Adult-onset Still's disease revealed by a pleuropericarditis


ABSTRACT: We report a case of adult-onset Still's disease (AOSD) revealed by pleuropericardial manifestations. A 40 yr old black woman was admitted for flu-like syndrome with pharyngitis, hectic fever, polymorphonuclear hyperleucocytosis and pleuropericarditis. The diagnosis of AOSD was supported by 3 major and 3 minor criteria after exclusion of Infectious, haematological and connective tissue diseases.

Pulmonary involvement is infrequent in AOSD, and consists of transient pulmonary infiltrates and chronic restrictive pattern. However, pleuritis, like pericarditis, is present in 25% of cases. Initial onset of pleuritis, associated with fever and hyperleucocytosis preceding articular manifestations could be responsible for a delay in diagnosis and a subsequent worsening in the prognosis of the disease. A rapid improvement is usually observed under nonsteroidal anti-inflammatory drug or corticosteroid treatment.


Adult-onset Still's disease (AOSD) is an uncommon connective tissue disease with symptoms similar to those described in children with Still's disease or systemic juvenile rheumatoid arthritis [1-3]. Three major symptoms are usually present: high-grade spiking fever, evanescent erythematous or salmon-coloured maculopapular rash involving the trunk and extremities and articular manifestations usually polyarthritis [3, 4]. However, a fever of unknown origin (FUO) can be the initial symptom and is sometimes associated with infrequent visceral involvement such as pleuropericarditis. This latter presentation suggests numerous diagnoses, namely infectious, neoplastic or connective tissue diseases. The difficulty of the diagnosis is illustrated by the present case report.

Case report

A 40 yr old Haitian woman was admitted on August 19, 1988, for a flu-like syndrome associated with a mediastinal chest pain. Her past medical history included an essential epilepsy treated with phenobarbital.

Since August 10, 1988, the patient suffered from headache, myalgias, sore throat and dry cough. She was treated with erythromycin (2 g qd). Eight days later, she developed a high grade fever (39°C) and a mediastinal chest pain. On admission, she was asthenic and had a recent 2 kg weight loss. The temperature chart (fig. 1) showed a 40°C spiky fever occurring in the late evening. Pulse rate was 90 per min, and arterial pressure 110/60 mmHg. The patient a pharyngitis involving several cervical lymph nodes. Cardiovascular and pulmonary examination was within normal limits. Arterial oxygen tension (PaO2) was 8.66 kPa and arterial carbon dioxide tension (Paco2) 4.6 kPa (room air). Chest X-rays showed a small bilateral pleural effusion and a mild cardiomegaly. An echocardiogram revealed a moderate pericardial effusion. Thoracic computed tomographic (CT) scan confirmed pleural effusion without interstitial feature. purified protein derivative (PPD) skin test was negative. Haemoglobin was 9.1 g·dl-1, white blood cells (WBCs) 14,000 per μl, 87% polymorphonuclear cells (PMNs),
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erythrocyte sedimentation rate (ESR) 110 mm-first hr
declared a clear yellow fluid with protein level 37 g·l², 2,000 cells-per µl with
95% nonimpaired PMNs and 5% lymphocytes. Fingerprint bronchoscopy, bronchial biopsies, and
bronchoalveolar lavage (10x10³ cells·ml⁻¹, 95% macrophages) were normal. Lymph node and bone
marrow biopsies disclosed nonspecific process.

Pleurophtericitis associated with hectic fever, hyperleucocytosis, pharyngitis and cervical lymph
nodes suggested infectious or immunological diseases. Thus, microbiological studies, including acid-fast
bacillus(AFB) were performed on blood, pleural and
bronchoalveolar fluid analysis (6 cases) demonstrate a clear to
cloudy, yellow exudate with a mild cellularity, mostly
neutrophils; glucose level is usually within normal limits
and contrasts with the low level observed in rheumatoiid arthritis pleural effusion. In two cases, pleural
histology has shown a nonspecific acute inflammation
[2, 7]. A complete recovery is usually achieved with
anti-inflammatory agents but clearing can occur
spontaneously. Pleuritis does not appear to be a worse
prognostic criteria. Parenchymatous lung involvement
has only been reported in 11 cases [2, 4, 7-11]. Clinical
manifestations are rare and nonspecific; an acute
respiratory failure is reported in two cases [7]. Usually,
pulmonary manifestations consist of radiological
findings, namely pulmonary infiltrates involving the
lower lobes. These infiltrates are eventually sensitive to
salicylates or corticosteroids but radiological recurrence
is possible. A chronic progression appears to be
uncommon. CORBETT et al. [10] reported a patient with
a chronic cough and exertional dyspnoea revealing a
left lower lobe infiltrate. Symptoms were associated with
a severe corticoresistant restrictive defect. Such a
restrictive pattern is mentioned in 7 out of the 8 AOSD
patients studied by TROUM et al. [11]. Three patho-
logical reports showed nonspecific findings: an acute
alveolitis [11], a nonspecific chronic inflammatory

