**Immunologic Lung Disease**

**Delphine Gamondès and Didier Revel**

**CONTENTS**

12.1 Introduction 197
12.2 Pulmonary Manifestations of Systemic Disease 197
12.3 Cardiac Involvement in Systemic Immunologic Diseases 198
  12.3.1 Pericardial Disease 198
  12.3.2 Valvular Heart Disease 198
  12.3.3 Myocardial Disease and Coronary Artery Disease 200
12.4 Conclusions 202
   References 202

**12.1 Introduction**

Immunologic lung disease regroups several entities with unknown origin, with pathophysiology involving an auto-immune process, and lesions (infiltrates or necrosis) that are often related to small- or medium-size vessels (vasculitis) and a biological profile with some immunological alterations, such as circulating immune complexes, antibodies, etc. Besides the lung, they can involve with a particular predilection the joints, the kidneys, the skin, the brain and, more particularly, the cardiovascular system. The prognosis is variable from one entity to the other, but the cardiovascular system is a major target in terms of life prognosis.

In this chapter we will concentrate on the more frequent systemic lung disease with cardiac involvement (pericarditis, myocarditis, valvular abnormalities) and in particular ischemic coronary events occurring very often at a younger age than in the normal population. The contribution of MDCT combining a lung parenchymal study with an examination of the cardiovascular system during the same procedure will be discussed.

**12.2 Pulmonary Manifestations of Systemic Disease**

Lung involvement in systemic disease may be a manifestation of the underlying pathological process, may be a complication of the underlying disease or may be related to the treatment. Imaging and in particular CT (HRCT) play a central role for the evaluation of lung involvement especially when clinically suspected. This aspect, which is beyond the scope of this chapter, has been extensively covered over the last 10 or 15 years in the international literature.

HRCT has several established roles:

- It may demonstrate subtle findings of pathology when the chest radiograph appears normal in patient studied for immunologic lung disease.
- It may participate in narrowing the differential diagnosis in case of overlapping immunologic syndrome.
- It may help to assess the treatment response by acquiring comparative examinations during follow-up.

Aside from the well-established role of HRCT in studying lung parenchyma in these various disor-


12.3 Cardiac Involvement in Systemic Immunologic Diseases

It is now well known that the heart is a frequent target of several systemic autoimmune diseases with associated lung involvement. The more frequently encountered entities are in particular systemic lupus erythematosus (SLE), rheumatoid arthritis (RA) and progressive systemic sclerosis (PSS) (Roman and Salmon 2003), although cardiac involvement could also be observed during several vasculitis such as Churg-Strauss syndrome (CSS) (Pela 2006).

Cardiac involvement is considered common with a variable frequency from one entity to the other and is given a high rank among the causes of morbidity and mortality. There is a common finding among all these systemic auto-immune diseases; regarding cardiac disease, all structures of the heart may be involved (Table 12.1).

**Table 12.1. Cardiac manifestations of several systemic diseases**

<table>
<thead>
<tr>
<th>Disease</th>
<th>Involvement frequency %</th>
<th>Pericardium</th>
<th>Endocardium valve</th>
<th>Myocardium</th>
<th>Coronary artery</th>
</tr>
</thead>
<tbody>
<tr>
<td>Systemic lupus erythematosus (SLE)</td>
<td>30–60</td>
<td>++</td>
<td>++</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Progressive systemic sclerosis (PSS)</td>
<td>7–15 50–70</td>
<td>+</td>
<td>0</td>
<td>++</td>
<td>++</td>
</tr>
<tr>
<td>Rheumatoid arthritis (RA)</td>
<td>1–10 30–50</td>
<td>++</td>
<td>+</td>
<td>0</td>
<td>+/-</td>
</tr>
<tr>
<td>Ankylosing spondylitis</td>
<td>40</td>
<td>+/-</td>
<td>++ (AI)</td>
<td>+</td>
<td>0</td>
</tr>
<tr>
<td>Systemic vasculitis</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Behcet</td>
<td>2–6 16</td>
<td>+</td>
<td>+/-</td>
<td>0</td>
<td>+</td>
</tr>
<tr>
<td>- Wegener and Chug-Strauss</td>
<td>10–15 6–44</td>
<td>++</td>
<td>+/-</td>
<td>+/-</td>
<td>++</td>
</tr>
<tr>
<td>- Takayasu</td>
<td>72</td>
<td>0</td>
<td>++ (AI)</td>
<td>++</td>
<td>+/-</td>
</tr>
</tbody>
</table>

12.3.1 Pericardial Disease

Pericardial disease, as a manifestation of serositis, is the most clinical cardiovascular manifestation (20 to 50%) (Cervera et al. 1992; Sturfelt et al. 1992) of SLE. Pericardial effusions may occur in the setting of active disease, but may be asymptomatic (Doherty and Siegel 1985). In case of mild pericardial effusions, typical electrocardiographic changes may be absent. Thus, the detection of a small amount of serositis with MDCT may be easier than with echocardiography (Fig. 12.1). In systemic sclerosis, limited to cutaneous disease, including CREST syndrome and diffuse cutaneous disease in which organ involvement (kidneys, lung, etc.) may be seen, pericardial disease is characterized histologically by chronic inflammatory changes. However, clinically important pericardial disease appears to be very rare (Hara 1990).

12.3.2 Valvular Heart Disease

Systemic auto-immune diseases are often complicated by structural or functional valvular abnormalities. Echocardiographic studies as well as autopsy