



## Case Report

# Mediastinal Pseudo-Tumor Tuberculosis Associated with Systemic Lupus Erythematosus

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**Abstract:** We report the singular association of mediastinal pseudo-tumoral tuberculosis [TB] with systemic lupus erythematosus [SLE]. D. I., 22 years old woman, is followed for a SLE since 2006 controlled by hydrochloroquine 'HCQ' 200mg twice/day and a thyroiditis of Hashimoto under levothyrox 125 mg/d. It does not relate to tuberculosis. She is hospitalized in pneumology for progressive dyspnea and chest pain. Objective clinical examination shows a conserved general condition, a pyrexia T° to 37, BMI to 21.1 (without notion of loss of weight), heart beat 94 / min, respiratory rate at 24 cycle/min and blood pressure at 110/60 mmHg. The pleuropulmonary and cardiovascular examination were without abnormality and thus the remainder of the clinical examination. The image of the thoracic face fails with a right para-cardiac opacity erasing the right edge of the cortex and the CT concludes to a mediastinal tumoral process, filling the gutter vertebral and extending from T6 to T10, with tissue density presenting areas of necrosis within it. Bronchial fibroscopy finds a thickening of inter-lobar spurs and bronchial biopsies identifies a bronchial mucosa site of chronic inflammatory remodeling with presence of multi-nucleated giant cells suggesting a tuberculoid process. The parameters of the hemogram were normal and the inflammatory syndrome was attested by both the increase ESR at 54 mm and the level of CRP at 12 mg/l. The smears, negative on direct examination, will prove positive for culture on Lowenstein medium. Antituberculosis treatment has been prescribed with good clinical, bacteriological and radiological progress. Long-term asymptomatic mediastinal pseudo-tumoral tuberculosis can be revealed at the compressive stage by dyspnea and chest pain. The fear of lymphoma requires biopsies with pathological examinations which sometimes reveal benign affections (tuberculosis, sarcoidosis in their tumor forms) which can enamel the autoimmune disorders characterizing a SLE.

**Keywords:** Mediastinal Pseudo Tumor, Tuberculosis, Systemic Lupus Erythematosus

## 1. Introduction

Systemic Lupus Erythematosus (SLE) is an autoimmune systemic disorder of unknown cause with various manifestations involving multiple organ systems. Genetic factors include specific genes and chromosomal defects, and hormonal and environmental factors, such as vitamin D, are

well-known immunomodulators of disease [1] [2].

Infections are the second leading cause of death in SLE patients (25%), immediately after the complications related to disease activity (26%) [3-6] and concerned most commonly tuberculosis with significant morbidity and mortality in endemic region, with more frequent and more extensive extra - pulmonary involvement [6-10].

## 2. Aim

To report a case of young female followed for SLE, 7 years ago, investigated for breathlessness and chest pain. Many diagnoses were initially evoked (pulmonary embolism, infection, shrinking lung syndrome ...) but the investigations (imaging, microbiology) established a rare and special diagnosis of pseudo-tumoral form of mediastinal tuberculosis.

## 3. Materials and Methods

### Case Report

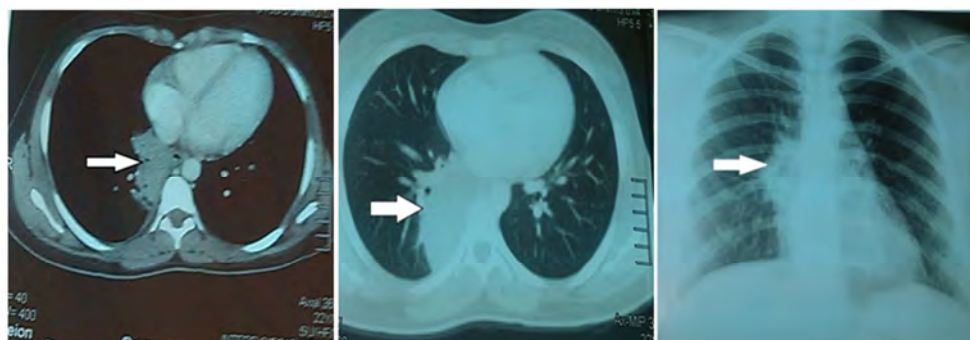
DI, a 22 -year-old female had been diagnosed with SLE since 2006. Her main symptoms consisted of polyarthritis, photosensitivity, oral ulcers, leukopenia, thrombopenia and positive antinuclear antibodies (ANA and anti-DNA). The SLE was stabilized for long time by low dose of corticosteroids (7.5 mg/day) and hydroxychloroquine (Plaquenil) as well for the Hashimoto thyroiditis which was controlled by 125 mg /d of Levothyroxine.

