

SWELLING FINGER REVEALING SYSTEMIC SARCOIDOSIS. A CASE REPORT

Melek Kechida (1), Mezri Maatouk (2), Manel Njima (3)

¹*Internal Medicine and Endocrinology Department, Fattouma Bourguiba University Hospital, Monastir 5000, Tunisia*

²*Radiology Department, Fattouma Bourguiba University Hospital, Monastir 5000, Tunisia*

³*Department of Anatomopathology, Fattouma Bourguiba University Hospital, Monastir 5000, Tunisia*

Abstract: Soft tissue localized sarcoidosis is rarely reported in the literature as well as associated cutaneous vasculitis. We present the case of a 34 year old patient with a history of a swelling in the distal left index finger phalanx evolving since 1 month who was admitted for a macular non pruritic rash of his legs. Laboratory findings and radiological investigations have guided the diagnosis of systemic sarcoidosis which was confirmed by finger biopsy. Cutaneous biopsy revealed associated vasculitis. The aim of this case report is to highlight that in case of progressive swelling in soft tissue associated to cutaneous vasculitis, systemic sarcoidosis is a possible differential diagnosis. Biological and radiological work out may guide the diagnosis and biopsy will confirm it.

Key words: sarcoidosis; fingers; vasculitis, leukocytoclastic, cutaneous

★ **Corresponding author.** Melek KECHIDA .Internal Medicine and Endocrinology Department, Fattouma Bourguiba University Hospital, 1st June street, Monastir 5000, Tunisia

Introduction:

Sarcoidosis is a systemic non caseating granulomatosis of unknown origin. Pulmonary manifestations are found in 90% [1.]Of the cases revealed by chronic cough or dyspnea on exertion. Extra pulmonary manifestations are various affecting heart, nervous system, gastro intestinal tract and eyes revealing sometimes the disease. Soft tissue involvement is rarely reported in the literature. Cutaneous manifestations can be specific or non-specific showing non caseating granuloma in biopsy. Leucocytoclastic vasculitis was rarely described as a cutaneous involvement associated to systemic sarcoidosis. Making the diagnosis of systemic sarcoidosis based on rare extra pulmonary manifestations may be difficult. We report a rare case of leucocytoclastic vasculitis revealing systemic sarcoidosis associated with soft tissue swelling with no bone involvement. Describing such a case allows highlighting the importance to have sarcoidosis as a differential diagnosis particularly in association with other systemic signs such cutaneous involvement needing further work up to confirm the diagnosis.

Case report:

A 34 year old man with a history of a swelling in the distal left index finger phalanx evolving since 1 month was admitted in the Internal Medicine Department for a macular non pruritic rash of his 2 legs since 2 days. On examination, he was afebrile, with no lymph nodes and could hardly flex his finger. X rays of the finger showed no bone involvement a part from enlarged soft tissue and chest X rays showed bilateral hilar opacities. Magnetic Resonance (MR) imaging of the left index showed marked,

well defined, thickening of the soft tissue surrounding the extensor digitorum in front of the proximal interphalangeal joint with no bone marrow oedema, bone destruction or joint involvement (fig 1 and 2). Body CT scan revealed bilateral and symmetric hilar lymph nodes with enlarged spleen (spleen index = 16 cm). Blood tests results are summarized in table 1. Cutaneous biopsy on the leg was consisting of leucocytoclastic vasculitis. A biopsy of the finger revealed a typical non caseating granuloma in favor of sarcoidosis (fig 3). Eye examination showed no uveitis or retinal vasculitis. Respiratory functional exploration indicated no reduced vital capacity. Treatment consisted in 0.5 mg/ kg / day of prednisone with a stepwise decrease. After 1 year of treatment, there were no lymph nodes or splenomegaly on a control CT scan.

Table 1: laboratory findings of the patient

Parameters	Laboratory results
Hemoglobin	12.7 g/dl
Platelets	210 * 10 ³ / mm ³
WBC	4600 Elts /mm ³
Lymphocytes	1100 Elts/ mm ³
ESR	04 mm / 1st h
Serum calcium	2.29 mmol/ l
Calciuria	0.18 mmol/kg/24 h
ACE	119 UI/l (19 -70UI/l)

WBC: white blood cell, Elts: elements, ESR: erythrocyte sedimentation rate, ACE: angiotensin converting enzyme

Legend of the figures:

