Rheumatoid disease and rib defects

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The presence of deformity of the ribs has been an infrequently reported finding in chronic rheumatoid arthritis.* Examination of the radiographs of patients attending our Rheumatoid Clinic suggests that this deformity may be more common than at present appreciated. The benign nature of this lesion is stressed and in this report the clinical and radiological findings in eleven patients are evaluated.


Clinical material

Lesions in the ribs were observed as incidental radiological findings in two patients with rheumatoid arthritis. As a result, the chest radiographs of fifty patients selected at random from the Rheumatoid Clinic were reviewed by one of us (W.M.P.). This search, together with close scrutiny of outpatients attending the clinic, revealed a further nine patients with rib lesions. Common aetiological factors were sought among these patients and relevant clinical data are tabulated. (Table I.)

Table I

Clinical features of eleven rheumatoid patients with rib lesions

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Sex</th>
<th>Age (yrs)</th>
<th>Diagnosis</th>
<th>Duration of arthritis (yrs)</th>
<th>Functional capacity (grade)*</th>
<th>Differential fixation test</th>
<th>Latex fixation test</th>
<th>Nodules</th>
<th>Shoulder involvement</th>
<th>Steroids given</th>
<th>Body build</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>51</td>
<td>RA</td>
<td>3</td>
<td>1/64</td>
<td>+</td>
<td>-</td>
<td>Moderate/severe restriction R &gt; L</td>
<td>Nil</td>
<td>Medium</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>51</td>
<td>RA</td>
<td>27</td>
<td>1/16</td>
<td>Neg.</td>
<td>-</td>
<td>Almost fixed R &gt; L</td>
<td>Nil</td>
<td>Medium</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>61</td>
<td>Psoriatic arthritis</td>
<td>25</td>
<td>1/16</td>
<td>Neg.</td>
<td>-</td>
<td>Moderate/severe Equal</td>
<td>7 yrs</td>
<td>Obese</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>52</td>
<td>RA</td>
<td>36</td>
<td>1/32</td>
<td>Neg.</td>
<td>-</td>
<td>Severe Equal</td>
<td>Nil</td>
<td>Medium</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>21</td>
<td>Still's disease</td>
<td>5</td>
<td>1/16</td>
<td>Neg.</td>
<td>-</td>
<td>Moderate/severe R &gt; L</td>
<td>Nil</td>
<td>Thin</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>61</td>
<td>RA</td>
<td>10</td>
<td>1/128</td>
<td>+</td>
<td>+</td>
<td>Fixed</td>
<td>Nil</td>
<td>Thin</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>63</td>
<td>RA</td>
<td>8</td>
<td>1/32</td>
<td>+</td>
<td>-</td>
<td>Fixed</td>
<td>Nil</td>
<td>Thin</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>72</td>
<td>RA</td>
<td>10</td>
<td>1/32</td>
<td>+</td>
<td>-</td>
<td>Almost fixed</td>
<td>Nil</td>
<td>Thin</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>55</td>
<td>RA</td>
<td>20</td>
<td>1/128</td>
<td>+</td>
<td>+</td>
<td>Fixed</td>
<td>4 yrs</td>
<td>Thin</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>F</td>
<td>67</td>
<td>RA</td>
<td>15</td>
<td>1/1,024</td>
<td>+</td>
<td>+</td>
<td>Fixed</td>
<td>Nil</td>
<td>Thin</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>F</td>
<td>60</td>
<td>RA</td>
<td>10</td>
<td>1/32</td>
<td>Neg.</td>
<td>-</td>
<td>Moderate/severe</td>
<td>10 yrs</td>
<td>Obese</td>
<td></td>
</tr>
</tbody>
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* Graded 1 to 5, Grade 1 representing normal functional activity and Grade 5 bedridden.
Review of the literature

In a paper on the pelvic lesions in rheumatoid arthritis, Martel and Duff (1961) mentioned, as an incidental finding, bone absorption at the posterior ends of the ribs in one patient close to the costovertebral articulations.

During a discussion at the XII International Congress of Rheumatology, Dixon (1964) noted the occurrence of lesions on the superior margins of ribs in several patients. Because the aetiology of the underlying abnormality is ill-understood a wide variety of explanations has been offered (Alpert and Feldman, 1964; Keats, 1967; Sargent and others, 1969).