She is explored in emergency department with a 2-week history of worsening chest pain associated with positional dyspnea, dry cough and dysphagia. He had no significant past

history of medication and denied any significant family history and notion of contagious tuberculosis.

On physical examination, the general condition of the patient was conserved with apyrexia temperature at 37°C, BMI to 21.1 (without recent loss of weight), CF to 94/min, RF to 24 cycle /min and blood pressure at 110/60 mmHg. There was no lymphadenopathy. Cardiovascular, respiratory and abdominal examinations were normal. There was no evidence of arthritis, rashes, alopecia or oral ulcers evoking involvement of SLE. The SLE Disease Activity Index (SLEDAI), measured at the diagnosis time of tuberculosis, was estimated at 11.

Chest radiography illustrated fails with a right para-cardiac opacity erasing the right edge of the cortex subsequent computed tomography of the thorax with contrast, revealed a mediastinal tumoral process, filling the gutter vertebral and extending from T6 to T10, with tissue density presenting areas of necrosis within it. Bronchial fibroscopy finds a thickening of inter-lobar spurs and bronchial biopsies identify a bronchial mucosa site of chronic inflammatory remodeling with presence of multi-nucleated giant cells in favor of a tuberculoid process. Echocardiography was normal (no pericardial effusion).



**Figure 1.** The image of the thoracic face fails a right para-cardiac opacity erasing the right edge of the heart. CT concludes a mediated tumor process, bridging the vertebral gutter and extending from T6 to T10, tissue density with areas of necrosis within it (arrow).

### Laboratory Values:

Hemogram showed level of hemoglobin at 9.1 g/dl, with MCV at 80.8 fl and MCHC at 30.4 g/dl; we noted a mild leucopenia with lymphopenia (total leukocytes, 3,000 / mm<sup>3</sup>; lymphocytes, 700 / mm<sup>3</sup>) and the level of platelets at 229 x 10<sup>9</sup>/l. The erythrocyte sedimentation rate (ESR) was at 64 mm/hr, a C-reactive protein within the normal range, an elevated lactate dehydrogenase of 455 U/L (normal range, 120 to 240 U/L), and a serum calcium of 7.2 mg/dL. Renal function tests were normal (serum creatinine, 0.7 mg/dL; creatinine clearance, 102 mL/min), and microscopic urinalysis was also without abnormalities. Total serum protein and albumin were decreased (5.53 g/dL and 2.36 g/dL), and 24-hour urinalysis showed a proteinuria of 10 g/24 hours. Total cholesterol was approximately at 250 mg/dL. The serum albumin was at 18 g/l [normal range (NR) 35-50]. Livers tests showed alkaline phosphatase at 207 IU/l (NR 30-300), alanine aminotransferase (ALT) at 34 IU/l (NR 5-35), aspartate aminotransferase (AST) at 28 IU/l (NR 5-35), gamma-glutamyltranspeptidase (γGT) at 23 IU/l (NR 11-51),

total cholesterol at 5.9 mmol/l (NR 3.6-5.2), triglycerides at 3.5 mmol/l (NR 0.4-1.5) and normal coagulation profile; viral hepatitis serology was negative. Serum iron at 54 μmol/l (50-175); vitamin D at 11.1 ng / ml (30-60) ng/ml. The thyroid tests showed level TSHus at 48.48 U ui/ml (0.2-4) FT4 at 15.44 pmol/l (8.02-24.5). Antibodies against double stranded DNA were positive at 454 IU/mL (0-7) and we observed hypocomplementemia (C3, 33 mg/dL (80-180) but C4 was normal, 22mg/dL (10- 40). The smears, negative on direct examination, will prove positive for culture on Lovenstein medium.

