OMCR

Oxford Medical Case Reports, 2020;7,238–241

doi: 10.1093/omcr/omaa054
Case Report

CASE REPORT

Diffuse Idiopathic calcinosis cutis: a case report in a 13-year-old Syrian boy

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Abstract

Calcinosis cutis is characterized by the deposition of calcium salts in the skin and subcutaneous tissue. It is divided into the following subtypes: dystrophic, metastatic, iatrogenic, idiopathic and calciphylaxis. In this case, we report a 13-year-old Syrian boy with idiopathic calcinosis cutis, the lesions were unusually widespread, unlike the common condition which is usually localized to one area. The case was unrelated to any systemic or local disorders, and the patient had no complications, so no treatment was planned.

As the best of our knowledge, there are very few cases of diffuse or widespread idiopathic calcinosis cutis. We analyzed the clinical, laboratory, radiographical and pathological characteristics of our patient, which helped us to reach the final diagnosis. We will discuss the pathogenesis, investigation and management of this disease.

INTRODUCTION

Calcinosis cutis is a rare disorder characterized by the deposition of calcium salts in the subcutaneous tissue in various parts of the body. Calcinosis cutis was first described in 1855 by Virchow. From the pathogenesis point of view, there are five types of calcinosis cutis: dystrophic, metastatic, iatrogenic, idiopathic and calciphylaxis [1].

To the best of our knowledge, only a few cases of diffuse Idiopathic calcinosis cutis have been reported in the literature. In our case, we report a 13-year-old Syrian boy with diffuse Idiopathic calcinosis cutis.

CASE REPORT

A 13-year-old Syrian boy presented to the dermatology clinic with complaints of a widespread yellowish-white subcutaneous nodule on his right thigh. During 2 years of follow-up, other lesions had appeared gradually on the forearm, elbow and brachium (Fig. 1). Then the lesions showed chalky discharge (Fig. 2A) and no similar lesions were observed elsewhere in the body. There had been no increase in the number of lesions since the last visit. Clinical examination revealed palpable firm nodules below the skin. There were no signs of inflammation, joint pain or photosensitivity. His past medical history did not reveal any underlying diseases, including metabolic, autoimmune, malignant or traumatic events. There was no family history of similar complaints.

We did full blood count for the patient and all values were within normal ranges (Table 1). serum calcium and phosphate levels as well as parathormone, vitamin D hormone levels (vitamin D was checked to exclude high levels and hypercalcemia); alkaline phosphatase levels were within normal limits. Also, a 24-hour urine collection test showed normal calcium and phosphate levels. Erythrocyte sedimentation rate and Rheumatoid Factor tests were done at first to investigate inflammatory and immunological causes and the results came negative (Table 1). Then we did more specific tests to screen the most

Received: April 23, 2020; **Revised:** June 5, 2020; **Accepted:** June 9, 2020

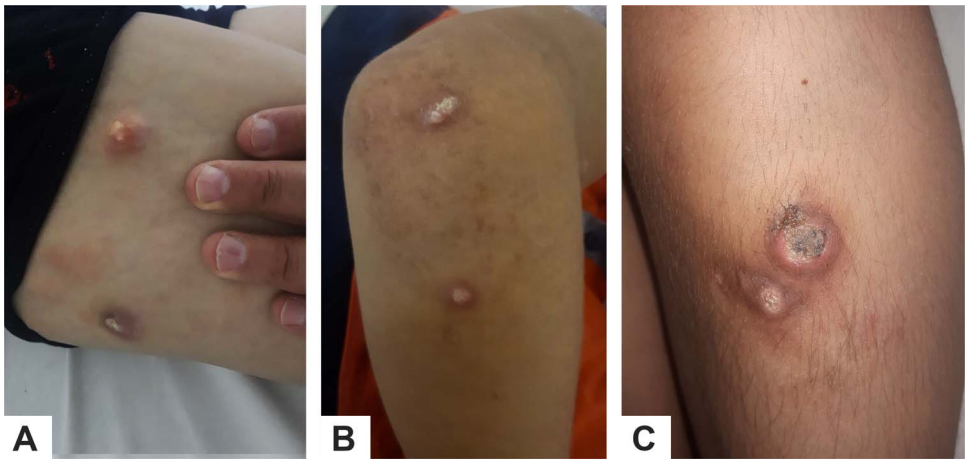

Figure 1: Gross images show widespread calcified nodules on right thigh (A) and forearm (B), nodules associated with ulcers in right brachium (C).

