

Idiopathic Calcinosis cutis - A rare case report

Dr. Satish Dalal¹, Dr. Sandeep², Dr. Deepika³, Dr. Vipul⁴,
Dr. Amandeep Saharan⁵, Dr. Vundavalli Sattibabu⁶

¹M.S. Professor, Department of General Surgery, Pt. B. D. Sharma Postgraduate Institute of Medical Sciences (P.G.I.M.S.), Rohtak-124001, Haryana, INDIA

²M.S. Senior Resident, Department of General Surgery, Pt. B. D. Sharma Postgraduate Institute of Medical Sciences (P.G.I.M.S.), Rohtak-124001, Haryana, INDIA

³M.D. Senior Resident, Department of Anaesthesia, Pt. B. D. Sharma Postgraduate Institute of Medical Sciences (P.G.I.M.S.), Rohtak-124001, Haryana, INDIA

^{4,5,6}Junior Resident, Department of General Surgery, Pt. B. D. Sharma Postgraduate Institute of Medical Sciences (P.G.I.M.S.), Rohtak-124001, Haryana, INDIA

ABSTRACT

Calcinosis cutis is characterized by the deposit of calcium salts (hydroxyapatite or calcium phosphate) in the skin and subcutaneous tissue. It has been classified into four types: metastatic, dystrophic, iatrogenic and idiopathic types. Idiopathic calcinosis occurs in the absence of any tissue or metabolic abnormalities and is a diagnosis of exclusion. Distinct anatomical areas, vulva, scrotum, penis, neck, and breast have been reported to develop idiopathic calcinosis cutis. We present a rare case of 27-year-old male with ill-defined nodules over both the buttocks. He was keen on surgical removal of the lesions. The lesions were removed and sent for histopathological examination. There was neither a history of trauma nor any inflammatory process prior to the development of lesions and no systemic abnormality was detected.

INTRODUCTION

“Calcinosis cutis” is characterized by the deposit of calcium salts (hydroxyapatite or calcium phosphate) in the skin and subcutaneous tissue.¹ Virchow described calcinosis cutis in 1855. When confined to a small area of the extremities and joints, it is described as calcinosis circumscripta. When diffuse, it is referred to as calcinosis universalis and affects the subcutaneous and fibrous structures of the muscles and tendons.²

It has been classified into four types: metastatic, dystrophic, iatrogenic and idiopathic types.³ The metastatic type occurs in normal tissue as the result of increased serum calcium and/or phosphorus levels and a calcium x phosphate product ≥ 70 mg/dL. Iatrogenic calcinosis occurs as the result of intravenous leakage of calcium gluconate or as calcium salt deposits in the skin following electromyography or electroencephalography. The dystrophic form of this condition occurs when calcium salts are deposited following damage or tissue devitalisation in the skin, subcutaneous tissue, muscles or tendons in the presence of normal calcium and phosphorus metabolism and no visceral involvement. In all cases of calcinosis cutis, insoluble compounds of calcium are deposited within the subcutaneous tissue due to local/systemic factors. These calcium salts consist primarily of hydroxy apatite crystals or amorphous calcium phosphate. Metabolic and physical factors are pivotal in development of calcinosis.⁴

Idiopathic Calcinosis cutis is caused by no underlying disease and a diagnosis of exclusion. Its chemical composition has recently been evaluated by vibrational microspectroscopy and according to the results of this study, type B carbonated apatite and beta carotene interspersing subcutaneous tissue.⁴ Many case reports of idiopathic calcinosis cutis of penis, scrotum and vulva have been reported worldwide. Although, surgical excision is the main choice of treatment, electrodesiccation or CO₂ laser ablation may be performed for small, multiple lesions.⁴