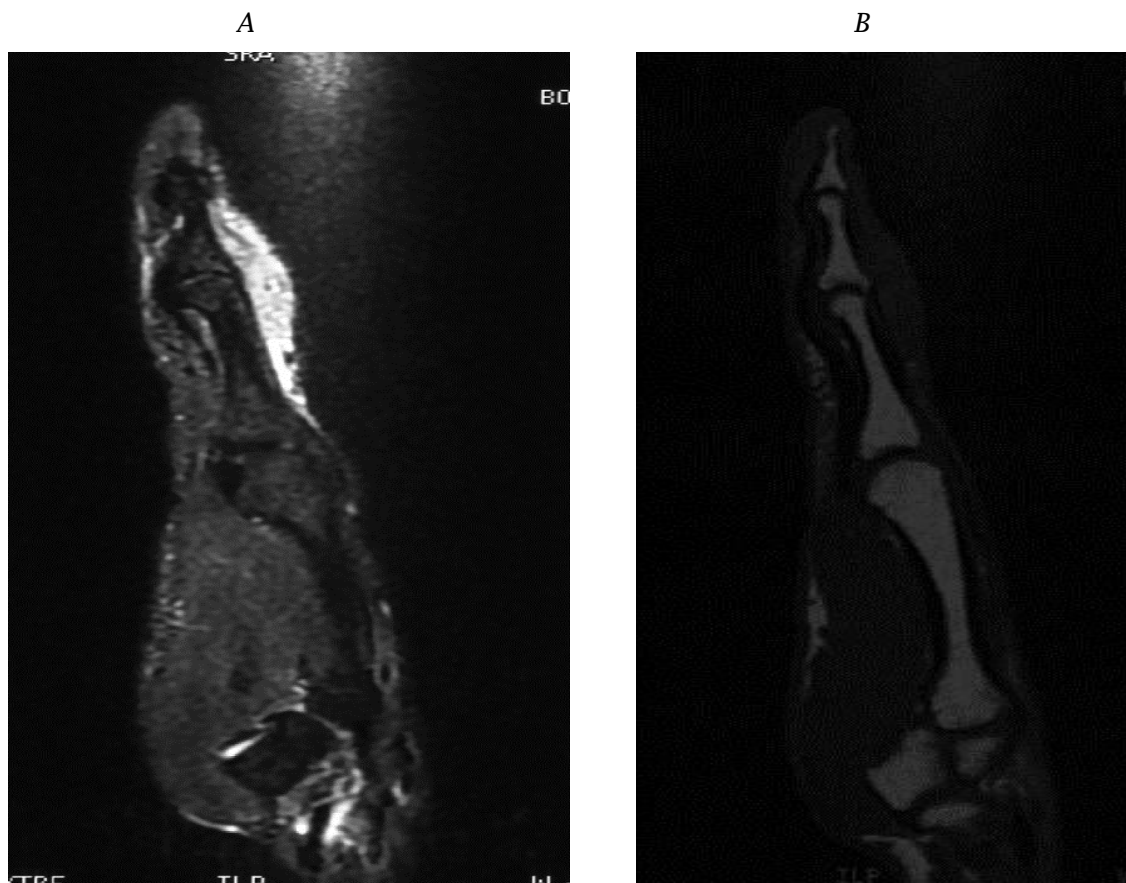


Figure 1: Magnetic Resonance (MR) imaging of the left index: sagittal fat saturated T2 weighted MR (A), sagittal T1 weighted MR (B): marked well defined thickening of the soft tissue surrounding the extensor digitorum in front of the proximal interphalangeal joint. There is no bone marrow oedema, bone destruction or joint involvement

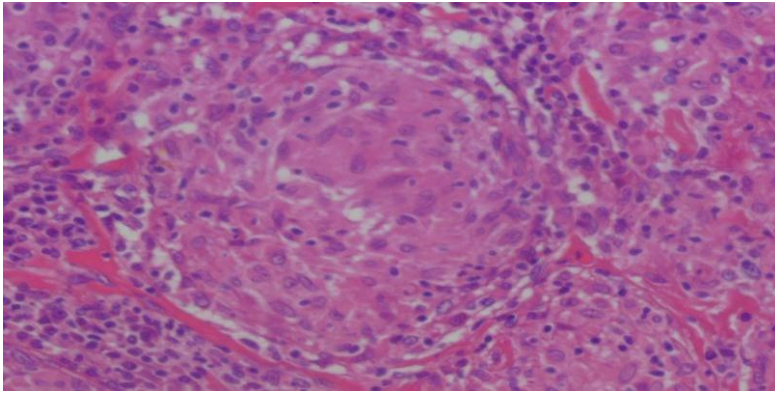


Figure 2: Granuloma infiltration of the dermis. Hematoxylin and eosin, original magnification x 4

Discussion:

We described a rare case of finger soft tissue involvement revealing systemic sarcoidosis associated to leucocytoclastic vasculitis. Sarcoidosis is a multisystem granulomatosis of unknown origin. Pulmonary involvement in sarcoidosis represents 90% of the cases but almost any other organ can be affected such as nervous system, heart, gastrointestinal tract and eye [1.]. Soft tissue involvement is rarely described in the literature. Gottlieb and Wenner reported the same case of our patient with soft tissue involvement of the finger without bone manifestations [2.]. Soft tissue involvement may have various differential diagnoses in that localization like foreign body granuloma, giant cell tumor, lipoma, neurofibroma synovitis and metastatic cancer. To establish the diagnosis of sarcoidosis, it's worth noting that it requires a series of clinical, radiological and biological arguments. Histopathological findings with non caseating granuloma will be the hall mark of the diagnosis. In our case, Computed Tomography (CT) scan findings including bilateral symmetric hilar nodes associated to enlarged spleen and biology finding consisting of hyper calciuria, lymphopenia, elevated LDH and angiotensin converting enzyme were arguments in favour of sarcoidosis. Biopsy findings confirmed the diagnosis.

Cutaneous manifestations in sarcoidosis are variable occurring in 20 - 25% of cases [3.]. Specific lesions are nodular or erythematous purplish macules. Non specific lesions are mainly erythema nodosum [4.]. In our case, leucocytoclastic vasculitis was the histological feature of the cutaneous eruption which occurred concomitantly to sarcoidosis. Vasculitis associated to sarcoidosis was described first in 1979 [5.]. Few other cases were than described. Such histological finding is suggested to be mediated by circulating immune complexes, explaining also extra pulmonary features of sarcoidosis [6.].

Conclusion:

Investigating progressive soft tissue swelling should take into account sarcoidosis as a differential diagnosis. Laboratory tests and radiological findings can guide the diagnosis. Histopathology findings will confirm the hypothesis. Leucocytoclastic vasculitis is another cutaneous manifestation during systemic sarcoidosis which should be known by physicians.

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