions particularly involving the face, thighs, abdomen, and peri-articular areas. Although poorly elaborated in the literature, the disease course has been described as benign with spontaneous recovery expected within weeks to months in all patients with few exceptions. In this report, we present a case of SHJCM with a review of the literature, defining the characteristics and histologic findings of this rare disease. To our knowledge, this is the second reported case in the Middle East region and first in Lebanon. It is also the first case with long-term follow-up of 10 years after diagnosis. This is important for highlighting the role of follow-up in this disease entity.

Case Report

A 12-year-old male, previously healthy, presented to the pediatric clinic for a 1-week history of bilateral palmar pruritus and plantar pain. Three days prior to presentation, the parents also noticed the appearance of a non-pruritic, erythematous patch on the posterior aspect of his neck. The patient had no fever, and no signs of focal infection. History was negative for any insect bites and the patient had no known allergies. On closer inspection, the erythematous patch extended from the neck to the upper back (Figure 1) with no evidence of any papules, pustules, or vesicles. His palms and soles were completely unremarkable. A comprehensive physical examination was also within normal limits. Laboratory findings revealed a normal blood cell count, hemoglobin levels, negative inflammatory markers including c-reactive protein, sedimentation rate and liver function tests. Serologies for Epstein-Barr virus, varicella-zoster virus, and parvovirus B19 were all negative.

One week after the initial presentation, the patient returned with frontal, periorbital, and upper lip edema and erythema. Concomitantly, peri-articular nodules appeared on the proximal interphalangic joints bilaterally (Figure 2), with a reported increase in pruritus and pain in the soles upon ambulation. Repeated laboratory tests remained within normal ranges with normal creatine phosphokinase and lactic dehydrogenase levels.

Figure 1. Upon presentation, an erythematous patch on the neck and upper back.

Figure 2. Periarticular nodules on proximal inter-phalangic joints bilaterally and peripheral cyanosis.
The patient was referred to a rheumatologist, and a clinical diagnosis of juvenile idiopathic arthritis was evoked despite negative serologies: antinuclear antibody (ANA), Anti-Scl (anti-topoisomerase), rheumatoid factor (RF). The patient was started on nonsteroidal anti-inflammatory drugs and desloratadine.

Three weeks after initial presentation, the patient was no longer able to walk due to the plantar edema and knee pain. Similar nodules also appeared on his nose (Figure 3), fingers, and left elbow. The parents also reported bluish discoloration of the fingertips (Figure 2).

Subsequently, an excisional biopsy of an elbow nodule was performed. It showed fibrovascular proliferation in a septo-lobular pattern, reminiscent of proliferative fasciitis, set within myxoid stroma, dissociating and surrounding some fatty islands (Figure 3). The tissue also showed polymorphous inflammatory cells and essentially numerous large gangliocyte-like cells (Figure 4A, 4B). Abundant mucin deposition was observed on histology (Figure 5). There was no evidence of epithelioid granuloma or other element in favor of specific inflammation or malignancy. These features, along with the presence of mucin deposition, were most consistent with the diagnosis of SHJCM.

The patient had progressive amelioration on symptomatic treatment only. Within 5 months of onset of symptoms, the patient had complete resolution of his nodules with no more pain.

Discussion

Self-healing juvenile cutaneous mucinosis (SHJCM) is an extremely exceptional disease. It was first described in 1973 in the French literature [1]. It was not until 1984 when it was first described in the English literature [2]. Since then, only 20 cases have been reported. Despite a worrisome presentation, it has a benign course and early detection relieves patients from unnecessary therapy and stress. All the cases reported shared 4 common characteristics: young age at presentation, similar pattern of cutaneous lesions, absence of systemic findings, and abrupt onset followed by spontaneous complete resolution [3]. Patients were in the pediatric age group (ages mostly ranging from 1 year old to 17 years old) with one exception of...
a 26-year-old patient diagnosed with SHJCM reported by de las Heras in 1996 [4]. We hereby present a table summarizing all case reports reported in the literature to date (Table 1). Patients with SHJCM have similar cutaneous manifestations with similar distribution. The skin lesions can be categorized into 3 types [5–8]. 1) Small, ivory colored, non-tender papular lesions involving the head, neck, trunk and periarticular spots. 2) Deeply located nodules on the face and periarticular

