

CASE REPORT

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Received: 28.08.2022

Accepted: 09.09.2022

Published: 25.12.2022

Citation: Shilpa MD, Harendrakumar ML, Dave P. Idiopathic Calcinosis Cutis Masquerading as Malignancy - The Mystery of Two Cases Revealed on Cytology. J Clin Biomed Sci 2022; 12(4): 147-150. <https://doi.org/10.58739/jcbs/v12i4.115>

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Funding: None

Competing Interests: None

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Published By Sri Devaraj Urs Academy of Higher Education, Kolar, Karnataka

ISSN

Print: 2231-4180

Electronic: 2319-2453



Idiopathic Calcinosis Cutis Masquerading as Malignancy - The Mystery of Two Cases Revealed on Cytology

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Abstract

Calcinosis cutis also commonly called as Calciphylaxis is one of the uncommon conditions characterized by precipitation and deposition of phosphate and calcium salts in the dermis and subcutaneous tissues. We report two different cases clinical diagnosed as malignancy and final diagnosis was made on FNAC which correlated with the histopathology. A 45-year-old women came with a compliant of swelling over lateral aspect of thigh on left side since 6 months. Provisional diagnosis of liposarcoma was made clinically and on FNAC given as Calcinosis cutis which correlated with histopathology. A 62-year-old women came with compliant of swelling over lateral aspect of thigh on right side since 10 months associated with pain. Provisional diagnosis of soft tissue sarcoma was made clinically and advised for FNAC clinically and on FNAC given as Calcinosis cutis which correlated with histopathology. Calcinosis cutis is a condition in which there is organized and localized deposition of calcium in the skin. The evolution of the lesions depends on the aetiology of the calcification. The pathogenesis still remains unclear, but several theories have been suggested for this. Calcinosis cutis is a potential mimic of a neoplastic lesion. FNAC is a simple, rapid and reliable technique and which helps in diagnosis of such lesions.

Keywords: Mimicker; Fine needle aspiration cytology; Diagnosis

Introduction

Calcinosis cutis also commonly called as calciphylaxis is one of the uncommon

conditions of systemic calcinosis characterized by precipitation and deposition of calcium and phosphate salts in the dermis and subcutaneous tissues.¹

The pathogenesis is not completely understood. We report two different cases clinical diagnosed as malignancy and final diagnosis was made on FNAC which correlated with the histopathology.

Case Report 1

A 45-year-old women came with compliant of swelling over lateral aspect of thigh on left side since 6 months. Initially the swelling was small later gradually increased to the present size of 5X3 cms. It was associated with pain. On examination an irregular swelling of 5X3cms size noted near the lateral anterior superior iliac spine which was hard, tender and non-mobile. Provisional diagnosis of liposarcoma was made clinically and advised for FNAC. The FNAC was performed using 22-gauge needle. On multiple aspirations 2 ml of chalky white material was aspirated. Air dried smears were stained with May Grenwald Giemsa stain and methanol-fixed smears were stained with Hematoxylin & Eosin (H & E) and Pap stain. On microscopy o smears showed flakes of amorphous granular material which stained basophilic on H&E (Figure 1) and deep blue on Giemsa stain along with few giant cells. Diagnosis was given as Calcinosis cutis. Other investigations like Serum calcium and serum phosphate was done and were within normal limits. Later the lesion was excised and sent for histopathological examination. On histopathological examination it showed large basophilic deposits of calcium within the cystic cavities in the dermis (Figure 2) with giant cell reaction at places was noted and final diagnosis was given as Calcinosis cutis.

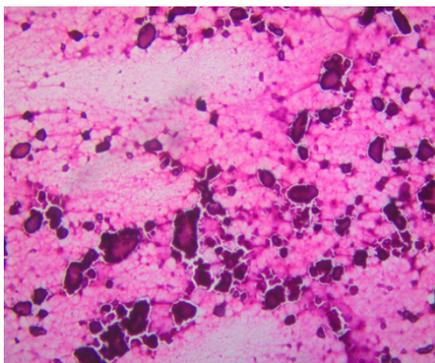


Fig 1. FNAC H&E smear (100X) showing flakes of amorphous material

Case Report 2

A 62-year-old women came with compliant of swelling over lateral aspect of thigh on right side since 6 months associated with pain. On examination an irregular swelling measuring 8X5cm in size noted near right iliac crest which was hard, tender and non-mobile. Provisional diagnosis of soft tissue

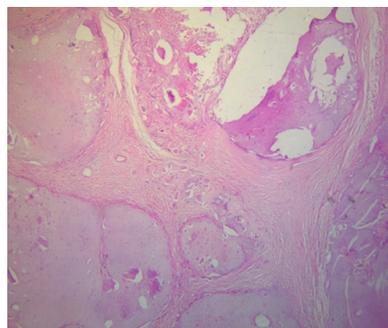


Fig 2. Histopathology H&E section (100X) showing cystic cavities filled with large basophilic deposits

sarcoma was made clinically and advised for FNAC. The FNAC was performed using 22-gauge needles. On multiple aspirations 5 ml of chalky white material was aspirated. Air dried smears were stained with May Grenwald Giemsa stain and methanol-fixed smears were stained with Pap stain and Hematoxylin & Eosin (H & E). Microscopy of the smears showed flakes of amorphous granular material that stained on Pap stain (Figure 3) and deep blue on Giemsa stain along with histiocytes and lymphocytes. Diagnosis was given as Calcinosis cutis. Further biochemical investigations like serum alkaline phosphatase, calcium and phosphorus levels were done and all were in normal limits. Later surgical excision was done and it was sent for histopathological examination. On microscopy large nests of basophilic calcific material separated by fibrous septa in the dermis along with giant cell reaction was noted (Figure 4). The overlying epidermis was normal. With these findings final diagnosis of Calcinosis cutis was given.

