Calcifying Lupus panniculitis in patients with No manifestations of Systemic Lupus Erythematosus

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Introduction

Lupus panniculitis named also as lupus profundus is characterized by one or several firm subcutaneous nodules with or without overlying epidermal changes (Figure 1). Sometimes, lupus profundus is manifested by erythematous plaques, some deep plaques and nodules ulcer involving the extremities, trunk, breasts, buttocks or face. Lupus panniculitis is considered a variant of lupus erythematosus cutaneous that primarily affects subcutaneous tissue [1]. Lesions appear while other may resolve slowly or may have long standing calcification.

Clinical Manifestations

Lupus panniculitis is estimated to occur with a frequency of 2-3% in patients with systemic lupus erythematosus (SLE) and between 10 and 50% of patients with lupus panniculitis will have or eventually develop SLE [2-4]. About 25% of patients with lupus panniculitis fulfilled the American College of Rheumatology criteria of SLE and antinuclear antibodies (ANA) are positive in 65% of patients in low titters [5]. Lupus panniculitis has been described associated to other entities and is not limited to patients with SLE. Prognosis is generally good, despite the association to systemic manifestations [6]. A panniculitis associated with a patchy lymphocytic infiltrate and deposition of mucin in the overlying dermis is suggestive of lupus paniculitis as shown in Figure 2 [7]. Lupus profundus received its name by Kren and Oppenheim in 1912 [8-10] and its diagnosis is made by clinical or histological findings. Signs of one or several firm, asymptomatic, well established and large subcutaneous nodules in patients with or without SLE is the most frequent clinical presentation [11]. It can lead to cutaneous and subcutaneous atrophy with occasional ulceration [12].

Histology

The histopathology consists in an inflammatory process with perivascular and perianxial lymphoid infiltrate predominance on lymphoid conglomerates that suggests germinal centres in addition to collagen hyalinization, fibrinoid necrosis, mucinosis, perivascularitis and microcalcifications in fat tissue; or may be large cumulates of calcifications which compromise lobules or septa [13,14]. Lesions will show variable degrees of calcification (it depends on the age of lesions) or sometimes with intense calcium deposits on previously damage fat lobules with hyaline necrosis frequently limited by a collagenic pseudocapsule [8]. Other changes described as common findings are foci of lymphocytes with or without germinal centers, hyalinization of septa, fat lobules, and lymphocytes within the vessel walls or on the perivascular tissue [15].

It is non-common to observe calcifications on deep soft tissues and patients suffered from pain when it happens sometimes along with the diagnosis of a renal transplantation and caciphylaxis [16]. Lesions have a chronic clinical course with remissions, recurrences and resolutions and are commonly accompanied by large areas of depression and lipoatrophy [17].

Pathogenesis

The Pathogenesis of calcium deposits is not clear but it is well documented that parathyroid hormone and vitamin D are not key factors. It was suggested that tissue alkaline phosphatase may activate extracellular pyrophosphatase (that normally inhibits calcium deposits) generating phosphates along with denaturized proteins of necrotic cells produced by inflammation of panniculitis [18,19]. This induces the production of phosphate calcium and calcareous deposits on lesions as is documented in dystrophic calcinosis [20].
The calcareous deposits damage the cytosolic sites producing cellular deposits and cellular death and may contribute to further calcareous deposition worsening the necrotic area, the acid environment and interfering on the action of the calcification inhibitors and the pyrophosphatases [21].

Treatment

Management of patients with lupus paniculitis includes antimalarials that were used the first time by Thurson and Curtis [10,22], azathioprine [23], cyclophosphamide [15] and dapsone. Thalidomide was recommended in lupus paniculitis by Burrows, especially when it is associated to partial C4 deficiency [24]. It has also been reported the management of older lesion calcifications with colchicine [25]. Some cases may respond to a combination of antimalarials such as hydroxychloroquine 200 mg and quinacrine 100 mg daily when a single drug is ineffective [26]. Other treatments include probenacid [27], low doses of warfarin [28-30] and Diltiazem [31]. Systemic glucocorticoids should be reserved for widespread or resistant lesions and Intraleisional glucocorticoids are usually ineffective because they exacerbate the atrophic healing process [32]. Adjuvant treatments include topical care and prevention from injury. Surgical debridement or resection of individual lesions may be attempted when all other modalities have failed and there is appreciable debilitation, presence of recurrent infection, painful masses, ulcerations, or local functional impairment [33]. It has been described lupus paniculitis associated to discoid lupus, subacute cutaneous lupus, and systemic lupus erythematosus. It has been described cases of acute calcifying paniculitis or secondary paniculitis associated to renal failure and/or calciphylaxis and also described in severe dystrophic calcinosis [34,35].

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