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Pathology

Idiopathic Calcinosis Cutis with Ossification in Rare Site: A Rare Case Report

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Abstract **Case Report**

We received a mass for histopathological examination from the upper right thigh in a 55 years old female patient presented with a hard mass over the thigh for the last 10 years. It was Painless Slow Growing swelling with no significant history of connective tissue disorder. Laboratory parameters including serum calcium and phosphorus level were within normal limits. Grossly mass was firm, gray white nodular tissue measuring 3.5x2x2cm. Greyish white areas were seen on the cut section. On microscopic examination, the calcified mass showed histopathological features of basophilic, finely granular and amorphous deposits in the subcutaneous tissue and dermis.There were presence of few foreign body type multinucleated giant cells. Some areas showed mature osteoids. We diagnosed the case as idiopathic calcinosis cutis with focal ossification by clinical, biochemical and histopathological examination. Idiopathic calcinosis cutis is extremely rare and the upper part of thigh is a very rare site for idiopathic calcinosis cutis. Diagnosis of this is very challenging because other soft tissue neoplasms mimic the same features. We confirmed the case by a special stain, von kossa which confirms the presence of calcium salts.

Keywords: Idiopathic calcinosis cutis, serum calcium, von kossa, rare site.

INTRODUCTION:

Calcinosis cutis is a rare pathological entity characterized by the deposition of calcium salts in the skin and subcutaneous tissue. It is broadly classified into dystrophic, metastatic, idiopathic, iatrogenic and calciphylaxis-related types, based on underlying etiopathogenesis. Dystrophic calcinosis cutis, the most common subtype, occurs in setting of normal serum calcium and phosphate levels and usually with prior tissue damage, inflammation, trauma, connective tissue disorders. Idiopathic calcinosis cutis causes subepidermal calcified nodule. Pathogenesis of idiopathic calcinosis cutis is generation and calcification of stoma, degranulation of mast cells and dystrophic calcification following dermal damage from unknown cause. Idiopathic calcinosis cutis may be a subepidermal calcified nodule, commonly occurrs in head- neck region. Scrotal type occurs in scrotum in case of male child. Tumoral calcinosis type usually is around the joints. The back of upper part of thigh is an uncommon site for idiopathic calcinosis cutis and lesion in this location often pose a diagnostic challenge due to their resemblance to other calcified or ossified soft tissue lesions such as tumoral calcinosis, myositis ossificans, benign and malignant neoplasms. The presence of ossification within calcinosis cutis is exceptionally rare and represents heterotopic bone formation secondary to long standing calcium deposition [1]. Calcinosis cutis shows no uniform gender predilection overall; however, dystrophic calcinosis demonstrates female preponderance due to association with connective tissue disease while idiopathic calcinosis cutis commonly affects children and young adults with no consistent sex bias [2].

CASE PRESENTATION:

We received a specimen of mass over the upper right thigh with the history of a female patient aged 55 years presented with slowly growing, painless nodules over the upper right thigh for a duration of 1.5 years.

There was no history of trauma, injection at the site, infection. There was no similar kind of lesion in other parts of the body, no known co-morbidities, no history of connective tissue disorders and no family history.

On clinical examination, a firm, well circumscribed subcutaneous nodule measuring 4x3cm was noted over the right side of the upper part of thigh.There was no pain or tenderness associated with it.The overlying skin was unremarkable. There was no discharge or ulceration associated with it.

On routine laboratory investigation, all blood parameters were normal. Serum calcium and phosphate levels were also within normal limits.

The lesion was surgically excised and submitted for histopathological examination.

Gross examination revealed a firm, gray white nodular tissue measuring 3.5x2x2cm. On the cut section, greyish white areas were seen.

On microscopic examination, the calcified mass showed histopathological features of basophilic, finely granular and amorphous deposits in the subcutaneous tissue and dermis.There were presence of few foreign body type multinucleated giant cells. Some areas showed mature osteoids. We diagnosed the case as idiopathic calcinosis cutis with focal ossification by clinical, biochemical and histopathological examination. We confirmed the case by a special stain, von kossa which confirms the presence of calcium salts.