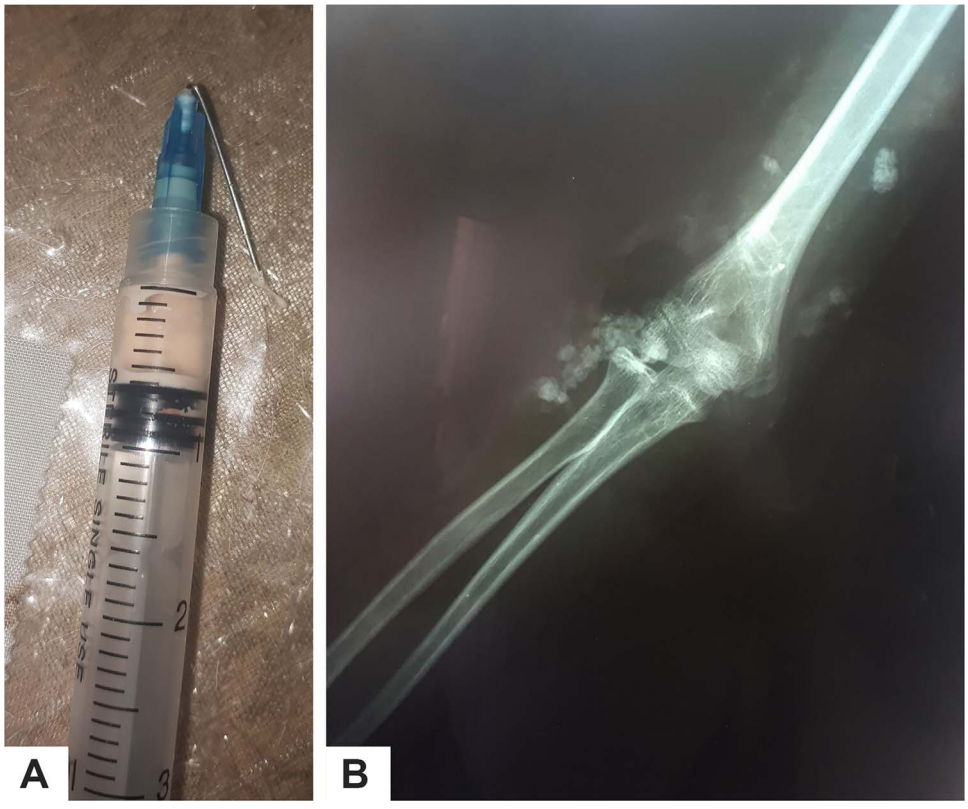

Figure 2: Drained chalky discharge from one of the nodules (A). Plain x-ray demonstrates subcutaneous calcifications around the elbow and brachium (B).

common conditions associated with the disease, Anti-Jo1 for dermatomyositis and it was negative (4.3), serological tests for systemic lupus erythematosus (SLE) and scleroderma, including antinuclear antibody (ANA) and anti-dsDNA were negative.

The possibility of familial hyperphosphatemia is unlikely because we checked-up parent's calcium and phosphate blood levels and the results were normal. Plain x-ray revealing calcification around the elbow and brachium separate from the adjacent bone (Fig. 2B).

Surgical excision was performed and histological examination of one of the nodules revealed thick, chalky discharge at the time of the procedure and microscopic massive calcium deposits (microscopic image was not available).

The patient had been seen in a dermatology clinic for the calcifications, which had been managed conservatively with regular follow-up visits for the last year and no increase in the number of lesions or changes in the patient's general health status. The patient was scheduled for follow-up annually.

DISCUSSION

Calcinosis cutis is divided into five subtypes: dystrophic, metastatic, idiopathic, iatrogenic calcification and calciphylaxis [2].