These lesions are not specific for rheumatoid arthritis and have also been described in scleroderma (Elke and Meier-Ruge, 1966), patients with poliomyelitis treated on a respirator (Bernstein, Loeser, and Manning, 1958), hyperparathyroidism (Noetzel and Steinbach, 1962), and Marfan's syndrome (Leak, 1966). Several mechanisms have been suggested but, so far as we have found, the only autopsy and histological findings which have been reported concern a patient with scleroderma.

Sargent and others (1969) classified these deformities on the basis of disturbances of osteoblastic and osteoclastic activity and considered the lesions in rheumatoid arthritis to be due to decreased or deficient bone formation. Pressure effect from rheumatoid nodule formation or granulation tissue has also been considered (Alpert and Feldman, 1964; Keats, 1967).

Elke and Meier-Ruge (1966) described the autopsy and histological findings in a patient with scleroderma. Changes similar to those of senile osteoporosis, but in a very severe form, were found. Interstitial pulmonary fibrosis was felt to be a contributory factor in this patient since torsion stresses on the affected ribs were aggravated by tachypnoea. Microscopically, lamellar bone formation was found in the outer rib margin with lacunar resorption at the inner and upper borders.

We feel that there is great similarity between our findings and those reported in patients suffering from poliomyelitis who require treatment for a long time on a respirator. The lesions in these patients, which are usually symmetrical, have been detected 20 months after the onset of treatment on the respirator (Bernstein and others, 1958). A generalized osteoporosis develops as a result of recumbency over a long period, and severe loss of muscle mass with restricted scapulohumeral and glenohumeral movement. The thoracic cage and mediastinal border of the scapula tend to lie in continuous apposition. It is postulated that the bone lesions in these patients result from pressure erosion of the scapula against the ribs in an already osteoporotic skeleton.

Similarly, Dixon (1964) suggested that the notches in rheumatoid arthritis might be due to recumbency and he further suggested the possibility of rheumatoid involvement of the gliding plane behind the scapula.

Related clinical findings in our patients

All the patients with rib lesions were female but since 84 per cent. of all of the cases reviewed were female these numbers are too few to be significant.

Most of the patients were middle-aged or elderly, but lesions were demonstrable in one patient aged 21.

Nine of the eleven patients had rheumatoid arthritis, classical or definite according to the criteria of the American Rheumatism Association, but one patient had psoriatic arthritis and another Still's disease. In most cases, the disease had been present for many years, but in two the duration of disease was 5 years or less.

Eight patients had a differential agglutination titre of 1:32 or more, but this was less than 1:256, except in one patient who had a titre of 1:1,024. Subcutaneous nodules were present in only three patients.

Ten of the eleven patients had severe impairment of functional capacity, mostly leading a chair/bed existence. In all cases, shoulder movement was either fixed or severely restricted and there was accompanying weakness and wasting of shoulder-girdle musculature. In addition, all but two patients were extremely thin and the two who were obese showed evidence of steroid side-effects. Six of the eleven patients had clinical kyphosis. Only three of the eleven patients were receiving steroids or had been treated with steroids within the preceding few years.

Ten non-rheumatoid patients who had undergone arthrodesis of the shoulder were reviewed and in no case were abnormalities of the ribs detected.

Radiological findings

The distribution of lesions in our eleven patients is presented in Table II. Of the thirty ribs affected, twenty were on the right, and the 3rd, 4th, or 5th ribs were involved in 25 instances.

Table II Radiological findings

<table>
<thead>
<tr>
<th>Rib no.</th>
<th>Side</th>
<th>Lesion</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>1</td>
<td></td>
<td>1</td>
</tr>
<tr>
<td>3</td>
<td>2</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
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<td></td>
</tr>
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<td>6</td>
<td></td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>7</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

The site of involvement tended to vary depending on the rib level affected. Most of the lesions tended to occur between 1 and 6 cm. from the costovertebral articulation. This corresponds to the area of most acute forward angulation of the rib.

We found three types of lesion. The commonest, occurring in 27 out of the thirty lesions, is rather...
difficult to describe or to illustrate (Fig. 1A). There is ill-defined bone loss which may result in one of two appearances. The rib outlines may remain intact but with obvious cortical thinning giving a vaguely hour-glass appearance. In other cases, localized bone loss may be so severe that the rib margins can no longer be identified. This latter feature was most prominent in Case 7, a patient who had suffered a long-standing hemiplegia in addition to generalized rheumatoid arthritis and persistently lay on her right side. A chest radiograph showed that the 3rd-7th ribs on the right were demineralized with loss of cortical outline. The 5th left rib was also very slightly affected. Autopsy radiographs revealed much more extensive change than had been previously appreciated and these will be described separately.

