Extensive Abdominal Wall Panniculitis. An unusual presentation of sarcoidosis

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Abstract

The involvement of the skin in sarcoidosis is present in around 5 to 35% of cases and it may be the initial manifestation of the disease.

The authors describe the clinical case of a 34 year old female patient, admitted in hospital with high fever, a 20kg [44 pounds] weight loss and an extensive panniculitis of the lower quadrants of the abdomen progressing for 2 months.

The cutaneous biopsy revealed a granulomatous lobular panniculitis without vasculitis and without caseous necrosis of the sarcoidosis type. All the other diseases that can cause a lobular panniculitis without vasculitis were excluded.

The patient evolved favorably taking oral corticotherapy and the clinical follow-up at 3 years excluded the involvement of other organs.

Key words: sarcoidosis, panniculitis, cutaneous manifestations.

Introduction

Sarcoidosis is a multisystemic granulomatous disorder of unknown etiology involving more often lungs, lymph nodes, mediastine, liver, spleen, eyes, parotid and skin. Less often, severe forms involve the central nervous system, the heart, the upper airways and the bones.[1,2]

The disease is characterized by the formation of non-caseous granuloma in the affected tissues,[1,2] showing typically a centre of epithelioid cells involved by CD 8^+ lymphocytes and mature macrophages.[3]

It is accepted that in its etiopathogeny there is a cell and humoral immune response (mediated by B and T lymphocytes) to an unknown antigen.

The cutaneous involvement in sarcoidosis is described in only 5 to 35% of cases[4,5,6] and lesions are classified as: specific when the histologic exam show the usual sarcoid granuloma (lupus pernio, infiltrated plaques, maculopapular rash, subcutaneous nodes, old scars infiltration) or non-specific (erythema nodosum, ichthyosiform lesions).[4,6,7,8] These lesions can be an initial and isolated manifestation of the illness, preceding for 6 months to 3 years the systemic involvement, or on the contrary, they may arise at a later stage, usually 6 months to 9 years after the generalized disease.[4,7]

Clinical case

A 34 years old female, Caucasian born and living in Lisbon, florist, healthy until a month and half before admission, date in which she started a daily fever (Axillary temperature 38ºC – 100.4 F), without shivers or chills, non-selective anorexia, with weight loss she quantified in 20 kgs [44 pounds], asthenia and adynamia, mentioning simultaneously the appearance of erythematous plaques, warm and painful, with 3 to 4 cms [1.2 – 1.6 inch] diameter, coalescing progressively, occupying the abdomen lower half.

She denied arthralgias, myalgias, headaches, changes on the intestinal tract, xerostomia, xerophthalmia, Raynaud´s phenomenon, breathing, eyes, urinary and other organs complaints.

Personal, family and epidemiologic history were irrelevant, denying bites or accidental traumas with plants or flowers during her working activity. Also denying exposure to heavy metals.

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TABLE I

Supplementary tests performed

1st line tests:

Red cells: 3.360.000/mm3

HB: 9.4 g/dL

HCT: 28.3%

MCV: 84rβ

Leukocytes: 6.500/mm3

w/ leukocyte formula N

Platelets: 330.000/ mm3

ESR: 80mm

Glucose: 80 mg/dl

Kidney liver F.: N

Iron: 51 ng/dl

Transferrin: 145 mg/dl

Ferritin: 442 ng/dl

Total protein: 6.3 g/dL c/ γ: 27%

Urinalysis II: no alterations

VDRL: non reactive

PA Thorax X Ray and ECG: no alterations

Subsequent exams:

Urocultures (3): E. coli

Mantoux (5 un): negative

CP reactive: 6.4 mg/dL

KB search (sputum and gastric fluid): Neg

Hemoculture (Brain/Bactec): Neg

Serology (Widal, Hudlesson, D. Lyme, Q, Fever, Bartonella, A, B and C Hepatitis and HIV1 and HIV2): Neg

C3, C4, CH50: Normal

ANA, antiDNA, antiSSA, antiSSB, antiSM, antiSCL70, LE cells: Neg

CIC: 4.2 mg/dL

TASO: 200U Tood

Band test: Neg

Other tests performed

Abdominal ultrasound: mild liver steatosis

Ultrasound (soft parts): marked diffuse thickness of the abdominal wall, with liquid lamina involving the subcutaneous fat, which assumes a lobular aspect suggesting edema

