Pulmonary and splenic sarcoidosis with atypical onset

Sarcoidoză splenică și pulmonară cu debut atipic

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Abstract

Sarcoidosis is a multisystemic disorder that can mimic other pathologies with multiorgan involvement, with a difficult differential diagnosis and delaying the definitive diagnosis. Sarcoidosis can take atypical clinical and radiological forms, often being mistaken for tuberculosis or neoplasia. The authors present the case of a 46-year-old male patient who underwent splenectomy for the suspicion of a lymphoma. The histopathologic examination confirmed the diagnosis of sarcoidosis. After one year of treatment with methylprednisolone, significant regression of the pulmonary micronodular opacities and lymphadenopathies (thoracic and abdominal) was revealed, with serum angiotensin convertase (ACS) returned to normal and the patient stationary at the functional respiratory tests. The specificity of this case is the atypical onset of symptoms only in the abdominal area, without respiratory manifestations (dyspnea or cough), contrary to the involvement of the pulmonary interstitium, being necessary a splenectomy to establish the diagnosis.
Keywords: ***atypical sarcoidosis, lymphoma, splenectomy***

Rezumat

Sarcoidoza este o afecțiune multisistemică ce poate mima alte patologii cu afectare multiorganică, ridicând dificultăți de diagnostic diferențial și întârziind diagnosticului de certitudine. Sarcoidoza poate îmbrăca forme atipice clinice și radiologice și se confundă cel mai adesea cu tuberculoza sau cu diverse neoplazii. Autorii prezintă cazul unui bărbat, de 46 de ani, care a fost supus unei intervenții chirurgicale pentru suspiciunea de limfom, dar examenul histopatologic a pus diagnosticul de sarcoidoză. Evoluția pacientului pe parcursul unui an de tratament cu metilprednisolon a fost favorabilă, cu regresia semnificativă a modificărilor interstițiale pulmonare și a adenopatiilor mediastinale și abdominale, cu angiotensin convertaza (ACS) revenită la normal și cu pacientul staționar din punct de vedere funcțional respirator. Particularitatea cazului o reprezintă debutul fără simptomatologie respiratorie, deși era prezentă afectarea interstițială pulmonară, mai mult, necesitând splenectomie pentru confirmarea diagnosticului.
Cuvinte-cheie: ***sarcoidoză atipică, limfom, splenectomie***

Introduction

Sarcoidosis is an inflammatory systemic disease of unknown etiology characterized by the presence of non-caseating granulomas with the involvement of any organ[(1)]. The primary location of the disease is the pulmonary system, in more than 90% of cases[(2)]. Although abdominal involvement is uncommon, it can more easily mimic an infectious or a neoplastic pathology, causing real differential diagnosis issues[(3)].

Case presentation

The authors present the atypical case of a 46-year-old male patient with no previous pathology, describing the insidious onset of a left hypochondrium pain and facial hyperemia. The patient was referred to the hematology department where a whole-body computed tomography (CT) scan revealed bilateral micronodular pulmonary opacities (Figure 1A), mediastinal and abdominal lymphadenopathies, significant spleen enlargement (Figure 1B), findings suggestive for non-Hodgkin malignant lymphoma. An osteomedullar biopsy was performed and showed the presence of normoblastic erythroblastic hyperplasia, possibly of neoplastic etiology.

The patient was referred to the department of general surgery for splenectomy and the pneumological preoperative assessment excluded pulmonary active infection with *Mycobacterium tuberculosis*.

A laparoscopic splenectomy and lymph node biopsy of the splenic hilum were performed, with initial unfavorable postoperative outcome and the onset of fever (38-39 °C) on day one.

A second chest and abdominal CT scan showed the presence of newly formed bilateral pulmonary consolidation areas, with bilateral pleural effusion, more on the left side (Figures 2A and 2B). Left chest tube drainage and broad-spectrum antibiotic therapy were initiated, with the remission of fever and the resolution of pulmonary radiologic elements of an infectious substrate (Figures 3A and 3B).

