Rheumatoid resorptive arthropathy

P.V. Voulgari, S.C. Efremidis¹, A.A. Drosos

**Case presentation**
(P.V. Voulgari)
The patient was a 42-year-old, single woman with a six-year history of symmetrical polyarthritis. She presented to our outpatient Rheumatology Clinic with a poor grip. During the preceding years, as a result of an emotional disturbance, she had refused to visit any physician or to receive medication for her arthritis. Clinical examination revealed hypermobility of the finger joints. The digits appeared shortened and excessive skin folds were present. The phalanges could be inserted into one another and then be pulled out into abnormally long extensions (telescope-like action). A small subcutaneous nodule was also present over the extensor surface of the second digit of the right hand (Fig. 1). No signs of psoriasis were found.

Laboratory evaluation showed a hemoglobin (Hb) value of 9.1 gr/dl with features of anemia of chronic disease (low serum iron and normal ferritin levels). The C-reactive protein (CRP) was 42 mg/dl (normal value: < 6 mg/dl) and the erythrocyte sedimentation rate (ESR) was 65 mm/hr. Serum rheumatoid factor (RF) was positive at a titer of 1/640 (latex fixation test). Finally, HLA-typing revealed HLA A2-28, B12-17, DR4-2 alloantigens.

**Radiological findings**
(S.C. Efremidis)
Hand and wrist radiographs showed severe erosive arthropathy affecting the wrists, metacarpophalangeal (MCP), and proximal interphalangeal (PIP) joints bilaterally. More specifically: 1) the carpals of the right wrist were completely resorbed and the distal ulna presented a whittled appearance; 2) there were severe resorptive changes with whittled metacarpals and subluxations of the MCP joints; and 3) there were extensive erosive changes in the PIP joints. Osteopenia was also present (Fig. 2).

**Differential diagnosis**
(A.A. Drosos)
The differential diagnosis in this case included an aggressive untreated rheumatoid arthritis (RA), psoriatic arthritis (PSA) and scleroderma resorptive arthropathy.
throphathy. Reliable differentiation between RA and PSA in this patient was based on the absence of psoriatic rash and the absence of erosive changes at the distal interphalangeal (DIP) joints, with the characteristic pencil-in-cup deformity (1). In addition, the presence of RF and the subcutaneous nodule favored the diagnosis of RA. The absence of Raynaud’s phenomenon and skin changes despite her long-standing arthritis, as well as the absence of acrosclerosis, erosive changes of the distal tufts, or resorption of the soft tissues of the fingertips, and finally the absence of subcutaneous calcifications, ruled out the possibility of scleroderma as the underlying cause of the disease in this patient (1).

The common radiological findings in RA consist of fusiform soft tissue swelling, juxta-articular osteoporosis, diffuse joint space loss and marginal subchondral bony erosions. The most serious result of rheumatoid involvement of the hand is resorptive arthropathy, defined as severe resorption of the bone that begins at the articular cartilage and spreads along the diaphysis of the involved phalanges. Erosive damage in RA patients is known to occur early in the course of the disease (2). Factors which may influence disease progression and erosive changes are: RF, rheumatoid shared epitope, the levels of acute phase response proteins (APR), early treatment with disease modifying antirheumatic drugs (DMARDs) and the effectiveness of drug therapy. It is generally accepted that high titers of RF are associated with more severe disease and with the presence of extra-articular manifestations (EAM) including rheumatoid nodules (3). The APR, measured either directly by the determination of CRP concentrations or indirectly by the ESR, is a sensitive and objective indicator of both the activity and the extent of synovial inflammation (4). Studies have shown that persistent reductions in ESR and CRP values are accompanied by the slow progression of radiological lesions (5).

The association between human leukocyte antigen (HLA) class II genes and RA is well established. Many studies have demonstrated that RA is associated with the DRB1-encoded alleles HLA-DR4 and HLA-DR1 (6). Weyand et al. reported that the severity of RA, as defined by the EAM and destructive arthritis, was correlated with the “dose” of the rheumatoid epitope (7). In contrast, Boki et al. showed that only a minority of Greek RA patients carries the rheumatoid epitope and that DR4 is not associated with disease severity in these patients (8, 9).

DMARDs have been shown to be effective in suppressing synovitis and in delaying radiological disease progression in recent-onset RA (10). In addition, it seems that even in patients with established RA the early introduction of DMARDs is more effective than delayed treatment with second-line agents (11-13). Other factors which may influence the progression of RA are listed in Table 1 (14, 15).

Our patient suffered from a very severe form of RA. There were many contributing factors which led to the above described catastrophic resorptive changes: the long-standing untreated disease; the presence of EAM, like rheumatoid nodules and anemia of chronic disease; the high titers of RF; the high levels of APR; and finally the presence of rheumatoid epitope, indicating an unfavorable prognosis and outcome.

Table I. Factors determining the progression of rheumatoid arthritis.

<table>
<thead>
<tr>
<th>Early</th>
<th>Disease progression with symmetrical polyarthritis</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Systemic manifestations (low grade fever, fatigue)</td>
</tr>
<tr>
<td></td>
<td>Increased ESR, CRP</td>
</tr>
<tr>
<td></td>
<td>Positive RF</td>
</tr>
<tr>
<td></td>
<td>Presence of the “rheumatoid epitope” (DR4, DR1)</td>
</tr>
<tr>
<td>Later</td>
<td>Polycartilaginous involvement</td>
</tr>
<tr>
<td></td>
<td>Increased morning stiffness</td>
</tr>
<tr>
<td></td>
<td>RF &gt; 1/160</td>
</tr>
<tr>
<td></td>
<td>Anemia</td>
</tr>
<tr>
<td></td>
<td>Thrombocytosis</td>
</tr>
<tr>
<td>Definitive</td>
<td>Presence of rheumatoid nodules</td>
</tr>
<tr>
<td></td>
<td>Early radiological erosions of hand x-rays</td>
</tr>
</tbody>
</table>

ESR: Erythrocyte sedimentation rate; CRP: C-reactive protein; RF: rheumatoid factor.

Acknowledgments
The authors wish to thank Mrs. E. Horti for her secretarial assistance.

References
3. WOLFE F, SHARP JT: Radiographic outcome of recent-onset rheumatoid arthritis. A 19-year