Discussion

Since the first description of AOSD in 1971 by
BYWATERS [1] and BUJAX et al. [2], about 250 cases have been reported [3-5]. The diagnosis of AOSD can be
readily done when the three main symptoms are present
in association with hyperleucocytosis (60-70%). Other
clinical manifestations include peripheral lymph node
enlargement which appears in approximately 50% of
cases, sorethroat (40%), splenomegaly (42%),
hepatomegaly (27%) with normal or abnormal liver
enzymes (30%), pericarditis (30%) or even carditis (3%) 
[3, 4, 6].

Pleurisy is revealed by chest pain or by systematic
chest roentgenogram. In most of the patients, pleural
effusion is bilateral and frequently associated with
pericarditis. As observed in our patient, pericardial and
pleural fluid analysis (6 cases) demonstrate a clear to
crushed, yellow exudate with a mild cellularity, mostly
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Table 1. – Adult-onset Still's disease: diagnostic criteria

<table>
<thead>
<tr>
<th>Major criteria</th>
<th>Minor criteria</th>
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</thead>
<tbody>
<tr>
<td>Hectic fever &gt;2 wks</td>
<td>Arthragias</td>
</tr>
<tr>
<td>Cutaneous rash</td>
<td>Myalgias</td>
</tr>
<tr>
<td>Arthritis</td>
<td>Pericarditis</td>
</tr>
<tr>
<td>Hyperleukocytosis &gt;12,000-per µl (x2)</td>
<td>Increased GOT, GPT serum levels</td>
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<tr>
<td>Past history of childhood Still's disease</td>
<td>Pharyngitis</td>
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</tbody>
</table>

Exclusion criteria

Microbiological identification
Antibiotic therapy efficacy
Other connective tissue disease or malignancy

The diagnosis is assessed by 4 major criteria or 3 major and 3 minor criteria and
absence of exclusion criteria [3]. GOT: glutamic oxalo-acetic transaminase; GPT:
glutamic pyruvic transaminase.
infiltrate with patchy interstitial fibrosis [10] and some epithelioid granulomas in one case [7]. Bronchoalveolar lavage showed a mixed alveolitis (lymphocytes 15%; neutrophils 16%; eosinophils 6%) in one case [7] and was normal in our patient.

These pleuropulmonary manifestations can be related to AOSD when other criteria are present (table 1). Infections and malignancy as well as connective tissue diseases, i.e. vasculitis, systemic lupus erythematosus, rheumatoid arthritis or sarcoidosis, have to be excluded.

Treatment of the respiratory manifestations is based upon anti-inflammatory agents. In 20% of cases, salicylates or nonsteroidal anti-inflammatory drugs give improvement, but most of the patients require corticosteroids at dosage of 1–2 mg·kg⁻¹ body weight qd. In those patients requiring prolonged high-dose corticosteroid treatment to control the disease, slow-acting or sparing-agent can be required. Antimalarial agents, gold salts, penicillamine or immunosuppressive agents such as cyclophosphamide, azathioprine, or methotrexate [3] have been used.

Respirologists should be aware of this condition of AOSD which associates pleurisy, fever and hyperleucocytosis, all symptoms leading us to rule out infectious and other causes of chronic FUO. Therefore, diagnosis criteria of AOSD could enable clinicians to avoid recourse to invasive procedures.

References

Maladie de Still de l’adulte révélée par une pleuro-péricardite.
RÉSUMÉ: Une femme de race noire, âgée de 40 ans, a été hospitalisée pour un tableau clinique associant un syndrome grippal avec pharyngite, une fièvre hémicte avec hyperleucocytose et une pleuro-péricardite. Le diagnostic de maladie de Still a été porté sur la présence de 3 critères majeurs et de 3 critères mineurs, après avoir exclu une infection systémique, une maladie hématologique ou une connectivité définie. Les manifestations pulmonaires sont rares au cours de la maladie de Still et comportent des infiltrats pulmonaires transitoires et une atteinte restrictive. Cependant, pleurésies et péricardites sont observées dans 25% des cas. Une pleurésie révélatrice, accompagnée d’un syndrome fébrile et d’une hyperleucocytose avant l’apparition de manifestations articulaires, peut être responsable d’un retard diagnostique à l’origine de complications. Les anti-inflammatoires non stéroïdiens et surtout la corticothérapie générale ont habituellement un effet rapidement favorable.