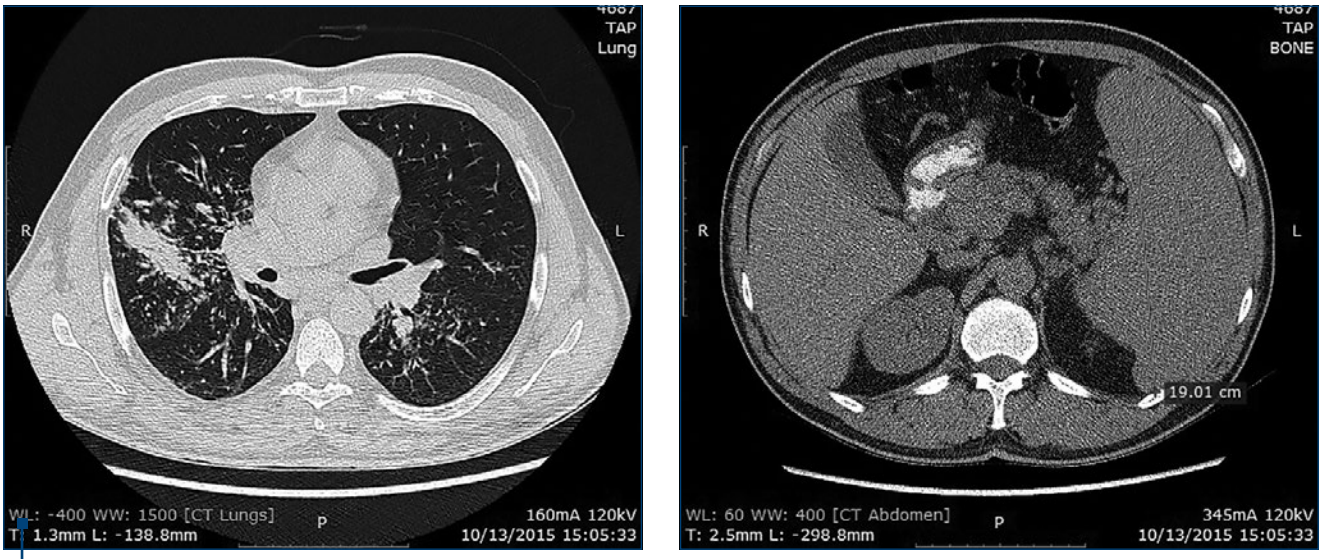
The histopathological and immunohistochemistry examinations of the removed spleen and splenic hilum lymph node revealed non-caseating chronic granulomatous inflammation, positive CD68, CD4, CD8, CD4/CD8 ratio = 5.2, suggestive features of sarcoidosis, but that cannot rule out tuberculosis. For a differential diagnosis, polymerization chain reaction (PCR) from the patient's sputum once again excluded the infection with *Mycobacterium tuberculosis*.

The patient was referred to our pneumology department for further investigations and treatment at 5 months from the onset of symptoms.

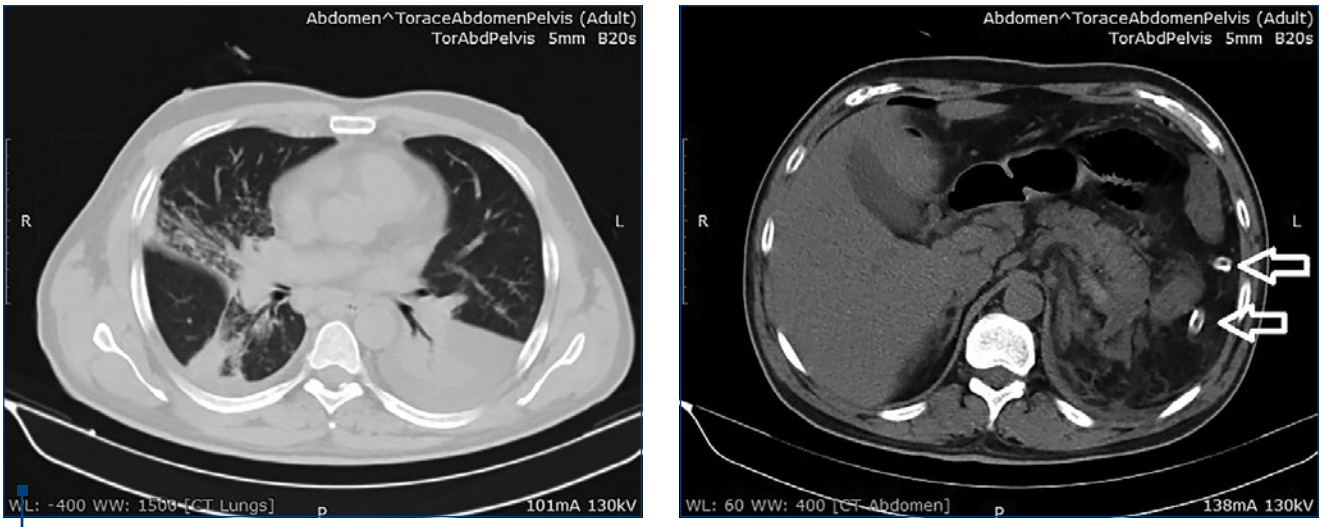
Bronchoscopy with bronchial-alveolar lavage showed the predominance of lymphocytes – 49%, CD4/CD8 ratio =4.1, and serum angiotensin convertase (ACS) of 67 U/L

Table 1 Comparative values of lung function testing, ACS serum level and thoracic and abdominal CT scan features before and after methylprednisolone treatment