Localized deformity resulting in a slightly sigmoid downward curvature of the ribs and localized notching on the superior margin was much less frequent (Figs 2A and B); the former was found to affect seven ribs and the latter three.

Fracture of the rib was not observed on any occasion even in the most demineralized situations. None of the eleven patients had either lung granuloma or evidence of parenchymal disease.

In one patient, an interesting progression of changes was found after a period of 42 years. The initial radiograph, in 1964, showed a notch on the superior margin of the 3rd right rib and the 4th and 5th ribs on the same side were thinned (Fig. 2c). In Figs 2A and B, on subsequent examination in 1968, these ribs have developed a curious slightly sigmoid downward curvature with clearly defined notches on the 5th and 6th ribs on the left which were not previously present.

**Radiological comments**

Previous reports have stressed certain characteristic features which clearly differentiate deformities in the ribs of patients suffering from rheumatoid arthritis from the defects found in coarctation (Boone, Swenson, and Felson, 1964; Röser, 1928) and neurofibromatosis (Holt and Wright, 1948). These have mostly been described as occurring on the superior margins of the ribs and invariably close to the mid-line; they are often unilateral but if bilateral are not necessarily symmetrical. Any ribs from the 2nd to the 9th, i.e. those lying in close proximity to the medial border of the scapula, may be involved, but the 2nd, 3rd, 4th, and 5th are most frequently affected. It is interesting that no similar abnormality has been reported along the borders of ribs at other sites.

Only three ribs in our series showed clearly defined superior margin notches. In one patient, the 5th and 6th ribs were affected on the left side. The notch on the 6th rib lay in a line approximately 1 cm lateral to the defect on the rib above. This corresponded to the position of the medial border of the scapula. Radiographs taken with the arm resting by the side and abducted showed that the scapula glided in and out of these rib grooves quite readily. With the arm at rest, the scapula lay within the rib deformities.

It is not completely clear, although most of the deformity occurs on the external rib surface, why this should appear on a radiograph as deformity of the superior or even inferior margins. Perhaps this may be because in cross-section the rib has a triangular configuration with the apex pointing downwards and partly undergoes torsion to achieve forward angulation.

An antero-posterior radiograph of a normal rib shows that it has a well-defined upper cortical margin, but the lower edge is less well defined because of the overhanging subcostal groove. The frequency of reporting of superior marginal defects may simply reflect the relative ease of detection of local bone loss of the well-demarcated rib margin.

Because changes may be subtle, care must be taken not to overinterpret rib lesions in patients with anatomical irregularity of cortical thickness and rib diameter. Clearly defined measurable reduction of rib width over the affected segment has been suggested as a reliable guide to rib deformity (Sargent and others, 1969). It is clear that, although none of our patients had lung disease, minor rib changes could be obscured by overlying conditions such as honeycomb lung.

Finally, we have found that rib defects are more consistently shown on radiographs of the shoulder, and it is now our practice to include the posterior ends of the ribs when this region is being examined. Similarly, we have found that a chest x-ray taken in the antero-posterior rather than the more usual postero-anterior view will often show these defects more clearly, since the ribs are closer to the x-ray plate and there is less superimposition by the clavicles.

**Fig. 1A** Case 7. Antero-posterior chest radiograph. The 3rd to the 7th ribs on the right are thinned and the cortical outline is lost, particularly in the superior margin. The 5th left rib is also slightly affected.

**Fig. 1B** Case 7. Autopsy radiograph. On the right, the 2nd to 7th ribs show, in addition to thinning, curious 'cystic' and soap-bubble changes. The 5th left rib is thinned with local irregularity of cortex, but there are no cystic changes comparable to those on the right.
Rheumatoid disease and rib defects

Image 1A

Image 1B
Rheumatoid disease and rib defects

Pathological findings

One patient, Case 7, came to autopsy and the pathological diagnosis was recorded as 'chronic severe rheumatoid arthritis with cachexia and toxaemia from bed sores'. She was bedridden and had suffered a long-standing hemiplegia and persistently lay on her right side.

The posterior 8-10 cm. approximately of the right 3rd to 8th ribs inclusive and the 5th left rib were removed for further study. All were covered externally by variable amounts of soft tissue and internally by the pleura. The specimens were x-rayed in various positions, and transverse sections were made of the central part of each rib except the 4th and the 7th which were macerated. Sections in the long axis of each end of the 3rd, 5th, and 6th ribs were also prepared.