CASE REPORT

A 27 years old male presented to the surgery OPD with multiple firm, nontender, nodules measuring approximately 2x1 cms over both the buttocks without any skin ulceration. They had slowly grown in size during the last 3 years and then remained static. Complete hemogram showed following results; Hb 13.5 g/dL, WBC 5.2 x 10³/mL, PLT 270 x 10³/mL, and sedimentation rate 9 mm. Other investigations revealed within normal limits i.e. serum calcium levels of 9.5 mmol/dL, magnesium levels of 2.4 mg/dL and phosphorus levels of 2.9 mmol/dL. Blood sugar, uric acid,

electrolytes, liver function and kidney function tests and routine urine examination were normal. Antinuclear cytoplasmic antibodies, ANF, alkaline phosphatase and gamma GT were also within normal limits. Anti-DNA, anti-RNP, anti-Jo1 and Anti-Mi were nonreactive. The masses were excised under local anaesthesia and there was no recurrence at 1 year follow up. Skin biopsy specimen demonstrated dermal calcification with a diagnosis of idiopathic calcification. Any special staining except H and E, was not applied.

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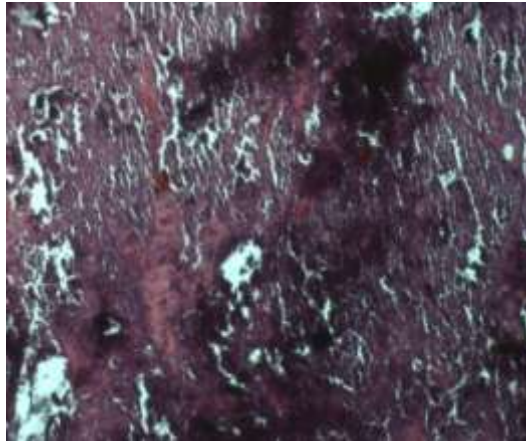


Figure 1: showing calcinosis cutis

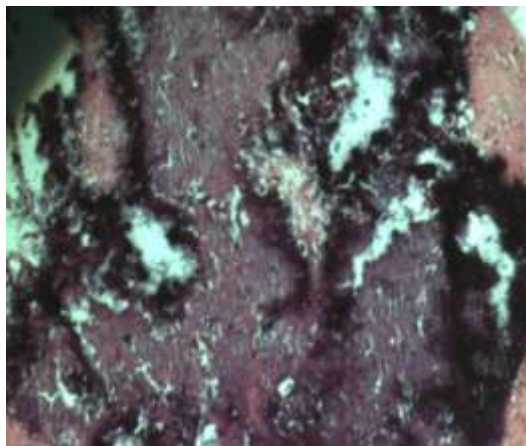


Figure 2: showing calcinosis cutis

DISCUSSION

Distinct anatomical areas, vulva, scrotum, penis, neck, and breast have been reported to develop Idiopathic calcinosis cutis.⁵ The main observation is absence of any abnormality in phosphorus or calcium metabolism and to make a diagnosis of idiopathic calcinosis cutis, one should also workup to rule out collagen vascular disease, as dystrophic calcinosis cutis is caused usually by connective tissue disease. The onset of swelling may be in childhood or adulthood. Nodular subcutaneous forms of calcinosis cutis with congenital origin have been reported.⁶ This type of nodular idiopathic calcinosis cutis was also seen at infrapatellar region in an athlete of marathon runner, showing that the occurrence of calcinosis cutis may be related with chronic repeated physical activity.⁷

When reviewing the literature, it can be seen that the definition of idiopathic calcinosis cutis with several different forms and association with different clinical statements makes it an ill defined clinical entity. Strong evidences is needed for a clear description of etiopathologic mechanism. An interesting clinical form of idiopathic calcinosis cutis is milia like form of which two thirds of the reported cases have been associated with Down syndrome.⁸

At now, histopathological detection of large calcium deposits within subcutaneous tissue with no other pathology is enough to make the diagnosis of idiopathic calcinosis cutis. Histopathologic findings include calcific deposits beneath epidermis without any inflammatory infiltration. There may be histopathologically vascular changes lead to fibrosis, intimal hyperplasia, and occasionally thrombosis of the vessel in severe cases of calciphylaxis. Von Kossa and Alizain

stain is used to confirm the presence of calcium encrusting degenerated collagen bundles throughout the reticular dermis.

Surgical excision is the main choice of treatment, electrodesiccation or CO2 laser ablation may be performed for small, multiple lesions.⁹

CONCLUSION

Idiopathic calcinosis cutis is a rare diagnosis and should be kept in mind in subcutaneous swellings.

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