Pelvic ultrasound: w/ alterations

Myelogram: no alterations

Echocardiography: normal

Colonoscopy: w/ alterations

Small intestine enema: w/ alterations

Thorax CAT scan: w/ alterations

Bronchoalveolar wash with Bronchoscopy: w/ alterations

Eyes exam (slit-lamp examination): no changes

Respiratory functions tests: no changes

Objective exam: obese patient (1,65m/115kgs – 5ft4/18st11), without palpable adenopathies. Blood pressure: 110/70mmHg (lying down, right arm), Pulse: 80 b/m regular, rhythmic, wide. Breathing rate: 14 cycles/m. Axillary temperature: 38,2ºC. Pale skin and mucosa. Anicteric sclera, tongue and oropharynx without changes.

Breasts without alterations. Cardiopulmonary auscultation also without changes. Globous abdomen, with clear stretch marks, and in the abdominal wall and occupying almost all the totality of the infra-umbilical quadrants, a warm erythematous plaque with marked edema with non-defined borders and painful to palpation could be seen. There were no organomegalies. Rectal touch without alterations.

Bilateral malleolar edema was also present (++). Neurologic, gynecologic and ophthalmic exams were normal.

It was admitted the cellulitis diagnosis of the abdominal wall, and the patient received endovenous Flucloxacillin and Gentamicin for 10 days, without a significant clinical improvement.

The histopathological exam of the cutaneous biopsy has revealed "lobular panniculitis without vasculitis" and all the remaining supplementary tests (including bacterial, immune and imagiology) have only shown a normocytic-normochromic anemia and increased sedimentation rate (ESR) *(Table I)*.

As the diagnosis was inconclusive, it was performed a second biopsy, now taken from the subcutaneous cellular tissue of the abdominal wall, which has revealed: "granulomatous lobular panniculitis of lymphohistiocytic predominance, with sarcoid type granulomas without vasculites and without identifiable microorganisms through the Ziehl-Nielsen or Fite staining".

Of notice occurred three E. coli urinary infections.

After starting therapy with prednisolone, dosage of 60mg/day, it was seen apyrexia at the end of the 2nd day, with progressive improvement of cutaneous lesions, from the 4th day onwards and being discharged on the 7th day, asymptomatic with Hb 12,1g/dL and ESR 20mm.

Corticotherapy was withdrawn after 6 months, due to a total regression of the cutaneous lesions, with the patient asymptomatic and without evidence of systemic involvement or other organs at the end of three years.

Discussion

The case presented has shown to be a difficult differential diagnosis, with scarce clinical manifestations and an unspecific histopathologic first exam.

The possibility of cellulitis in the abdominal wall was excluded by the clinical condition persistence (fever, weight loss, panniculitis), in spite of the implemented therapy, and by the association with normocytic-normochromic anemia and increased ESR we accepted we were before a systemic disease.

In such circumstances, the differential diagnosis of lobular panniculitis without vasculitis includes pyogenic infections and *Mycobacterium tuberculosis,*[9] a pancreatic disease, a deep lupus, the Sweet syndrome, the subcutaneous lymphoma, leucemia,[10] Weber-Christian disease and sarcoidosis.

Without clinical or laboratorial evidence of pancreatic disease, of pyogenic infection (negative hemoculture) or tuberculosis (direct and cultural expectoration exam, gastric fluid, cutaneous fragment, thorax CAT scan were negative), subcutaneous lymphoma and leukemia were excluded through cutaneous biopsy, by the myelogram and the osteomedullary biopsy. Also the location and features of abdominal lesions and absence of leukocytosis and neutrophilia did not favor Sweet disease.

On the other hand, the second biopsy without polymorphonuclear infiltration and without subcutaneous nodes, have excluded Weber-Christian disease, reinforcing a sarcoidosis diagnosis.

Although the clinical presentation of sarcoidosis is more often characterized by the pulmonary and ganglionar involvement, the cutaneous lesions may precede in years the compromise of other organs, which is described in 25% of cases. In such circumstances it is crucial a prolonged monitoring of these patients.

This panniculitis histological aspects, the thorough exclusion of other etiologies and the excellent response to corticotherapy, enables us to state a diagnosis of sarcoidosis.

Due to the semiology scarcity, and low frequency rate, it seems to us justified the publication of this case. ■

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