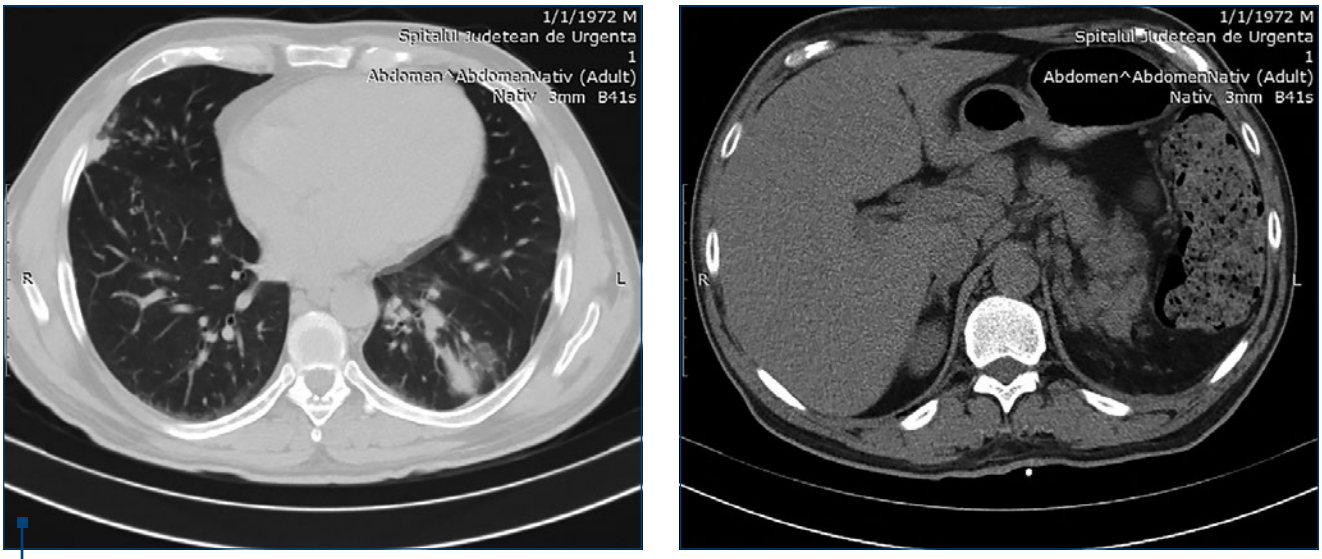
			Before Treatment	After Treatment
Pulmonary function testing	**Spirometry**	TLC	5.35 L (86.5 %)	5.51 L (88 %)
		FVC	3.04 L (74.6 %)	3.16 (76.8 %)
		FEV1	2.46 L (73.1 %)	2.77 L (81.8 %)
		FEV1/FVC	85.75 %	85.93 %
		PEF	5.83 L (69%)	7.22 L (85.3 %)
	Diffusing capacity	DLCO	8.29 (87.2 %)	9.16 (95.9 %)
		KCO	1.95 (126.7%)	1.96 (128.3%)
Serum levels	**ACS**		67 U/L	17 U/L
Thoracic and abdominal CT scan features	**Spleen**	Measurements	19/8/18 cm	excised
		Splenic index	2736	
		Splenic volume	1617 mL	
		Splenic weight	1698 g	
	Abdominal lymphadenopathies	Mesenteric	15/13 mm	15/6 cm
		Hepatic hilum	28/15 mm	normal
		Splenic pedicle	15/10 mm	excised
		Left paraaortic	20/16 mm	normal
		Pancreas hilum	14/8 mm	normal
	Thoracic lymphadenopathies	Right paratracheal	26/12 mm	16mm
		Aortic arch	22/8 mm	normal
		Prevascular	34/14 mm	normal
		Below carina	18/10 mm	normal
		Carinal level	18/10 mm	normal
		Right pulmonary hilum	19/8 mm	normal
		Left pulmonary hilum	14/8 mm	10mm
	Liver	Measurements	At upper normal limit	normal



