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## Unusual Presentation Of Sarcoidosis: Case Report

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### ABSTRACT

Sarcoidosis is a multisystem granulomatous disease with polymorphic manifestations, of unknown etiology, that can affect any organ in the body, but most commonly the lungs [1,2,3]. Extrapulmonary involvement is common, can occur in association with or in the absence of intra-thoracic disease, and all organs can be involved, but it is rare to find an isolated extrapulmonary disease (less than 10% of patients) [3,4]. On occasion, the presentation of sarcoidosis may be atypical.

We report a case of systemic sarcoidosis, with particular clinical data: extrathoracic onset with subdiaphragmatic lymph nodes, hepatomegaly and splenomegaly.

Keywords: sarcoidosis, hepatomegaly, splenomegaly

### Case report

A 37-years-old man, farmer by profession, was admitted to the Department of Surgery for upper abdominal pain, fatigue, weight loss, with a high suspicion of gastric cancer.

Personal history and family history were insignificant.

The onset was insidious, with two months prior this admission, with progressive weakness to which weight loss (10 kg) and nonspecific upper abdominal pain were associated later.

His physical examination revealed only hepatomegaly and splenomegaly.

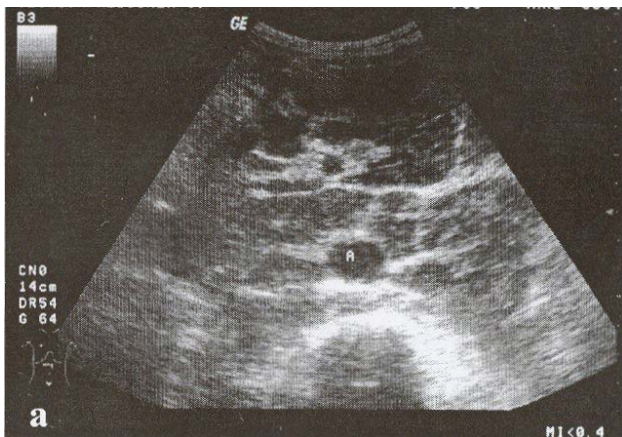
Laboratory studies indicated biological inflammatory syndrome (ESR, slightly elevated fibrinogen), hypergammaglobulinemia and a negative tuberculin purified protein derivative (PPD) skin test.

Abdominal ultrasound and CT scan documented multiple subdiaphragmatic lymph nodes (periaortic, inter-aorto-caval and iliac), with diameter from 1.75 to 2.5 cm (Figure 1).

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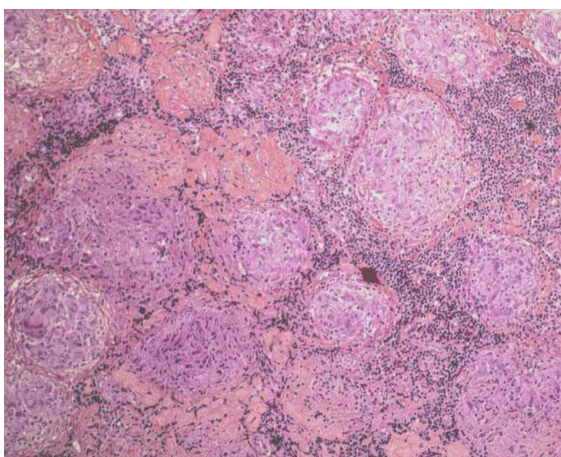


*Figure. 1: Abdominal ultrasound: multiple periaortic lymph nodes*

Because the findings were highly suspicious for a digestive tumor, GI endoscopy ruled out a neoplasia with this location.

Chest radiographs, chest CT scan and pulmonary function tests were performed, but they were all normal.

At this point, the patient underwent a diagnostic laparotomy. Splenectomy, lymph node and liver biopsy nodule were performed. Pathology report showed chronic non caseating epithelioid granulomas, with giant Langhans cells and accentuated fibrosis (Figure 2).



