Sclerotic Changes of the Manubrium Sterni

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Abstract. Six females with nearly identical sclerotic and hyperostotic changes of the manubrium sterni are reported. Malignancies, bacterial inflammatory processes, and Paget disease, which were first suspected, could be excluded. The youngest patients also had sclerotic changes of other bones, including the lumbar spine, the pubic bone, and the clavicle, and may be classified as having “chronic recurrent multifocal osteomyelitis” (CRMO). The two oldest patients had ossification of the costoclavicular ligament(s) and may be classified as having “inter-sterno-costoclavicular ossification” (ISCCO). One had only hyperostotic and sclerotic changes as seen in “sterno-costoclavicular hyperostosis” (SCCH). The pathogenesis of these uncommon diseases is unknown, but they are all frequently associated with pustulosis palmo-plantaris and have similar clinical courses and laboratory abnormalities. None of the present patients had HLA-B27. The similarity of the radiological abnormalities of the manubrium sterni suggests that the diseases themselves may be similar, but with different courses depending on age, CRMO being present in children and young adults and ISCCO or SCCH in older adults.

Key words: Sternum – Sclerosis – Hyperostosis – Pustulosis palmo-plantaris – Multifocal osteomyelitis

Structural changes of the manubrium sterni and its vicinity, characterized by sclerotic reactions, have been described recently and termed “sterno-clavicular hyperostosis” (SCCH) [3, 4, 6, 7, 11–13]. This entity is rare in Caucasians, only 13 cases have been reported from Europe and America, but a similar condition appears to be more common in Japan [16], where initially it was termed “inter-sterno-costoclavicular ossification” (ISCCO) [15] and subsequently “pustulotic arthro-ostitis” (PAO) [16, 17]. Sclerosis of the manubrium sterni also has been described as part of the syndrome “chronic recurrent multifocal osteomyelitis” (CRMO) [10] which mainly occurs in children and young adults [2, 8–10, 14]. The purpose of the present investigation is to call attention to these sclerotic changes and to characterize and discuss the different forms.

Materials and Methods

During a 10-year period six females with sclerotic changes of the manubrium sterni were studied in our departments. The radiological investigations included one lateral view of the sternum and posterior-anterior tomographic sections through the sternum, the sternoclavicular joints and the sternal ends of the clavicles, together with routine studies of the chest, spine, sacroiliac, and peripheral joints. Biopsy and thorough clinical and laboratory investigations were performed in all patients. Five of these females were followed for a mean of 8.9 years (range 4–15 years) and were examined in detail at the end of the follow-up period.

Results

When first seen, two patients (Cases 1 and 6) had diffuse sclerotic changes of the manubrium sterni. Four had only sclerosis of the upper part, but at the end of the follow-up period all had a widened and diffusely dense manubrium with uneven, blurred, articular facets at the sternoclavicular joints. Some other characteristics are recorded in Table 1 and Figs. 1–6. None of the patients had spondylitis, sacroilitis, or peripheral joint changes.
Table 1. Some characteristics of the patients

<table>
<thead>
<tr>
<th>Case number</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at onset of the disease (years)</td>
<td>31</td>
<td>34</td>
<td>24</td>
<td>49</td>
<td>57</td>
<td>67</td>
</tr>
<tr>
<td>Duration of the disease when first examined (years)</td>
<td>0.5</td>
<td>5</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>0.5</td>
</tr>
<tr>
<td>Follow-up period (years)</td>
<td>4</td>
<td>9</td>
<td>8.5</td>
<td>8</td>
<td>15</td>
<td>0</td>
</tr>
</tbody>
</table>

Radiological abnormalities

Sclerotic changes:
- Manubrium sterni: + + + + + +
- Sternal body: + + + +
- Clavicle: +
- Vertebrae body: +
- Pubic bone: +

Ossification of:
- Synchondrosis sterni: + + + + +
- Costoclavicular ligament(s): + + + +
- First costal cartilage: + + + + +

Other diseases
- Pustulosis palmo plantaris: + + + +
- Psoriasis: +

Laboratory abnormalities:
- Elevated ESR: + + + +
- $\gamma$ globulin: +
- ASK: + (+)
- ASH: (++)
- Positive CFC: +

CFC = complement fixation test for clamydia; ( ) = slightly elevated

All patients had complained of constant discomfort in the upper anterior chest region and had experienced recurrent episodes of pronounced pain and some swelling in the region of the manubrium sterni. These episodes often occurred in cold and/or damp weather or in connection with infections. In association with these exacerbations one patient with additional sclerotic changes of the lumbar spine had low back pain, and another with sclerosis of the right pubic bone had slight discomfort in this region.

The course was prolonged over several years in the five females followed. Three patients had recurrent exacerbations of pustulosis palmo plantaris (PPP) often in connection with pronounced upper chest pain. One had psoriasis which progressed independently of the sternal disease. Low-grade fever was observed in association with an episode of pain in only one patient (Case 5).

In all patients surgical biopsy from the manubrium revealed spongiosclerosis, but no signs of malignancies. In one patient (Case 1) small areas with lymphocytes, plasma cells, and single eosinophil leucocytes were present in the bone marrow, but the other patients had no signs of inflammation and none had increased osteoclastic activity as seen in Paget disease. Bacteriological investigations were negative.

Moderately elevated erythrocyte sedimentation rates (ESR $\leq 52$ mm/h) were found in four patients, and ESR was permanently elevated in those followed. In all patients normal serum calcium and alkaline phosphatase levels were present. Tests for the histocompatibility antigen-B27 (HLA-B27) and rheumatoid factor were consistently negative.

Discussion

The sclerotic changes of the sternal manubrium observed in the present series of six females were virtually identical. All patients developed a widened and diffusely dense manubrium with uneven blurred articular facets at the sternoclavicular joints. Five had ossification of the first costal cartilage. None had hyperostosis of the clavicle, but one clavicle became sclerotic.

However, the patients were different in other respects. The two oldest females (Cases 5 and 6) had ossification of the costoclavicular ligament(s) and may be classified as having ISCCO [15]. This disorder always includes abnormal ossification in the region between the clavicle and the first rib. On the basis of X-ray findings it has been classified into three stages according to the extent of ossification. Stage 1: localized to the area of the costoclavicular ligament; stage 2: generalized with the interspace between the clavicle and the first rib filled with an abnormal ossified mass and irregularity of the inferior margin of the clavicle and the superior margin of the first rib; stage 3: hyperostotic with hyperostotic changes of the medial parts of the clavicles and in severe cases also hyperostotic affection of the manubrium. It has been claimed that the disease always progresses from stage 1 to 3 [15]. In Case 5 the disease began in the region of the costoclavicular ligaments, and 17 years later ossified plaques were present between the clavicles and the first ribs. In addition there were pronounced changes of the manubrium, but no hyperostotic or sclerotic changes of the clavicles. Case 6 had only slight ossification between the right clavicle and first rib, but pronounced changes of the manubrium. In both cases the radiological changes were thus not quite typical for ISCCO, but both patients had PPP in conformity with ISCCO/PAO [15-17].

Case 4 had no ossification of the costoclavicular...