Examination of the radiographs (Fig. 3) and the macerated specimens (Fig. 5) showed that the essential change in all the ribs was a rather sharply demarcated loss of bone extending laterally for about 6-7 cm. from a point some 1-2 cm. anterior to the tubercle, the maximum loss being around the centre of this zone. The upper three ribs were more...
FIG. 3 Case 7. Isolated ribs. When radiographed from above, defects in the external margins of two ribs are seen to extend laterally from the tubercles. (Upper = 7th, middle = 4th, lower = normal 6th rib as control.)

FIG. 4 Case 7. Section of inferior margin of 4th right rib, showing perforation in costal groove. Haematoxylin and eosin. × 40

FIG. 5 Case 7. From above, downwards the external surfaces of the 7th and 4th ribs and a control rib (6th). The arrow points to the anterior edge of the zone of bone loss. × 2.
severely affected than the lower three and the 5th left rib was least affected. The changes occurred on the external surface and were most prominent in the normally thinnest part, the costal groove, though in some the upper border was also involved. Here the bone was reduced to a translucent shell containing perforations ranging from the barely visible to holes 3 mm. in diameter (Fig. 4). In the most severely affected 4th rib, the costal groove had virtually disappeared and bone loss from the upper border was also marked, so that this rib was only 1-2 mm. thick and 5 mm. wide (Fig. 5). In the macerated specimens and in the sections (Figs 3 and 6) it was evident that most of the bone loss had occurred from the external surface, though in some the upper border was also involved and loss of bone had occurred internally near the upper border of the 3rd rib.

Histologically, the 3rd to 8th ribs all showed evidence of osteoclastic subperiosteal erosion externally and degrees of periosteal new bone formation internally. Thus the overall effect was a rib consisting mainly of a thickened inner compact cortical shell (Figs 6 and 7). The erosive process was active in the 3rd rib but mainly inactive in the others, some of which showed evidence of attempts to repair the old scalloped surface. Externally there was no inflammatory cellular infiltration in the extraosseous soft tissues.

The 5th left rib showed least involvement. The outer cortex though eroded was largely preserved, so that apart from perforation of the costal groove the general shape of the rib was unaltered. This was the only rib without periosteal thickening of the internal face in the regions examined histologically.

In sections of the 5th left and 3rd and 6th right ribs, parts of a bursa with a fibrous wall but without inflammatory cellular infiltration were present, the appearances suggesting an old inactive bursitis (Fig. 8, overleaf).
strongly suggests local frictional forces between the scapula and the ribs. If this were the principal cause of the bursa, its persistence may, of course, be partly due to involvement by the coexistent rheumatoid disease, though there was no unequivocal evidence of this at necropsy. The thickening of the internal cortex is probably part of the adaptive remodelling induced by progressive loss of bone from the external surface. Aetiological comments All our patients were female and although most were middle-aged and had suffered from an inflammatory arthritis for many years, the rib lesions were also found in a young patient and in patients with disease of relatively short duration. It is noticeable that, in most cases, functional capacity was severely impaired; the patients had very restricted mobility and were generally thin with clinical evidence of muscle wasting. Kyphosis was also common in the group with rib lesions. Without exception, glenohumeral movement was severely restricted, equally in eight of the eleven cases but predominantly on the right side in the remaining three. Perhaps surprisingly, only one patient was strongly seropositive and only three had subcutaneous nodules.

We feel that persistent pressure of the scapula against the ribs is the most likely explanation of the lesions found in these patients. Certainly, in one, it was possible to elicit crepitus as the medial border of the scapula glided over the rib deformities. It is reasonable to suppose that muscle wasting, kyphosis, and gleno-humeral shoulder movement will aggravate this rib erosion in an already generally demineralized skeleton.

We were surprised that strong seropositivity and subcutaneous nodule formation did not occur more frequently. It is well known that such patients readily develop nodules at pressure sites. Although we have not demonstrated nodule formation in relation to these rib defects, it would be reasonable to suppose that this might occur in some patients.

In conclusion, it would appear that the main significance of this type of rib lesion lies in its recognition as a benign process. None of our patients complained of symptoms that were directly attributable to these lesions. It might be difficult in some instances to differentiate this from neoplastic disease if a subpleural lung nodule were present in close relation to rib defects.

Summary An analysis is presented of radiographic rib defects correlated with clinical findings in eleven rheumatoid patients. Histological and clinical features suggest that the rib defects are due to pressure effect from...
the scapula, and that they may be a relatively common finding in a mild form in patients with chronic rheumatoid arthritis who are thin and kyphotic with restricted shoulder movement.

References