Table 1: Laboratory tests on admission

	Variable	Result	Normal range
Full blood count	WBC	8.4 10^3/ul	3.5–10 10^3/ul
	GRA%	62.3%	35–80%
	LYM%	29.7%	15–50%
	MID%	8%	2–15%
	GRAN	5.2 10^3/ul	1.2–8 10^3/ul
	LYM	2.5 10^3/ul	0.5–5 10^3/ul
	MID	0.7 10^3/ul	0.1–1.5 10^3/ul
	RBC	4.73 10^6/ul	3.50–5.50 10^6/ul
	HGB	12.1 g/dl	11.5–16.5 g/dl
	HCT%	35.1%	35–55%
	MCV	74.2 fl	75–100 fl
	MCH	25.6 pg	25–35 pg
	MCHC	34.5 g/dl	31–38 g/dl
	RDW%	11.7%	11–16%
	RDWa	42.5 fl	30–150 fl
	PLT	314 10^3/ul	100–400 10^3/ul
	MPV	7.5 fl	8–11 fl
	PCT%	0.23%	0.01–9.99%
	LPCR%	11.9%	0.1–99.9%
	PDW	9.9 fl	0.1–99.9 fl
Immunological tests	ANA (method: Immunofluorescence Antibody—IFA)	Negative	
	Anti-dsDNA (method: Immunofluorescence Antibody—IFA)	Negative	
	Anti-Jo1	4.3	Negative: up to 12 Positive: <18
Blood tests	25(OH) Vitamin D	18.40 ng/ml	30–100 ng/ml
	Alkaline phosphatase (ALP)	159 U/l	100–290 U/l
	Alanine aminotransferase (ALT)	17 U/l	10–60 U/l
	Calcium	8.3 mg/dl	8.8–10.5 mg/dl
	Creatinine	0.5 mg/dl	0.2–1.3 mg/dl
	Glucose	97 mg/dl	65–110 mg/dl
	Phosphorus	4 mg/dl	1–4.5 mg/dl
	Urea	36 mg/dl	5–50 mg/dl
	TSH	2.98 mIU/ml	0.4–6.2 mIU/ml
	ESR	Normal	
	PTH	Within normal limits	
	Rheumatoid factor (RF)	Negative	
Urine	24 h urine calcium	225 mg/24 h	100–300 mg/24 h

Dystrophic calcification present as a result of local tissue damage or abnormalities such as connective tissue disorders (syndrome, scleroderma and dermatomyositis). This type is associated with normal calcium and phosphate levels in the serum. Metastatic calcification is characterized by abnormal calcium and/or phosphate metabolism, causing the deposition of calcium in cutaneous and subcutaneous tissues. Iatrogenic calcinosis is a complication of intravenous administration of calcium or phosphate. Calciphylaxis is a calcifying vasculopathy affecting the small vessels [2, 3].

Idiopathic calcification occurs without any metabolic disorder or tissue damage. This type includes subepidermal calcified nodules, tumoral calcinosis and scrotal calcinosis. Idiopathic calcinosis cutis characterized by normal calcium and/or phosphate serum levels (except tumoral calcinosis) [4].

The subepidermal deposition usually occurs in children on the head and extremities, mainly as solitary, hard and white-yellowish papules. This calcification is most commonly localized to one area, whereas in our patient, calcified nodules were widespread [2, 5].

In order to reach our final diagnosis, we excluded other types of calcinosis cutis by following this algorithm: calciphylaxis, metastasis and tumoral calcinosis cutis were excluded because we have normal calcium and phosphate levels. Dystrophic calcinosis cutis was not expected based on normal immunological tests (Table 2) and a lack of clinical symptoms.

The previous investigations exclude any causes for dystrophic or metastatic calcinosis as well as the calcinosis universalis, leaving as with idiopathic calcinosis with an unusual widespread presentation, wherein the etiology is unknown [2, 4].

Medical and surgical treatments are options to cure calcinosis cutis, a variety of drugs, mainly bisphosphonates, intralesional corticosteroids, aluminum hydroxide, warfarin and diltiazem, have been tried with limited success. Most medical treatments, though beneficial in some cases, are not recommended for children as the risks outweigh the benefits. The local excision of painful or ulcerated nodules is a therapeutic option but local recurrence is common. The present case was unrelated to any systemic or local disorders, and the patient had no complications, so no treatment was planned [1, 3].

Subepidermal Idiopathic calcinosis cutis is most commonly localized to one area, only few previous cases of unusually diffuse calcinosis cutis have been described in the literature, and here we report a new one.

From this point, more future studies should be made about the diffuse form of the disease, we believe that studies could lead to a new classification of Subepidermal Idiopathic

calcinosis cutis: localized and widespread form, which in turn helps physicians to make a more accurate diagnosis.

FUNDING

None.

ETHICAL APPROVAL

No approval was required.

PATIENT CONSENT

Written informed consent was obtained from the patient's legal guardian for publication of this case report and any accompanying images.

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