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Case Report

A Rare Association of Calcinosis Cutis with Rheumatoid Arthritis in an Elderly Woman – A Case Report with Cytology and Histopathology Correlation

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Abstract

Calcinosis cutis is a rare disorder noted in the skin and subcutaneous tissue with the presence of insoluble calcium salts. Amongst the various subtypes of calcification, the most commonly found one is dystrophic calcification, which is mostly associated with connective tissue diseases such as dermatomyositis and systemic sclerosis. However, dystrophic calcification associated with rheumatoid arthritis is extremely rare. The condition results in substantial morbidity with pain and limitation of movement with extreme debilitation. We report a rare association of good controlled rheumatoid arthritis with dystrophic calcinosis cutis, in an elderly woman.

Keywords: Calcinosis Cutis; Dystrophic; Rheumatoid Arthritis; Elderly; Hips

Abbreviations

DC: Dystrophic Calcinosis; RA: Rheumatoid Arthritis; DMARDS: Disease-Modifying Antirheumatic Drugs

Introduction

Calcinosis cutis is a rare disorder of skin and subcutaneous tissue, wherein insoluble calcium salts are deposited. Various subtypes of calcinosis cutis are described in literature, such as dystrophic calcification, metastatic calcification, idiopathic, iatrogenic, and calciphylaxis [1]. Dystrophic calcinosis (DC) is the most common subtype noted, occurring in devitalized tissues in the presence of normal serum calcium and phosphate levels. It is most frequently associated with connective tissue diseases such as dermatomyositis and systemic sclerosis [2]. However, there are very few reports on dystrophic calcification associated with rheumatoid arthritis (RA) [2]. The condition results in substantial morbidity with pain and limitation of movement with extreme debilitation. We hereby report the case of an elderly woman with RA, controlled with treatment, who developed calcinosis cutis in her bilateral hips.

Case details

A 69- year-old woman presented with complaints of swelling involving bilateral hips for a duration of two years. The swellings were associated with itching and later she developed ulceration over the lesions. She gives a history of gradual increase in size of the swellings. No association with pain was reported. Patient is a known case of rheumatoid arthritis for the past 10 years. She had regular follow-up and had been noted to be in remission. Her rheumatoid arthritis was controlled on disease-modifying antirheumatic drugs (DMARDS) including leflunomide and hydroxychloroquine. The patient did not give a history of any new complaints, suggestive of disease progression. She did not have any other known concurrent diseases such as diabetes mellitus or hypertension. She did not smoke or consume alcohol. She also had a negative family history with regards to connective tissue diseases.

Investigations

The routine blood investigations revealed that the patient had anemia, with hemoglobin of 10.4 gram/deciliter, and normal red blood cell indices. Her total leucocyte count was towards the lower limit of normal, 4600 cells/ cubic millimeter and a differential count with 60% neutrophils, 32% lymphocytes and 8% monocytes. Her platelet count was adequate, 2.1 lakh/cubic millimeter. She also was noted to have erythrocyte sedimentation rate of 10 millimeter/hour. Her serum calcium level, upon evaluation turned out to be within normal limits, 9 milligram/deciliter.

On local examination

Multiple hard nodules, firm to hard in consistency with overlying thickened skin noted over bilateral hips. The nodules on left hip were noted over an area of 6 x 4 cm, located 1 cm behind anterior superior iliac spine, while the right hip nodule measured about 4 x 4 cm (Figure 1a and 1b).

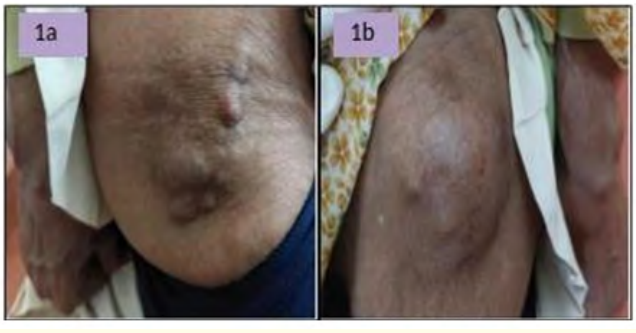

Figure 1: (1a) Photograph of swellings over left hip and (1b) swellings over right hip, while patient is in supine position. Multiple nodules are seen with hyperpigmented overlying skin.

Materials and Methods

Clinical data was collected from the request forms received for the purpose of cytology and histopathology evaluation. Cytology smears, biopsy specimens and the histopathology slides of the same specimen was studied extensively.