Images:

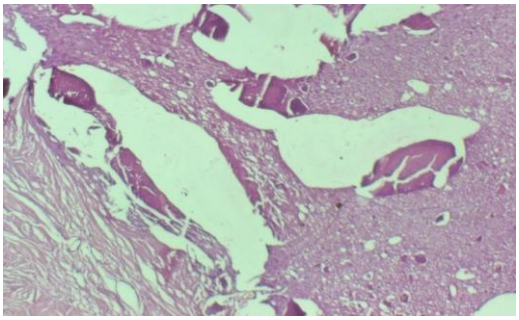

Figure 1: Idiopathic calcinosis cutis; amorphous granular calcium deposition along with multinucleated foreign body giant cells; H&E stain X 40

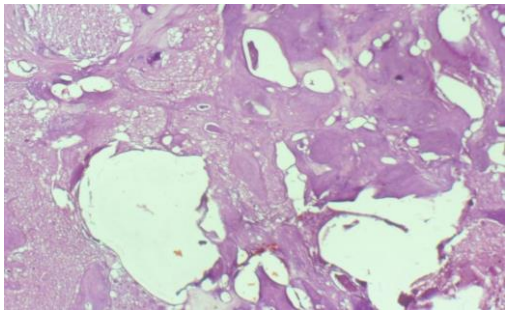

Figure 2: Idiopathic calcinosis cutis; amorphous granular calcium deposition along with focal osteoid formation; H&E stain X 40

DISCUSSION:

Idiopathic calcinosis cutis is a rare entity characterized by calcium salt deposition in the dermis and subcutaneous tissue without underlying metabolic derangement or tissue injury. From a pathologic perspective, diagnosis primarily on histomorphological features, as clinical features are nonspecific.

Histologically, lesions show irregular basophilic, amorphous calcium deposits, commonly located in the dermis or subcutis, often surrounded by a foreign body giant cell reaction. Long standing lesions may demonstrate focal ossification, indicating chronicity.The overlying epidermis is usually unremarkable unless secondary ulceration or transepidermal elimination is present. Special stains such as von kossa can confirm the presence of calcium salts. In our case diagnosis of idiopathic calcinosis cutis with focal ossification is challenging regarding site as well as extremely rare disease. Diagnostic features of idiopathic calcinosis may be similar with dystrophic calcification, metastatic calcification, calcified epidermoid cyst, and pilomatricoma and osteoma cutis. Correlation with normal serum calcium and phosphate levels and absence of associated systemic disease is essential to establish the idiopathic nature of the lesion.

We reported this rare case of calcinosis cutis with focal ossification involving the thigh to emphasize its histopathological features, rarity of presentation and the importance of considering the entity in the differential diagnosis of calcified cutaneous lesions at unusual anatomical sites. Calcinosis is often associated with autoimmune connective tissue diseases such as scleroderma [1]. Very limited cases of idiopathic calcinosis cutis with focal ossification have been reported. Exact incidence is difficult to report as this entity is usually described in the form of individual case reports and limited numbers of case series are available in the literature. Tian-yu E *et al.,* has reported a case of idiopathic calcinosis cutis where a 51 years old male presented with long standing hard nodules in the buttock with pruritus and no other comorbidities [3]. Tejashwini Kotian *et al.,* reported a case series of 6 cases of idiopathic calcinosis cutis where patient's age ranged from 25 to71years, 3 males and 3 females and lesions were on the scrotum, scalp, axilla, iliac region with no other comorbidities[4]. Choudhari Nidhi *et al.,* also reported a case of idiopathic calcinosis cutis where a 19-year-old female presented with a hard lesion in her left toe with no other associated disorder [5].

CONCLUSION:

This case highlights the importance of histopathological evaluation in diagnosing idiopathic calcinosis cutis with focal ossification a rare and often clinically misleading condition specially when there are no other associated abnormalities and very rare site. Here in India and especially in West Bengal we get this case

on a very rare occasion. Early recognition and complete excision ensure favorable outcomes and prevent unnecessary investigations.

Declaration by Authors:

Consent: Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Conflict of Interest: The authors declare no conflict of interest.

Source of Funding: None.

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