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Idiopathic pleural panniculitis with recurrent pleural effusion not associated with Weber-Christian disease

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Abstract: A 82-year-old patient with dyspnea and a recurrent history of pleural effusion was admitted into our unit. He performed a chest computed tomography showing right pleural effusion. Video-assisted thoracoscopy (VATS) exploratory showed parietal pleural thickening of adipose tissue. The surgical procedure consisted, therefore, in the execution of multiple biopsies of the parietal pleura which appeared covered, on the whole surface, by islands of adipose tissue, without macroscopic pathological aspects. After the procedure was performed pleurodesis with talc. The definitive histological examination consisted of normal mesothelial cells surrounded by fatty tissue infiltrated by small lymphocytes in a patient without skin lesions or visceral or systemic signs of inflammatory involvement of the adipose tissue. We reported a rare case of idiopathic pleural panniculitis with recurrent pleural effusion not associated with Weber-Christian disease.

Keywords: Panniculitis; Thoracoscopy; Exploratory excision; Weber-Christian disease

1 Introduction

Idiopathic panniculitis is a systemic inflammatory non suppurative disease of adipose tissue of unknown etiology [1-4]. This clinical evidence is often observed in Weber-Christian disease (WCD) associated with cutaneous lesions (recurrent subcutaneous inflammatory painful nodules) and systemic manifestations (fever, malaise and, sometimes, flogistic involvement of the lungs, heart, gastrointestinal tract, spleen, kidneys, and adrenal glands). The diagnosis of lobular panniculitis, in our case, was obtained on pleural biopsy with histological examination consisting of normal mesothelial cells surrounded by fatty tissue infiltrated by small lymphocytes without skin lesions or visceral or systemic signs [6-15]. In our case, we reported a pleural panniculitis with recurrent pleural effusion not associated with Weber-Christian disease.

2 Case Report

A 82 year old man was hospitalized because of fever, dyspnea and right pleural effusion. He had an history of repeated thoracentesis, diabetes (NIDDM) and COPD. On hospital admission, clinical general conditions showed a pulse rate 80 and respiratory rate 34 per minute without hepato-splenomegaly neither painful erythematous subcutaneous nodules. A chest roentgenogram showed a right-sided pleural effusion. A subsequent total body computed tomography (CT) confirmed the pleural effusion with no evidence of pancreatic abnormality. Laboratory data showed an erythrocyte sedimentation (VES)
rate of 15 mm·h⁻¹ and C-reactive protein (CRP) of 3.0 mg·dL⁻¹. White blood cell count was 7,500 cells·mm⁻³ (7.5 x 10⁹·L⁻¹) with Haemoglobin concentration of 130 g·L⁻¹, and 240,000 platelets·mm⁻³ (240 x 10⁹·L⁻¹). Glutamic oxaloacetic transaminase (GOT) was 34 IU·L⁻¹, and glutamic pyruvic transaminase (GPT) 40 IU·dL⁻¹. Lactate dehydrogenase (LDH) was 650 IU·L⁻¹, and creatine phosphokinase (CPK) was 350 IU·L⁻¹. Serum amylase was 85 IU·L⁻¹, and lipase was 30 IU·L⁻¹. Electrophoretic protein pattern was 6.0g·dL⁻¹, and albumin 3.0g·dL⁻¹. No antibodies against itself were produced (tests for antinuclear factor, rheumatoid factor and immunocomplex) and urinalysis was normal. Alpha1-antitrypsin (A1AT) was 184 mg·dL⁻¹. QuantiFeron test was negative. A cardiology evaluation showed no heart failure manifestations. Right-sided thoracentesis showed a sterile pleural exudate with pleural adenosine deaminase (ADA) level not elevated. Respiratory function showed a moderate obstructive deficit. Thoracoscopic examination revealed parietal pleural thickening of adipose tissue. The surgical procedure consisted, therefore, in the execution of multiple biopsies of the parietal pleura which appeared covered, on the whole surface, by islands of adipose tissue, without macroscopic pathological aspects (Fig. 1). After the procedure we performed pleurodesis with talc. The definitive histological examination consisted of normal mesothelial cells surrounded by fatty tissue infiltrated by small lymphocytes. The patient had no skin lesions or visceral or systemic signs of flogistic involvement of the adipose tissue. Treatment with prednisolone (50 mg·day⁻¹ for the first week, then 25 mg for the second week) was initiated. A one month-follow up showed no pleural effusion and a complete resolution of symptoms.

**Ethical approval:** The research related to human use has been complied with all the relevant national regulations, institutional policies and in accordance the tenets of the Helsinki Declaration, and has been approved by the authors’ institutional review board or equivalent committee.

**Informed consent:** Informed consent has been obtained from all individuals included in this study.

### 3 Discussion

Weber-Christian disease (WCD) [16-24] characters (recent non-suppurative nodular panniculitis [25-30], fever and painful cutaneous nodules, malaise, arthralgia, hepatosplenomegaly, weight loss and anorexia [31-38]) were described by Weber first, and Christian subsequently. While lobular panniculitis associated with extensive visceral involvement may lead to a severe prognosis and eventually to death, due to sepsis, hepatic failure, hemorrhage, and thrombosis, only cutaneous involvement was a positive prognostic indicator. Pleuritis is a rare complication of this systemic panniculitis [39-44]. Deficient levels of pleural ADA were not found. The thoracic complications, in our case, were not associated with extensive visceral involvement. A diagnostic thoracoscopy, performed for recurrent pleural effusion, showed pleural thickening on the whole surface with
islands of adipose tissue, without macroscopic pathological aspects.

Pleural thickening biopsy allowed us to diagnose of idiopathic pleural panniculitis.

In our case, thoracoscopic biopsies allowed us to diagnose a rare case of idiopathic pleural panniculitis with recurrent pleural effusion and fever not associated with Weber-Christian disease.

4 Conclusions

Idiopathic pleural panniculitis is a very rare occurrence because in the literature are reported only few cases of pleural involvement secondary to idiopathic pulmonary panniculitis, as in the case of Weber-Christian disease or cases of benign pleural lipoma, in which is frequent the involvement of the mediastinum, lung, bronchus, but, rarely, however, the only involvement of the parietal pleura. The presence of pleural panniculitis form with parietal pleural thickening of adipose tissue, without pathological aspects of mesothelial cells, and the presence of lymphocytic infiltrates within the adipose tissue, in a patient with no cutaneous or visceral lesions or signs of systemic inflammatory disease of the adipose tissue, is an occurrence not reported so far in the literature.

Conflict of interest: The authors declare that they have no conflict of interest or any financial support.

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