### Table 1. Summary of reported cases of SHJCM in literature.

| Reference number | 1st author | Age & sex | Case characteristics |
|------------------|------------|-----------|----------------------|
| 1                | Colomb D  | 13 years old (F) | • Previously healthy  
• Characteristic lesions on scalp, hands, elbows, knees |
| 2                | Pucevich MV | 13 years old (M) | • Arthralgia myalgia  
• Loss of appetite, hoarseness,  
• Carpal tunnel syndrome  
• Lesions on face, trunk, knees, hands, elbows |
| 4                | De las Heras ME | 26 years (F) | • Healthy  
• Abrupt eruption of papular lesions involving scalp, face, neck, trunk, hands |
| 5                | Caputo R  | 5 years old (M) | • Previously healthy  
• Scleroderma of the face, knees and hands  
• Periarticular papular lesions and arthralgias  
• Fever, myalgia, weakness |
| 6                | Bonerandi JJ | 14 years old (M) | • Arthralgia  
• Elevated blood pressure  
• Papular lesions on hands, face, knees, trunk, neck |
| 7                | Carder KR  | 6 years old (M) | • Previously healthy  
• Asymptomatic  
• Lesions on arms and thighs |
| 8                | Hershko K  | 21 months old (F) | • Healthy  
• Papular lesions on face, trunk, periarticular regions |
| 9                | Cowen EW  | 3 years old (M) | • Arthralgia, elevated blood pressure  
• Myositis and subcutaneous edema  
• Multiple lesions on axillae, legs, scalp, face, hands, trunk, knees |
| 10               | Wadee S   | 8 years old (M) | • Known to have  
• Nephroblastoma  
• On chemotherapy  
• Plaques and papules on scalp, face, sacrum, arms, dorsal hands |
| 12               | Nagaraj LV | 18 month (F) | • Healthy, asymptomatic  
• Numerous nodules on forehead, extremities |
|                  |            | 7 years (M) | • Healthy  
• Multiple subcutaneous lesions  
• Skin nodules on scalp, trunk, and extremities |
|                  |            | 6 Years (F) | • Healthy  
• Subcutaneous nodules on forehead, pruritic, non-tender skin lesions |
| 13               | Kolodziejczyk B | 4 years old (F) | • Painful lower extremities  
• Fever, cervical lymphadenopathy and hepatosplenomegaly  
• Edema of the face  
• Fine, hard, whitish nodular lesions on frontal, occipital, cervical, axillary regions and extensor areas of joints |
| 14               | Konwalter BE | 14 months (F) | • Healthy, asymptomatic  
• Multiple, firm, non-tender skin colored scaly erythematous nodules on scalp, forehead, axillae, lower legs, abdomen, hands |
| 15               | Aydingoz IE | 15 years old (M) | • Nodular lesions on trunk, face, forearms, scalp, hands and knees |
areas. 3) Non-pitting edema involving the zygomatic and peri-orbital regions.

Mild inflammatory symptoms including pyrexia, muscle fatigue, and myalgia usually coincide with the eruption of cutaneous lesions. More importantly inflammatory symptoms seldom occur but may be associated with the cutaneous lesions. These include temporary hypertension, tender joints and arthrits, and significant edema of the joints and hands. Lesions are associated with absent to mild inflammatory symptoms such as arthralgia, fever, weakness and muscle tenderness, and lack extracutaneous involvement. Other symptoms include painful polyarthritis or transient hypertension, as well as swelling of the knees, elbows, and hands [5,6,8]. With reference to our case, the diagnosis of SHJCM was mainly based on the clinical course coupled with the histologic findings. For the majority of reports, the histologic examination was performed on excisional biopsies of the popular lesions. The histologic conclusions included edema of the reticular and papillary layers of the dermis along with evidence of separated collagen bundles. Abundant monocytes and fibroblasts with perivascular infiltrates were also evident. Mucin deposition, mostly hyaluronic acid, has been identified with Alcian blue staining prepared at pH of 2.5 in the upper reticular dermis. Alcian staining at pH 0.5 and periodic acid Schiff staining are usually negative [2,5,7,9]. Wadee et al. described histologic findings from nodular lesions. The slides revealed mucin in the reticular dermis and numerous stellate shaped fibroblasts.

The initial cause of excess mucin manufacturing and increased fibroblast production is still unknown [10–13]. Some authors postulated that it may be due to a constant stimulation of an antigenic response caused by an infection or inflammation at the level of initial fibroblast and mucin production [2,11].

Contrary to other types of mucinosis, SHJCM has not been associated with systemic diseases such as systemic lupus, thyroid disorders, monoclonal gammopathy, or bone marrow plasmacytosis. However, thyroid-stimulating hormone (TSH) level, antinuclear antibody (ANA) titer, erythrocyte sedimentation rate (ESR), and serum protein electrophoresis should be ordered to rule out such associated disorders in the cases where the diagnosis is uncertain [4–9].

In all of the cases reported in the literature, SHJCM had a complete spontaneous resolution over a varied time interval, ranging from a few weeks to 3 years. [3]. Exceptions were reported whereby 2 cases relapsed into fibroblastic rheumatism and auto-inflammatory rheumatologic disease [13].

**Conclusions**

In our patient, the diagnosis was based on the clinical presentation compatible with SHJCM which included the unique pattern of the eruption, the typical lesions, the patient’s age, the absence of systemic disorders, and the spontaneous resolution of symptoms. The diagnosis was further confirmed with dermatopathology studies that were highly in favor of SHJCM. To our knowledge, this is the second case of SHJCM reported in the Middle East region and the first to be reported in Lebanon. It is also the first case reported to have the longest follow-up period; a time span of 10 years of follow-up with no new findings or relapse.

In conclusion, SHJCM is a rare disease whereby awareness of its symptoms and presentation may help in diagnosis and prevention of unnecessary testing and aggressive treatment for a rather benign disease.

**Conflict of interest**

None.

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