Our Dermatology Online

Case Report

Rare splenic calcifications in a child with cutaneous systemic lupus and a review of the literature

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ABSTRACT

Lupus erythematosus is a complex autoimmune multisystemic disease that can affect the skin and other organs including the spleen where Various anatomical changes can occur but the recognition of spleen calcifications in lupus patients is really a rare event which may be a marker of poor prognosis and renal involvement of lupus, our observation is the 13th described in the literature, and the to our knowledge the first one that concerns a child which is even more unusual.

Keys words: Lupus; Spleen calcifications; Renal involvement; Child

INTRODUCTION

Lupus erythematosus is a complex autoimmune multisystemic disease that can affect the skin and other organs including the spleen [1]. Various changes can occur in the spleen affected by the inflammatory process such as splenomegaly, hyposplenism, infarction, spontaneous rupture, functional asplenia, and periarterial thickening in an "onion skin" pattern [2].

The recognition of calcifications in systemic lupus have been unusually reported concerning other organs like the brain [3], the skin [4], the breast [5,6], the coronary system [7] and submandibular gland [8], in periarticular [9] and in Soft tissues, but their existence in the spleen of patients with systemic lupus is really a rare event since our observation is the 13th described in the literature, and the occurrence of this feature in a child as the case of our patient is even more unusual.

CASE REPORT

A 14-years-old child admitted in our department for scaly erythematous lesions in the face and the hands with acrocyanosis and photosensitivity that started 3 months before; the dermatological examination revealed a maculopapular erythematous patches of the nose and the malar area (butterfly mask),in the palmar surface and interarticular surfaces of the dorsum of the hands and feet with buccal mucosal ulcerations. Cutaneous lupus was confirmed histologically (Figs. 1 and 2).

The results of paraclinical examinations of blood and urine had revealed a systemic lupus with hematological (anemia, thrombocytopenia and leucopenia), immunological (positive antinuclear antibodies with diffuse pattern anti-dsDNA) and renal involvement (hematuria, proteinuria) confirmed histologically as focal lupus nephritis Class III.

The rest of laboratory investigations showed a high erythrocyte sedimentation rate and a hypocomplementemia CH50, C3, C4.

On a chest radiography taking the diaphragmatic domes, we had noticed calcifications in the splenic area with some calcifications in the chest (Figs. 3 and 4). The abdominal ultrasound revealed extensive calcifications over the entire height of the spleen.

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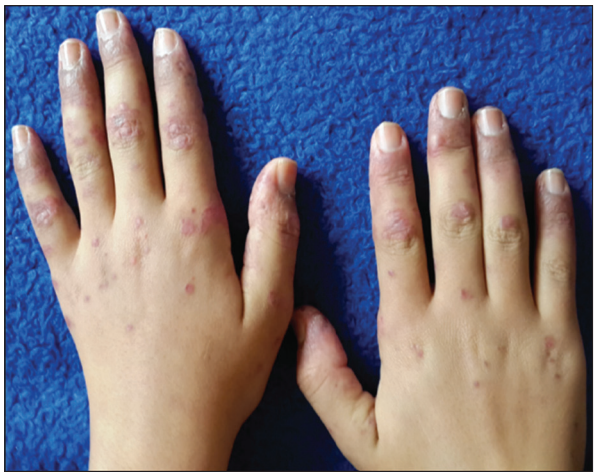

Figure 1: Clinical image showing patches of lupus on the dorsum of the hands.

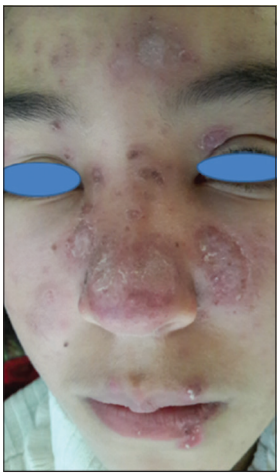

Figure 2: Clinical image showing erythematous patches of the nose and the malar area (butterfly mask).

During all the investigations, the serum calcium, phosphorus, and alkaline phosphatase levels were normal.

The patient was treated initially with bolus corticosteroids during 3 days for the renal lupus and then oral corticosteroids, in association with hydroxychloroquine 200 mg/d, and we transferred the child to the department of pediatrics for appropriate management of his systemic lupus.

DISCUSSION

Splenic calcifications have been reported in connective tissue disorders such as rheumatoid arthritis, systemic sclerosis, infections, sickle cell disease, splenic haemangiomas, and cysts, and in B-cell lymphoma [10].

