ORIGINAL RESEARCH

Idiopathic calcinosis cutis: A case series

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ABSTRACT
Calcinosis cutis is an abnormal deposition of calcium in the skin and subcutis. Skin is not a site for collection of calcium but always a pathological phenomenon. Calcinosis cutis can involve any part of the skin. In this study we are presenting 4 cases of calcinosis cutis which were clinically misdiagnosed and sites affected were right iliac fossa and scrotum.
Key words: Calcinosis cutis, calcium deposition, Idiopathic calcinosiscutis

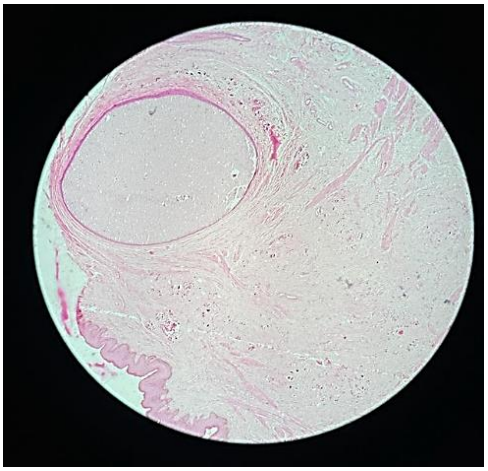
INTRODUCTION

Calcinosis cutis was first described by Virchow in 1855. The disorder is characterized by the pathological deposition of calcium in the skin and subcutaneous tissue. There are five subtypes: dystrophic calcification, metastatic calcification, idiopathic calcification, iatrogenic calcification, and calciphylaxis[1].The typical sites involved are fingers, forearms, and elbows. It may present as a painful swelling[2].Idiopathic calcinosis cutis occurs in the absence of known tissue injury or systemic metabolic defect and is present in the genital skin of vulva, penis, or scrotum. 7 In all our cases serum calcium levels were normal. It can be treated with pharmacotherapy, surgical or combined therapy based on the clinical characteristics[4].

CASE SERIES

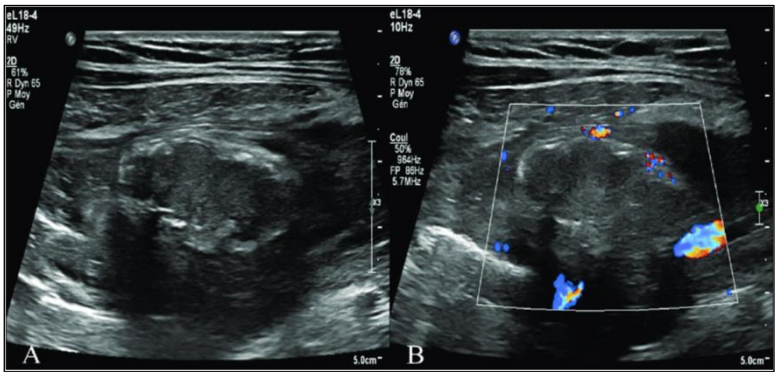
CASE 1

A 13 year old male child presented in skin OPD with history of appearance of verrucous papules three in number (one on thumb, one on right iliac fossa and one on dorsum of right foot) since 3 years. Patient was relatively asymptomatic. Clinical diagnosis was made as tuberculous verruca cutis. Excisional biopsy was done from the lesion on the right hand and sent to histopathology lab. Received greyish brown two tissue pieces firm to hard in consistency. One conical shaped skin covered tissue piece shows ulceration measuring 0.7x0.6 cm. Second wedge shaped skin covered soft tissue piece shows ulceration measuring 1x1x0.7 cm. On cutting intra lesion gritty, calcific deposits measuring 0.2 cm is seen. Microscopic examination showed keratinized stratified squamous epithelium revealing marked hyperkeratosis, parakeratosis and irregular acanthosis. Stratum corneum reveals deposits of calcium and abscess formation. Throughout dermis exhibits presence of calcium deposits along with foreign body type of giant cell reaction and focal necrosis.
PAS stain-No spores/hyphae seen.
AFB stain for TB bacilli seen.
Histopathologicalfeatures are suggestive of Calcinosis cutis with superadded infection.



