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A middle-age woman with subcutaneous plaque and hilar adenopathies

Communication

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Abstract: Subcutaneous sarcoidosis (SS) is an unusual and specific subtype of nodular sarcoidosis 1. The presence of SS with no elements of systemic manifestations is a rare condition: it is reported only in 1.4% to 6% of patients with systemic sarcoidosis, with the trunk being the most predilected area. Such cases with rare presentation are challenging for physicians because it can mimic several chronic infections, amyloidosis, hypothyroidism, lysosomal storage diseases and other conditions. Typical imaging (specially bilateral hilar adenopathies), histological exam and laboratory findings are the baseline to establish the diagnosis of sarcoidosis. In our case, the presence of subcutaneous manifestations avoided the performance of invasive procedures to get confirmation from other target organs: The epithelioid cells granulommas in subcutaneous fat and the representative radiological images were enough features to make the certain diagnosis. The first-line therapy for SS is oral steroids (20-40 mgr/day) with responses observed only 4-8 weeks after initiation of the treatment2. Prognosis of SS is good with spontaneous remission in some cases; however, when granulommas or fibrosis involves vital organs sarcoidosis can be life-threatening. Physicians should consider diagnosis of SS in patients with clinical suspicious history as sometimes skin manifestations are the first sign of systemic presentation of disease

Keywords: *Subcutaneous sarcoidosis • Gallium intake*

A 57-year-old female presented with an asymptomatic erythematous unique plaque over the right upper limb. This lesion appeared 10 months before and has gradually increased in size and number. She denied neither history of insect bite nor trauma. There were no further systemic features. On clinical examination, the plaque was barely visible (Figure 1, arrows), however it was detected easily by palpation: It was non-tender and firm in consistency with 15 cm x 8 cm measurements. Light palpation of left upper limb revealed a smaller lesion similar in consistency, with no cutaneous associated lesions (Figure 1, down). Cellulitis was considered as possible diagnosis and amoxicillin 1gr/day was initiated for 10 days. However, the lesion kept growing. Cultures of the tissue for bacteria, fungi and mycobacteria were negative. A detailed hospital checkup was performed, but it only showed slightly accelerated ESR and thrombocytosis. Levels of ANA, ENA, ANCA and complement were normal as well as protein electrophoresis which showed no signs of monoclonal gammapathy. Chest radiograph demonstrated bilateral hilar fullness. At this point the levels of angiotensin-convert enzyme (ACE) and serum calcium were measured, showing elevated ACE (150 U/L) and hypercalcemia (corrected calcium 15.4 mg/dl). With all these data, suspicion of sarcoidosis was considered and a skin biopsy of the plaque on the forearm was performed demonstrating non-caseating bacillus negative granulomas which suggested the diagnosis of sarcoidosis as the most likely one (Figure 3). A gallium scan was performed and revealed abnormal uptake in both forearms (Figure 2, down, arrows) and in the lung hila bilaterally which is typical of sarcoidosis –*lambda pattern*- (Figure 2 up, arrows). No significant intake was seen on legs (Figure 2 down right). Oral steroids (0.5 mg/ kg/ day) were initiated and after 2 weeks, lung and subcutaneous lesions improved with complete

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Figure 1. Right forearm shows a light erythematous plaque (Arrows). No cutaneous signs are observed on left forearm (Down).