Results and Discussion

Results: cytology and histopathology evaluation

The fine needle aspiration cytology (FNAC) was utilized for the evaluation of the bilateral hip lesions of the patient. Aspiration yielded 5 milliliters of chalky white fluidic material (Figure 2a), which did not dissolve either with alcohol or chloroform. Smears were prepared from the aspirated material, demonstrating acellular aspirate with irregular basophilic flakes of amorphous calcium (Figure 2b), suggestive of calcinosis cutis. The nodules were subsequently excised. Excision biopsy of nodules over both left and right hips identified skin and subcutaneous hard tissue with surface nodularity (Figure 3a and 3b). Cut-section revealed multiple chalky white deposits and areas of cystic degeneration. Microscopy showed dermis with basophilic calcified material and a few scattered inflammatory infiltrates (Figure 3c and 3d). Final diagnosis of calcinosis cutis was made.

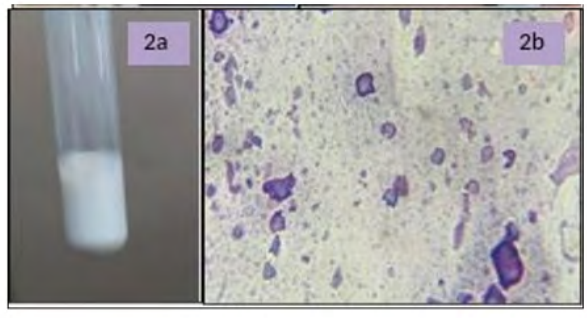

Figure 2: (2a) Photograph of aspirate from the bilateral hip swellings yielded chalky white material; (2b) Photomicrograph showing acellular aspirate with irregular basophilic flakes of amorphous calcium deposits (Papanicolaoustainx400).

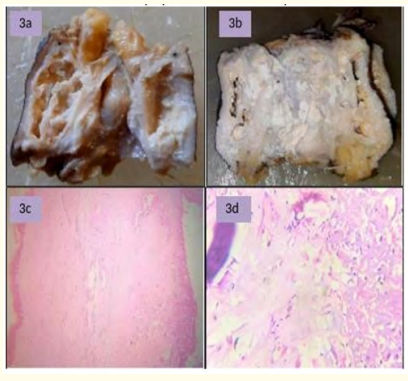

Figure 3: (3a) Photograph of excision biopsy specimen of swellings over left hip and (3b) right hip lesion. Cut section shows overlying skin, dermis with multiple chalky white deposits with areas of cystic degeneration.
(3c) Photomicrograph showing epidermis lined by stratified squamous epithelium and dermis with basophilic calcified material and scattered inflammatory infiltrates (H&E stain x400). (3d) Photomicrograph showing areas of calcificationinthe dermis(H&Estainx400).

Discussion

Out of the five described subtypes of calcinosis cutis, the most common subtype noted is dystrophic calcinosis (DC), which has been found to be in association with systemic diseases such as systemic sclerosis, dermatomyositis, or lupus, that results in tissue damage which creates a nidus for development of calcification [1]. Thus, dystrophic calcification is diagnosed with normal serum calcium and phosphorus levels, unlike metastatic calcification wherein there are altered serum calcium and phosphorus levels.

Idiopathic subtype of calcification includes tumoral calcinosis, and subepidermal calcified nodules. Iatrogenic calcification is a result of administration of agents containing calcium or phosphate. Calciphylaxis is seen in association with chronic renal failure and dialysis, with calcification of small and medium-sized vessels [3].

Autoimmune diseases are usually associated with dystrophic calcifications. The likelihood of developing DC is found to vary from one autoimmune connective tissue disease to another. A review of literature has revealed no specific incidence or frequency of occurrence of calcinosis among the numerous connective tissue disorders.

However, DC is frequently noted in patients with systemic sclerosis, and dermatomyositis. Some reports have concluded that in a span of ten years from the initial diagnosis of the limited systemic sclerosis, 25 to 40% of patients develop calcinosis cutis [4]. Another conclusion was that patients suffering from systemic lupus erythematosus can present with calcification around joints (33%) and in soft tissue (17%) [5].

Rheumatoid arthritis is a debilitating autoimmune disease characterized by both progressive as well as symmetric joint inflammation. The various complications include bone erosion, destruction of cartilage, with progressive loss of function, and concluding in joint deformity [4,6]. Only a few cases of calcinosis cutis associated with rheumatoid arthritis have been reported to date in the literature [2,7,8].

Our patient with a well-controlled rheumatoid arthritis on DMARDS, normal serum calcium levels, presented with an extensive dystrophic calcification. She had minimal limitation of right and left hip joint movement without significant complications like septic arthritis or ulceration as reported in few of the case reports. The patient has no further complications and is on regular follow-up.

Conclusion

Dystrophic calcification occurs in most connective tissue disorders, while limited case reports of the same are noted in association with rheumatoid arthritis. Patients on treatment for rheumatoid arthritis, should be monitored with serum calcium levels and radiology, if they present with cutaneous hard nodules. Regular follow-up should be ensured in all cases of autoimmune connective tissue diseases, with a special focus on development of calcinosis cutis, which by itself can reduce the quality of life of patients.

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Conflict of Interest

Nil.

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