*Figure. 2: Non caseating epithelioid granulomas, with giant Langhans cells and accentuated fibrosis*

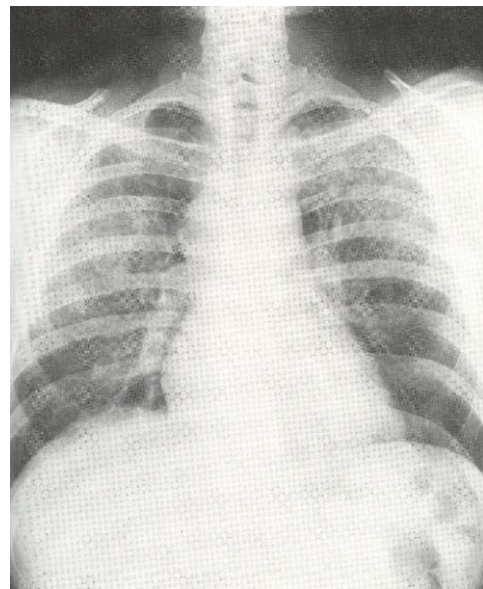
Positive diagnosis of extrapulmonary sarcoidosis was made.

Differential diagnosis included extrapulmonary tuberculosis, digestive tract neoplasia, malignant lymphomas, leukemias.

Steroid therapy was instituted (prednisone 1 mg / kg / day for 30 days, then dose was gradually reduced to 10 mg every 2 days), treatment followed for 18 months. After three months of therapy, complete regression of lymph nodes, hepatomegaly and general signs were shown. 18 months later, the treatment was discontinued due to favorable evolution, the patient following to be evaluated every 3 months, indication which he did not comply with. One year and a half after treatment discontinuation, the disease has relapsed consisting of recurrence of subdiaphragmatic lymph nodes, hepatomegaly but, this time, lung impairment was associated.

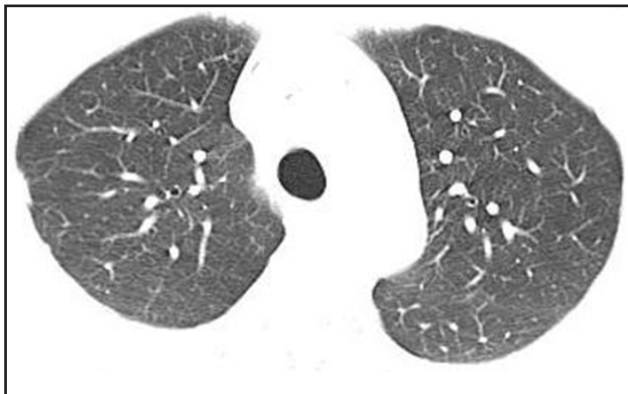
Impairment of the lungs was documented by:

1. Chest radiograph: reticulo-nodular fibrosis especially in the upper 2/3 of the lung fields (Figure 3);



*Figure 3: Chest radiography : Diffuse pulmonary fibrosis located predominantly in the upper 2/3*

2. CT pulmonary : **bilateral** reticular fibrosis in the upper 2/3 (Figure 4);



*Figure. 4: CT scan shows reticular opacities with upper zone distribution*

3. Pulmonary function tests: restrictive ventilatory dysfunction (FVC = 36% of predicted value, FEV1% = 41% of predicted value, IPB = 88%) with decreased transfer factor(57,1%) of predicted value;

4. Bronchoscopy showed only intense hyperemia of bronchial mucosa, with intense capillary vessels (Figure 5); bronchoalveolar lavage fluid (BAL) examination: lymphocytic alveolitis (lymphocytes 46% of the total number of cells), the cytology was negative for neoplastic cells.



*Figure 5: Bronchoscopy: intense hyperemia of bronchial mucosa, visible capillary vessels.*

5. Serum angiotensin convertase (CEA) was dosed with a positive result, 33.5 U/L (N: 15-28 U/L).

Treatment with prednisone was resumed, 1 mg / kg / day 30 days, with further reduction of the dose. After 3 months of treatment subdiaphragmatic lymph nodes disappeared and respiratory function tests were slightly improved, which allowed the progressive reduction of corticosteroid dosage to a maintenance dose of 20 mg to 2 days.

During treatment, there was an iatrogenic complication – cortisone induced diabetes, which required hygienic-dietary measures and treatment with oral antidiabetic agents (repaglinide).