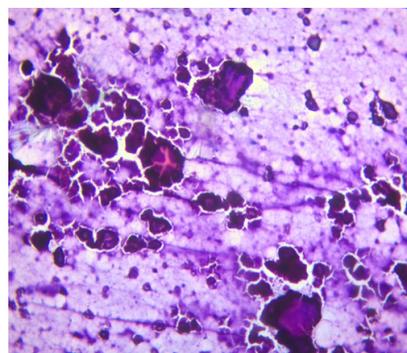


Fig 3. FNAC Pap smear (400X) showing flakes of amorphous material

Discussion

Calcinosis cutis is a condition with localized and organized deposition of calcium in the skin. It was first described by

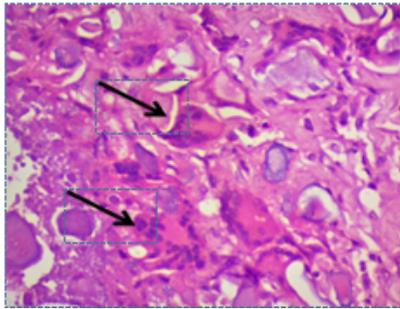


Fig 4. Histopathology H&E (400X) showing giant cell reaction (arrow)

Rudolf Virchow in 1855.² It is divided into four major types based on etiology and they are metastatic, dystrophic, idiopathic and iatrogenic. Other rare types are tumoral calcinosis, calcinosis cutis universalis, calcinosis cutis circumscripta and transplant associated calcinosis cutis.³ Most lesions are asymptomatic and develop gradually.⁴

The sites most commonly affected are the superior and lateral shoulder, posterior elbows, and lateral hip and gluteal regions. Rarely has it been noted in the hands, feet, spine, temporo mandibular joint & knee.¹

The development of these lesions depends on the causes for the calcification. The patients with history of chronic renal failure with abnormal serum and phosphate levels usually develop metastatic type Calcinosis. The patients with history of any trauma or an underlying disease with normal serum calcium and phosphate levels most commonly have dystrophic type calcinosis. The patients with iatrogenic calcinosis cutis will generally provide history of recent hospitalization.⁵

The cases with idiopathic calcinosis cutis will not associated with any underlying disease, previous trauma and recent hospitalization like in our both the cases. The pathogenesis of idiopathic calcinosis cutis is still unknown.⁴

In all the cases of Calcinosis cutis due to local and systemic factors the insoluble compounds of calcium get deposited. These calcium salts include primarily of amorphous calcium phosphate and hydroxyapatite crystals. The pathogenesis for this is still unknown and several theories have been suggested. Hypercalcemia and hyperphosphatemia have been described as the underlying cause in some patients while local trauma has been implicated in a few cases.³

In the present both the cases all the investigations to evaluate abnormal calcium metabolism revealed results within normal limits and the diagnosis made was based on the cytomorphological findings and it was confirmed on histopathological examination.

The sample yield on FNA is chalky white granular material and the following differential diagnosis have to be considered

which includes Epidermal cyst with calcification, Sarcoidosis, Pilomatricoma, calcified fibrous pseudotumor, tuberculosis, osteitis fibrosa cystica and extra skeletal osteosarcoma. Calcified epidermal cyst shows nucleate and anucleate squames^{6,7} whereas Sarcoidosis and Calcified tuberculosis show granulomatous reaction.^{6,8} Pilomatricoma shows mainly ghost cells, basaloid cells and multinucleate giant cells. Abundant hyalinised collagen, fat and neovascular bundles along with calcification are the features of calcified fibrous pseudotumor.^{6,9} Lymphoepithelial lesions show a polymorphous

lymphoid cell population along with histiocytes and calcification. Absence of tumor cells rules out extraskelatal osteosarcoma. The clinical history helps in evaluation of osteitis fibrosa cystica.

Anupama *et al.* studied the cytological features of idiopathic calcinosis cutis of the back and noted dense basophilic amorphous granular material without any evidence of epithelial cells.¹ In both of our cases there was no history of trauma, any underlying disease, hospitalization, previous parenteral therapy or any preceding pathological lesion at the site. Also normal serum calcium and phosphorus levels clearly excluded the possibility of metastatic, dystrophic and iatrogenic causes and hence a final diagnosis of idiopathic calcinosis cutis was made.

The treatment for both large, localized lesions and small calcified lesions is surgical excision, and it also allows for further histopathological examination which is required for the confirmation of diagnosis. Whereas for disseminated and extended calcinosis systemic therapy is used. Various treatment modalities have been tried which includes warfarin, bisphosphonates, minocycline, diltiazem, intralesional steroids, carbon dioxide laser and extracorporeal shock wave lithotripsy and they have been shown to have beneficial effects.² In our both the cases surgical excision was the treatment of choice and both the patients were on follow up and doing well.

After through literature search only very few case reports have been reported till date on FNA cytology of idiopathic calcinosis cutis. Correct cytomorphological diagnosis helps in determining cases requiring surgical rather than medical treatment.

Conclusion

The present cases are highlighted to alert the pathologists and they should consider Calcinosis cutis as the one of the differential diagnosis in any swelling with a chalky white granular aspirate on FNA technique as it is a potential mimic of a neoplastic lesion. FNAC is a simple, rapid and reliable technique and is of great diagnostic investigation especially in soft tissue lesions like calcinosis cutis. FNA features along with the serological and biochemical findings can help in effective patient management.

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