CASE 2

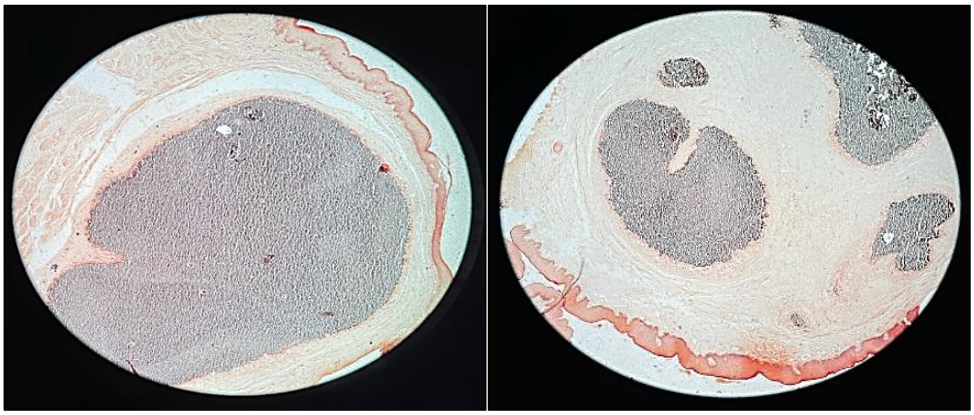
A 65 year female presented in skin OPD with history of well-defined nodular, firm swelling on right iliac region for the last 1 year and is associated with mild pain. Clinical diagnosis was made as lipoma. USG was done for this case which revealed superficial intraperitoneal mass in right iliac fossa, non-compressible, hyperechoic, surrounded by subtle hypoechoic line. Skin punch biopsy 5mm was done from right iliac region and sent to histopathology lab. Received a single skin covered tissue piece measuring 0.8x0.4x0.3 cm.Microscopic examination showed keratinized stratified squamous epithelium with increased melanin pigment in the basal layer. Papillary and mid dermis is unremarkable. Reticular dermis and subcutaneous tissue shows large masses of calcium deposits along with histiocytes and foreign body type of giant cell reaction. Histopathological features are consistent with Calcinosis cutis.



A) Superficial intraperitoneal mass in right iliac fossa, hyperechoic, surrounded by subtle hypoechoic line. B) without internal vascularity.

CASE 3

A 27 year male presented in surgery OPD with history of multiple cysts on scrotum. Clinical diagnosis was given as sebaceous cyst. Excisional biopsy was done and sent to histopathology lab. Received multiple skin covered nodular tissue bits together measuring 3x2.5x0.6 cm. Central areas of every nodule is chalky white in color with size ranging from 1-4 mm. Microscopic examination showed keratinized stratified squamous epithelium revealing hyperkeratosis, acanthosis and increased melanin pigment in the basal layer. Dermis shows numerous variable sized nodules having amorphous, homogenous to granular, globular basophilic material with foreign body giant cell reaction at the periphery, along with few lymphocytes and histiocytes. Histopathological features are consistent with Idiopathic Calcinosis cutis- scrotum. Von kossa was positive for calcium.



CASE 4

A 75 year female presented in surgery OPD with history of swelling in right iliac region which is slowly progressive in nature, non-tender and shows no discharge. Overlying skin is normal and not fixed to underlying structure. Sebaceous cyst was given as clinical diagnosis. Excisional biopsy was done and sent to histopathology lab. Received skin covered single grey brown firm tissue piece measuring 5x3x2 cm. External surface shows haemorrhagic areas at places. On cut section gritty, calcific deposits seen. Microscopic examination shows keratinized stratified squamous epithelium revealing hyperkeratosis, follicular plugging and mild acanthosis. Dermisfibrocollagenous tissue and presence of skin appendages. In addition variable sized lobules of calcification with foreign body giant cell reaction is seen. Histopathological features are suggestive of Calcinosis cutis.

DISCUSSION

Calcinosis cutis is broadly divided into 2 categories i.e.calcinosis cutis circumscripta and calcinosis cutis universalis. Calcinosis cutis circumscripta is isolated to small areas on extremity or joint, while diffuse involvement of skin, subcutaneous tissue and muscles is known as calcinosis cutis universalis[5].Calcification of the skin and subcutaneous tissue is known to occur in a variety of disorders and may be classified as dystrophic, metastatic, idiopathic or iatrogenic calcification, and calciphylaxis. Dystrophic calcification results from local tissue damage or abnormalities, such as alterations in the collagenous, elastic, or subcutaneous fat tissue. This form shows serum calcium and phosphate levels within normal ranges[6].Metastatic calcification is related to abnormal calcium or phosphate metabolism and is generally associated with hypercalcaemia and/or hyperphosphatemia. Iatrogenic calcinosis cutis arises as complication of a therapeutic or diagnostic procedure. It has been associated with subcutaneous injection of calcium-containing heparins, extravasation of calcium gluconate and use of calcium containing electrode compounds for electromyographic or electroencephalographic examination. Idiopathic calcinosis cutis occurs in the absence of known tissue injury or systemic metabolic defect[7].Idiopathic calcinosis maybe present in skin of vulva, penis, or scrotum. Idiopathic calcinosis cutis of the scrotum (ICCS), also called idiopathic scrotal calcinosis, is an uncommon entity and was first described by Lewinski in 1883. It is benign, mostly asymptomatic condition. patients generally present during the third to fourth decade of life. The disease presents as slow-growing, variable sized single to multiple yellowish nodules on the scrotum[8].Calcification of small blood vessels of the dermis or subcutaneous fat is termed calciphylaxis. It is often the result of changes in calcium/phosphate metabolism as is seen in hyperparathyroidism, and end stage renal disease[9].Calciphylaxis is associated with severe pain, an increased risk of infection, and a high mortality rate[1].Therapeutic options include diltiazem, warfarin, bisphosphonates, aluminum hydroxide, probenecid, intralesional steroids, sodium thiosulphate and surgical excision[10].Early diagnosis and treatment of this disabling entity is of paramount importance to improve the quality of life of these patients[5].

CONCLUSION

In all our cases calcium and phosphorous levels were normal.

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CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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