## 4. Results and Discussion

### 4.1. Management and Out-come

Antituberculosis treatment has been prescribed for 6 months – referring to the local recommendations- with good clinical, bacteriological and radiological progress (Figure 2) without any adverse effects referring to the clinical context

(not acute SLE observed particularly).



**Figure 2.** Complete cleaning of the lung lesion after 06 months of treatment.

#### 4.2. Discussion

*Mycobacterium tuberculosis* undergoes lympho-heamatogenous dissemination after entering the respiratory tract. Typically the mediastinal and hilar lymph nodes are the first lymphatic tissues. *M. tuberculosis* will encounter and it may then spread to other organ systems [11] [12].

In immunocompetent individuals, tuberculum organisms are ingested by macrophages, which subsequently present the antigen load to T-lymphocytes. Macrophages are activated through the actions of T-cell-mediated antibody production enabling them to destroy the bacilli [13].

What factors predisposed Systemic Lupus Erythematosus patients to infection? (table 1)

The susceptibility to infections in SLE patients may be explained by several intrinsic and acquired defects in the immune system, related to the disease itself or to immunosuppressive [5]. These include various mechanisms as impairment of B lymphocytes activity, intrinsic defects in the innate immune function such as defects in chemotaxis, abnormalities in phagocytosis, decreased immune complex clearance, and delayed hypersensitivity [13-16]; lymphocytes from SLE patients are responsive to suppressor factor produced by normal adherent cells treated with *M. tuberculosis* and SLE lymphocytes so treated will suppress lymphocyte activation to PHA and a variety of mitogens and antigens. Adherent cells from SLE patients, however, when treated with *M. tuberculosis* were unable to produce the factor which would activate either normal or SLE suppressor lymphocytes [13] [17].

In addition, deficiencies in early and late complement components, and carriage of variant alleles of the mannose-binding lectin, predispose SLE patients to infections [8] [16] [18-20]. The most important factors suggesting community acquired infections in SLE patients are immunosuppressive therapy (intravenous pulses of

methylprednisolone and intravenous monthly cyclophosphamide) [5] [7] [10] [17] [19] [20] and the most recent treatment of autoimmune diseases as biotherapy (anti-CD20, anti-TNF...) [20] [21] while antimalarial drugs seem to have a protective effect [5] [19] [20].

**Table 1.** Risk factors for infection [4] [10].

| Leukopenia                    |
|-------------------------------|
| Acquired hypocomplementaemia  |
| Genetic complement deficiency |
| MBL deficiency                |
| Hypogammaglobulinaemia        |
| Splenectomy                   |
| Functional hyposplenism       |
| Prednisolone dose             |
| Immunosuppressive medication  |
| Biologicse. g., rituximab     |

The Mediastinal masses are routinely encountered in clinical practice and a wide range of differential diagnoses must be considered (lymphoma; sarcoidosis) [17]. A less common cause is tuberculosis, a disease recognized for its atypical presentations attributed to its ability to affect almost any organ system ( anterior mediastinal mass)[12] particularly in this context (autoimmune disease, long time of corticosteroid therapy, lymphopenia and tuberculosis endemic area).

#### 5. Conclusion

As it happened with SLE patients, the clinical suspicion of TB is hindered by several factors. On the one hand, the atypical presentations of TB, in a military pattern, or with mediastinal lymphadenopathy, or as extra pulmonary disease, pose a great diagnostic challenge, since these presentations may point to a bacterial etiology or to other diseases such as lymphoma.

Mediastinal lymphadenopathy is rare in post-primary tuberculosis. Regardless of this rarity, the occurrence of mediastinal pseudo tumor in clinical history of no-smoker SLE, physician always do to think of tuberculosis in the differential diagnosis especially in an endemic area of tuberculosis.

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