The particularity of the case is the extrathoracic onset with hepatomegaly, splenomegaly and subdiaphragmatic lymph nodes manifestations that are rare in sarcoidosis. Pulmonary impairment appeared at first relapse, after 3 years of disease evolution, and included stage 3 of sarcoidosis, with pulmonary fibrosis..

## **Discussion**

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Sarcoidosis is a granulomatous disease of unknown aetiology with various clinical presentations depending on affected organs [1,3,9]. Lung and lymphatic system are the principal localizations [7,11,12]. Typical clinical manifestations are easier recognized, but unusual presentations, such as nodular lesions of spleen and/or liver, are difficult to diagnose in absence of clinical suspicious of sarcoidosis [1,3,9]. The diagnosis of sarcoidosis could be more complicated especially if abdominal sarcoidosis represents the exclusive involvement without



systemic disease [3,6,9]. Usually, extrapulmonary disease occurs in the context of more extensive chest disease [13,14,15]. Isolated splenic sarcoidosis or abdominal sarcoidosis without pulmonary or mediastinal involvement is rare, such as our patient [4,5,13]. We have described a patient with extrapulmonary sarcoidosis initially who later developed pulmonary involvement. Symptoms of extrapulmonary sarcoidosis are usually less characteristic for this localization. Abdominal pain and systemic symptoms, including fever, malaise and weight loss occasionally occur. Laboratory tests are usually normal, but some patients with splenic involvement can develop anaemia, thrombocytopenia and neutropenia [16,17,18]. Our patient had no blood count abnormalities only positive non-specific inflammatory markers and a slightly increased titer of ACE. Abdominal ultrasound scan and HRCT can easily detect the abdominal lesions. Splenomegaly (greatest splenic dimension >14 cm) is the most common splenic manifestation [19,20]. The organ is usually homogeneous, but multiple low-attenuating nodular lesions are occasionally noted and easily mistaken for lymphoma, metastases or infections [13,15,19]. Isolated or predominant hepatic nodular disease is less common than spleen involvement [2,5,13]. Besides spleen involvement, our patient presented also a hepatic nodule. In this case, for ascertain the correct diagnosis, splenectomy and hepatic nodule biopsy were required. The extent of pulmonary involvement, was evaluated both at the diagnosis and subsequent follow-up visits. At the time of diagnosis, the patient had no respiratory symptoms. We have seen that the progressive development of the disease is variable: the patient first described with initial diagnosis of extrapulmonary sarcoidosis but after that 3 years since diagnosis was made, he

presented pulmonary impairment. The treatment directed against the granulomatous inflammation is effective [3,18]. In this case, oral corticotherapy was administered for 18 months, which was successful with complete remission of abdominal lymphadenopathies and symptoms. This made possible to discontinue the therapy and to have a follow-up check every 3 months, but the patient missed his periodic evaluation and he presented 18 months later when he developed systemic sarcoidosis. When anti-sarcoidosis medications are withdrawn, relapse is very common and typically occurs 1 month to 1 year after therapy is tapered or discontinued [3,18].

## Conclusion

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Sarcoidosis is a chronic multisystem granulomatous disease. Atypical onset such as splenic or hepatosplenic involvement can occur and it is possible to be an exclusive involvement of these organs without pulmonary disease. Diagnosis of extrapulmonary sarcoidosis is difficult because, usually, is not suspected. In fact, hepatosplenic or/and splenic determinations are often asymptomatic or accompanied by unspecific symptoms and laboratory abnormal values [3,10]. In our case, US and abdominal CT scan identified pathological changes but the diagnosis was established only with the histological examination which is required to differentiate liver and/or spleen sarcoidosis from tuberculosis, primary biliary cirrhosis, metastasis or malignant lymphoma and other granulomatous diseases [3,10]. After diagnosis, continual follow-up for systemic manifestations is indicated. In some cases, recurrence

of the disease may be considered an acute exacerbation of the sarcoidosis but in our situation, the relapse may have been controlled if the patient would have been compliant to follow-up visits. On one hand, this case shows that the diagnosis of sarcoidosis can be difficult when only extrapulmonary impairment is involved. Furthermore, atypical presentations may result in a delay in diagnosis as well as unnecessary treatment or diagnostic testing [3,10].

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