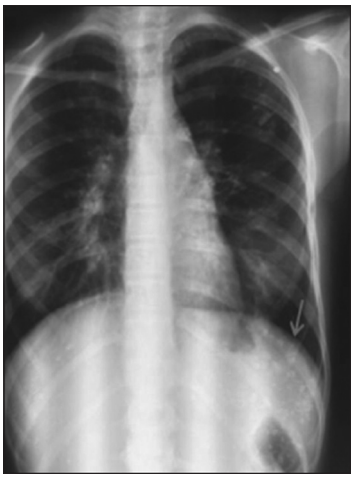

Figure 3: Radiologic image showing spleen calcification.

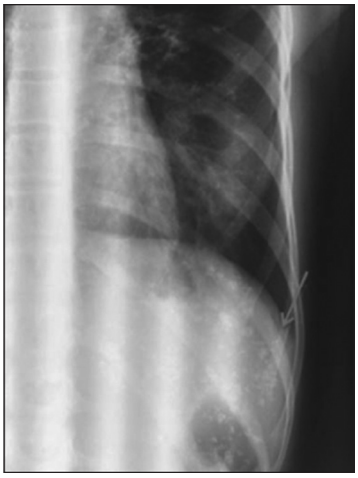

Figure 4: Radiologic image showing spleen calcification (close view).

Yet, these calcifications have barely been described among the anatomical and physiological changes in the spleen of patients having systemic lupus erythematous (SLE).

On the other hand, calcifications have been recognized in other organs in lupus patients, such as the brain[3], the skin [4], the breast [5,6], the coronary system [7] and submandibular gland [8], in periarticular [9] and in Soft tissues in general. The largest study carried out concerning the prevalence of soft tissue calcifications in patients with SLE had been done 15 years ago and had found that in 6 patients, the prevalence of ectopic calcification in SLE was 40% and that the incidence of lupus nephritis and nephrotic syndrome were significantly higher in the calcification positive group than the negative one. This renal involvement was also noticed in a woman having systemic lupus

with spleen calcifications [10] which is the same of our case, so, based on these results, we may deduce that these spleen calcifications could be a marker of poor prognosis and renal involvement especially in children where the lupus is known more dangerous [11].

Another important findings were reported in a patient having lupus erythematosus with diffuse soft tissue and vascular calcification in addition to a chronic kidney disease [12]. In this publication, the authors have explained these calcifications by the systemic disorder of mineral and bone metabolism which is related to chronic kidney disease.

As a result, if we combine all these facts, that the spleen calcifications are significantly related to the renal involvement of lupus and that a kidney disease could cause systemic disorder of mineral and bone metabolism, thus, it may cause calcium deposit in soft tissues like the spleen, but this theory does not explain why our patient whose renal disease was acute -instead of chronic- have developed these calcifications.

Moreover, three mechanisms have been proposed to explain the formation of calcifications as a whole in lupus patients, the first one was suggested by Neuman et al, that these calcifications could be enhanced by high alkaline phosphatase concentration since this alkaline phosphatase can remove the organic phosphate which inhibit the calcium deposit, another theory and the most logical one, has been demonstrated in necrotic tissue by Moss and Urist who proposed that the calcifications may be precipitated by chronic inflammation and tissue necrosis, injuries and ulcers, this explanation could explain calcifications in a spleen affected by the inflammatory process. Another Hypothesis for soft tissue calcifications is a pressure phenomenon producing ischaemia which was reported by Powell et al in 1974. While the exact significance of diffuse splenic calcification is still unknown, this unique radiologic finding may be a result of the disease process itself [13,14].

Furthermore, after a review of the literature from 1947 to 2015, 12 observations of these spleen calcifications were reported [1,13,14]. With a pathologic confirmation in an autopsy of the spleen in one patient [15], our observation is the 13th. The age of these patients ranged from 24 to 73 years old in contrary with the age of our patient (14 years old), this fact increase the originality of our case because as we already noticed, it may have a prognosis signification. Therefore, it may be important to choose invasive therapeutic options from the beginning in patients having these calcifications especially in children whose lupus is usually more severe [11].

CONCLUSION

The originality of our case is the occurrence of spleen calcifications in a child having cutaneous and systemic lupus with renal involvement which may confirm the theory that these calcifications have a prognosis signification.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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