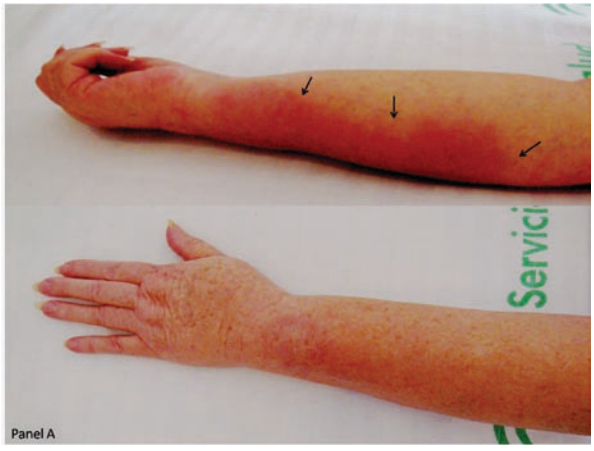
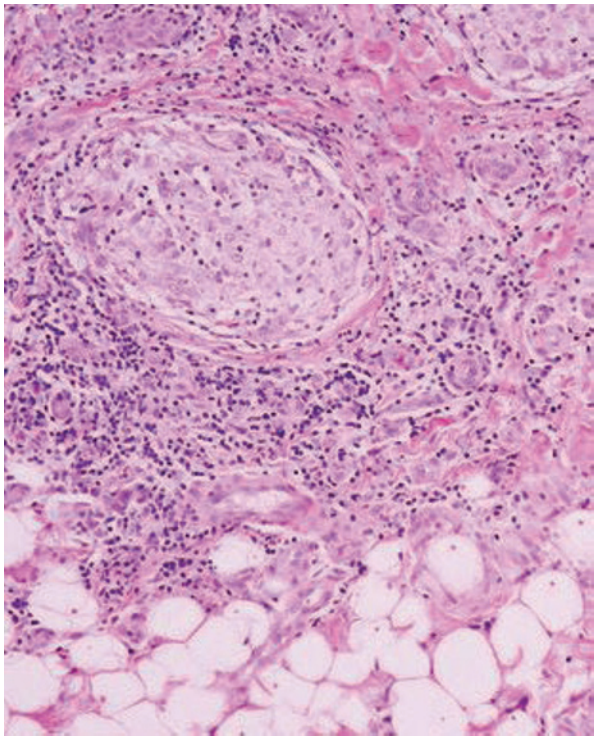
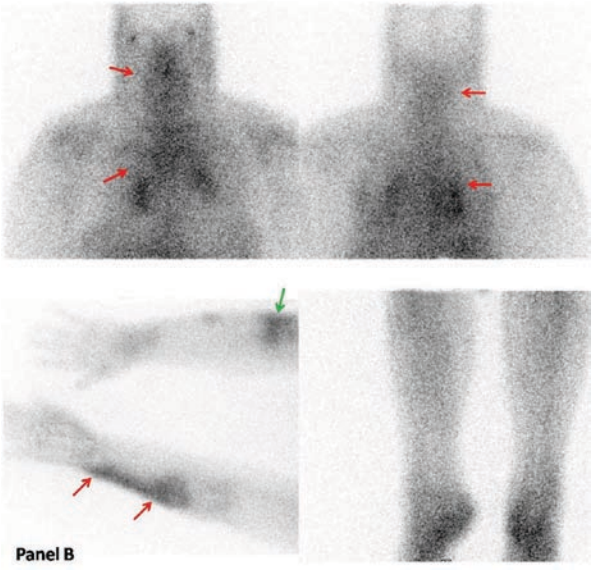


Figure 3. Non-caseating bacillusnegative granulomas in the deep dermis (Hematoxilin-eosine 20x).



response after 8 weeks. Follow-up laboratory values revealed that calcium was 8.4 mg/dl and ACE was 43 U/L. She has being followed- up for one year with no signs of recurrence. Subcutaneous sarcoidosis (SS) is an unusual and specific subtype of nodular sarcoidosis [1]. It was first described by Darier and Roussy in 1904 [2]. The presence of SS with no elements of systemic manifestations is a rare condition: it is reported only in 1.4% to 6% of patients with systemic sarcoidosis

Figure 2. Up: On gallium scan, anterior and posterior views of chest reveal abnormal intake on lung hila bilaterally, showing the typical lambda pattern of sarcoidosis. Down left: Right forearm shows abnormal intake, which was consistent with the clinical findings (Arrows). On left forearm, there is a small area of abnormal intake despite no cutaneous lesions were observed (Green arrow). Down right: No intake is observed on legs.



with the trunk being the most predilected area [3]. Our case presented with an uncommon location and it was misdiagnosed with cellulitis. Such cases with rare presentation are challenging for physicians because it can mimic several chronic infections, amyloidosis, hypothyroidism, lysosomal storage diseases and other conditions. Histological exam, typical imaging (especially *lambda pattern* in hilar adenopathies), and laboratory findings are the baseline to establish the diagnosis of sarcoidosis. In our case, the presence of subcutaneous manifestations avoided the performance of invasive procedures to get confirmation from other target organ: The epithelioid cells granulomas in subcutaneous fat and the representative radiological images were enough features to make the certain diagnosis. Although there are more accurate techniques such as computer tomography and positron emission tomography [4], we decided to performed an gallium scan because it is a helpful diagnostic tool when it shows the almost-unique pattern to sarcoidosis, but this can be attributed to its assistance in the determination the extent and distribution of the disease in cases with difficulties. Besides the lambda pattern, there is other specific pattern observed in the gallium scan: the *panda pattern*, which is due to the uptake of lagrimal and parotid glands. The first-line therapy for SS is oral steroids (20-40 mgr/day) with responses observed only

4-8 weeks after initiation of the treatment[2]. In general, prognosis of SS is good with spontaneous remission in some cases, however, when granulomas or fibrosis involve vital organs as lung, heart, liver or kidney, sarcoidosis can be life-threatening. Level of ACE has been found to be a useful assessment of disease activity and to evaluate the efficacy of the therapy [5]. In our patients, levels of ACE decreased dramatically with the therapy accompanied with the satisfactory response of the subcutaneous lesions. Physicians should consider the diagnosis of SS in patients with clinical suspicious history as sometimes skin manifestations are the first sign of systemic presentation of the disease [6].

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