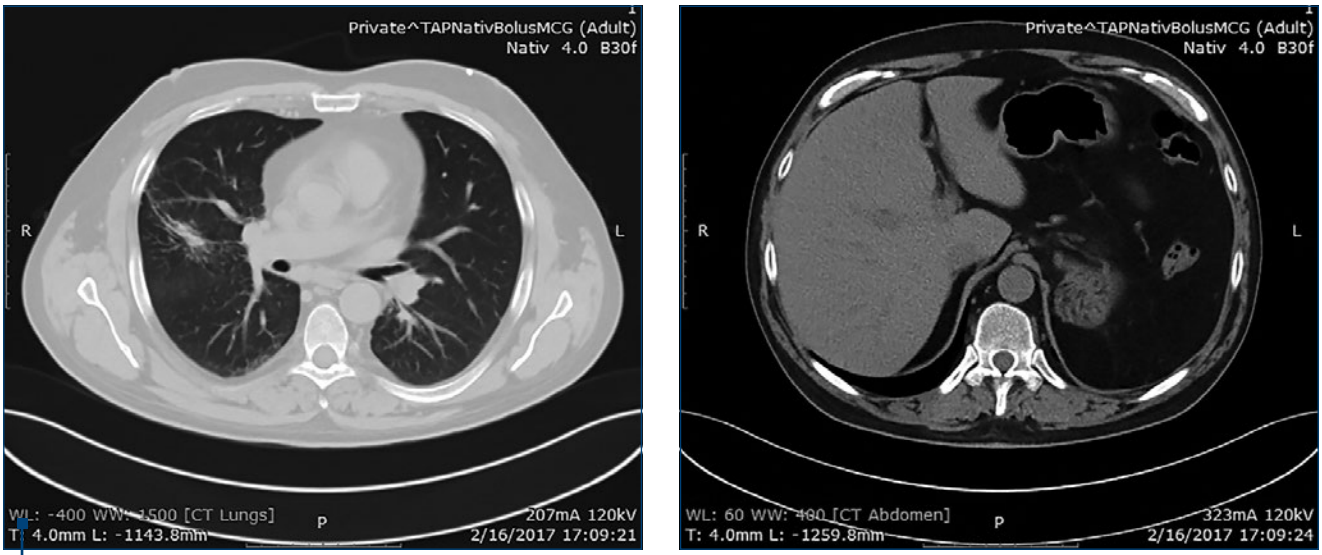
Figures 1A and 1B. *Axial high-resolution CT scan (parenchymal window) reveals bilateral micronodular pulmonary opacities and significant spleen enlargement (abdominal window)*



Figures 2A and 2B. *Axial high-resolution CT scan (parenchymal window) reveals newly formed bilateral pulmonary consolidation areas, with bilateral pleural effusion, more on the left side, and normal postoperative evolution after splenectomy, with the presence of two abdominal drainage tubes (abdominal window)*



Figures 3A and 3B. *Axial high-resolution CT scan (parenchymal window) reveals the resolution of pulmonary radiologic elements of infectious an substrate and uneventful recovery after splenectomy (abdominal window)*



Figures 4A and 4B. *Axial high-resolution CT scan (parenchymal window) reveals almost complete resolution of pulmonary opacities and long-term uneventful recovery after splenectomy (abdominal window)*

(laboratory normal values range 12-65 U/L). The respiratory functional samples showed mild restrictive ventilatory dysfunction and normal alveolo-capillary diffusion. The 6-minute walking test was within normal parameters and without desaturation. All investigations supported the histopathological diagnosis of sarcoidosis (Table 1).

Given the multisystemic involvement, with splenic, pulmonary and lymph nodes, the decision was to begin the therapy with methylprednisolone 32 mg per day, decreasing the dose progressively over a one-year period.

The patient was evaluated periodically, with thoracic and abdominal CT scans revealing significant regression of the pulmonary micronodular opacities and lymphadenopathies (thoracic and abdominal) towards the end of treatment (Figures 4A and 4B).

ACS was converted to normal values, and the respiratory functional tests showed stationary status. The follow-up at 12 months did not show signs of relapse (Table 1).

Discussion

The incidence of splenic sarcoidosis ranges between 10% and 50%, depending on the clinical judgement, imagistic findings and the histopathological examination of various resected specimens[4].

Spleen involvement is mostly asymptomatic, but occasionally pain has been described in the left hipocondrum, as well as non-specific symptoms such as sweating, fever and malaise.

In about 3% of patients with splenic sarcoidosis, massive spleen enlargement was described. In isolated cases, hypersplenism causes various grades of anaemia leukopenia, thrombocytopenia, to severe conditions including pancytopenia. The radiological findings of splenic sarcoidosis are sometimes multiple nodules throughout the parenchyma, that mimic abdominal carcinomatosis, a feature found more frequently in malignant lymphoma[5].

Most cases of splenic sarcoidosis do not require surgical treatment. The indications of splenectomy include abdominal pain due to splenomegaly, hypersplenism, functional asplenia or spontaneous splenic rupture[6].

The specificity of this case is the atypical onset of symptoms only in the abdominal area, with pain in the left hypochondrium, which is a general symptom, without pathognomonics of a particular disease. The patient didn't present respiratory symptoms (dyspnea or cough), contrary to the rather important involvement of the pulmonary interstitium revealed at the thoracic CT examination. Functional respiratory samples, however, showed mild restrictive syndrome.

In this case, the laparoscopic splenectomy was performed due to the high suspicion of lymphoma, taking into account the initial histopathological result of the osteomedullar biopsy and the isolated abdominal symptoms.

Similar cases of splenic sarcoidosis are cited in the medical literature, proving the variability of the clinic forms of sarcoidosis[6]. ■

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