Impressive Subcutaneous Calcifications in Systemic Lupus Erythematosus

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ABSTRACT

Dystrophic calcinosis cutis was commonly described in long-term dermatomyositis or systemic sclerosis, being rarely reported in other connective tissue diseases.

We report the case of a 65-years old woman with an only 5-years history of systemic lupus erythematosus, who presents with multiple, impressive subcutaneous calcified masses and biological normal serum calcium and phosphate levels.

Keywords: Calcinosis cutis, subcutaneous calcifications

CASE REPORT

A 65-year-old woman was referred to our department for polyarthralgia and functional impotence for the articulations of elbows, knees and ankles. Five years ago she was diagnosed with systemic lupus erythematosus (SLE), characterized by cutaneous, hematological and vasculitis involvements. Immunologically, she presented anti-dsDNA antibodies and anti-Ro antibodies.

The clinical examination showed small nodules in the fingers and multiple, indurate, various sized subcutaneous deposits in elbows, knees, thighs and gluteal muscles with a preferential periarticular distribution. Also, a ulcerate lesion in the gluteal muscles region, that revealed a chalky material.

Biologically, the serum calcium and phosphate levels were normal. The radiological examination showed large, impressive calcium deposits in elbows (Figure 1), knees, gluteal muscles region and thighs (Figure 2).

DIFFERENTIAL DIAGNOSES OF SUBCUTANEOUS CALCIFICATIONS

Calcinosis cutis (CC) is an entity characterized by the presence of insoluble calcium salts in the skin and subcutaneous tissue (1). Boulman et al. reweaving older classifications of CC proposed a characterization taking into account calcium/phosphorus levels and the background disease associated with the presence of CC. They divided the subcutaneous calcifications into five groups: metastatic calcification, tumoral calcification, dystrophic...
calcification, idiopathic calcification and calciphylaxis (2). Starting from this classification, Gutiérrez and Wetter spoke also about iatrogenic calcinosis as a separate entity and included tumoral calcinosis, a rare familial disorder into idiopathic calcification group and calciphylaxis, syndrome of vascular calcification into metastatic calcification group (3).

In Table 1 we tried to summarize the main etiologies that participate to the differential diagnosis at a patient with CC.

FINAL CLINICAL COMMENTS

CC, a clinical feature associated with CTD, was rarely reported at patients with SLE; until 2010 being only 36 such cases reported in English-language medical published work (4). In a retrospective study involving 78 patients with CC, Balin et al. found only 2 patients with SLE. On the contrary, CC was described in 23 patients with dermatomyositis (DM) and 21 patients with systemic sclerosis (SS) from the same series (5).

CC typically develops in patients with long-standing SLE (6) and might be an incidental radiologic finding because patients with CC and SLE tend to be asymptomatic (7). Local trauma or local ischemia might be involved as precipitating factors (3), the periarticular areas being most often involved (8).

In this paper we describe a rare association for CC, in a patient apparently without a long-standing CTD (only 5 years from the diagnosis of SLE), but with spectacular radiological images of massive calcium subcutaneous deposits (calcinosi cutis universalis).

The efficacy of different medical treatment for CC, like Warfarin, Bisphosphonates, Mynocicline, Ceftriaxone, Diltiazem, Aluminium hydroxide, Probenecid or Intravenous immunoglobulin, was reported only in single cases or small series, no treatment being generally accepted as standard therapy (9).

TABLE 1. Classification of calcinosis cutis.

<table>
<thead>
<tr>
<th>Category</th>
<th>Etiologies</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dystrophic (normal calcium or phosphorus)</td>
<td>Connective tissue diseases, Dermatomyositis, Systemic sclerosis, Mixed CTD, Lupus erythematosus, Undifferentiated CTD, Overlap CTD</td>
</tr>
<tr>
<td>Metastatic (elevated calcium or phosphorus)</td>
<td>Chronic renal failure, Hyperparathyroidism, Sarcoidosis, Milk alkali syndrome, Malignancies, Hypervitaminosis D, Calciphylaxis (abnormal calcium-phosphorus levels), Small vessel vasculopathy, Iatrogenic (ab/normal calcium or phosphorus levels), IV calcium-containing solutions, Sites of venipuncture</td>
</tr>
<tr>
<td>Idiopathic (normal calcium or phosphorus)</td>
<td>Subpidermal calcified nodules, Scrotal calcinosis, Tumoral (normal calcium and elevated phosphorus), Familial disorder</td>
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Abbreviations

CC-calcinosis cutis
CTD-connective tissue disease
DM-dermatomyositis
SLE-systemic lupus erythematosus
SS-systemic sclerosis.

Conflict of interests: none declared.
Financial support: none declared.
REFERENCES