Sternocostoclavicular hyperostosis: its progression and radiological features. A study of 12 cases

Peter Fritz, Gerhard Baldauf, Hans-Joachim Wilke, Irene Reiter

Abstract
Twelve cases of sternocostoclavicular hyperostosis were followed up over four to 16 years. The patients underwent repeated radiological examinations of the sternocostoclavicular joints and the sternum, and the extrasternal osseous manifestations of the disease were studied to show changes in the radiological features during long-term follow-up.

Five of 12 (41%) patients had extrasternal manifestations. With the exception of one patient extrasternal manifestations were first detected by scintigraphs because they were asymptomatic. With respect to the sternal manifestations the initial radiological diagnosis was made during an acute phase while painful swelling over the sternum and decreased mobility of the shoulders occurred. The radiological examinations showed the signs of a proliferative destructive arthritis in most patients. In contrast with the frequent occurrences of clinical symptoms, the radiological signs of progression take several years to become detectable.

There are no specific bacteriological, serological or histological findings. Usually a permanent increase in the erythrocyte sedimentation rate is found. Sternocostoclavicular hyperostosis is a slowly progressing disease, characterised by a chronic aseptic destructive sternoclavicular arthritis with a reactive low turnover sclerosis that begins in a similar way to an enthesopathy and ends after several decades with total ankylosis. The radiological identification of retrosternal proliferation of soft tissue by computed tomography was found to be a valuable criterion for the differential diagnosis from other benign hyperostotic processes of the sternoclavicular region.

In 1967, Sasaki reported the case of a patient with bilateral hyperostosis of the clavicles, interpreted as ‘osteomyelitis’, which was associated with pustulosis of the palms and soles (pustulosis palmaris et plantaris). In 1968, Kato et al. described a similar patient and in 1974 Sonozaki et al. reported four cases of symmetrical ossifications between the clavicles and first ribs.

In 1981, Sonozaki et al. published a review of the 1979 patient population of four Japanese dermatological clinics. From a total of 22 000 patients they found 12 cases of sternocostoclavicular arthro-osteoitis and 128 cases of pustulosis palmaris et plantaris; there was a 9.4% incidence of sternocostoclavicular arthro-osteoitis in patients with pustulosis palmaris et plantaris. By 1986, Japanese workers had published 147 reports of sternocostoclavicular arthro-osteoitis. In contrast by 1990 there were only about 80 published reports from European and American workers.

Koehler and coworkers were not aware of the earlier Japanese studies when they introduced sternocostoclavicular hyperostosis as a ‘previously unknown disease’. The clinical features of sternocostoclavicular hyperostosis are indicative of its relationship with reactive arthropathy and osteopathy in dermatological diseases such as acne conglobata, or pustulosis palmaris et plantaris and psoriasis.

Sternocostoclavicular hyperostosis has been referred to as an enthesopathy—that is, a disease of the insertions of tendons and ligaments. Some workers believe that sternocostoclavicular hyperostosis and chronic recurrent multifocal osteomyelitis represent one disease entity, the phenotype of which depends on the age at which the disease manifests itself. According to these workers, chronic recurrent multifocal osteomyelitis appears in infants and young adults, whereas sternocostoclavicular hyperostosis appears in middle life. Sternocostoclavicular hyperostosis and chronic recurrent multifocal osteomyelitis have the same hyperostotic changes in the sternum and clavicles, as well as additional foci of sclerosis in the vertebral column and pelvis. It has been suggested that sternocostoclavicular hyperostosis and chronic recurrent multifocal osteomyelitis are acquired hyperostotic syndromes.

At present, there can be little doubt that sternocostoclavicular hyperostosis is at the centre of a yet to be defined syndrome that is sometimes associated with dermatological and extrasternal osseous changes. Although the radiological features of sternocostoclavicular hyperostosis have often been described, the disease is relatively unknown and can be mistaken for cancer or another disease of the clavicles accompanied by sclerosis. There are few long-term studies on the progression of the disease.

This study emphasises the progression and accompanying radiological changes of sternocostoclavicular hyperostosis. In addition, the patients' clinical and serological findings are reviewed.

Patients and methods
From 1969 to 1984, 12 cases of sternocostoclavicular hyperostosis were detected at the...
Progression and radiomorphology of sternocostoclavicular hyperostosis were observed for extended periods of time, during which they underwent repeated radiological studies of the thorax. In all cases tomographs of the sternocostoclavicular joints and sternum, as well as scintigraphs, were repeatedly obtained. In two cases computed tomography scans of the upper aperture of the thorax were obtained (patients 3 and 9). In six patients, biopsy samples were taken of the sternocostoclavicular joints.

During the observation period, all patients underwent repeated evaluations of their erythrocyte sedimentation rates and levels of C reactive protein. Serum electrophoretic studies were repeatedly taken. All patients were checked for the HLA-B27 antigen, antinuclear antibodies, antibodies to DNA, and rheumatoid factors. In addition, serum enzyme and electrolyte levels were measured.

In 1990, a follow up examination was conducted on seven of the 12 patients. By that time one patient had died, another was ill, and three others were lost to follow up. As part of this examination, we obtained radiographs of the ribs, tomographs of the sternocostoclavicular region, and radiographs of extrameral osseous manifestations of the disease. All the serological and immunological studies conducted during the initial observation period were repeated.

This study is based on a retrospective review of the data gathered during the initial observation period together with the results of the 1990 follow up examinations.

Case histories

Eight men and four women were studied. All of them had had the first symptoms of sternocostoclavicular hyperostosis during the fourth and fifth decade of life. At the time of the 1990 follow up, the case histories were available for seven to 30 years (table 1). On average, patients had had symptoms for 15 years. It took a mean of 5-5 years after the first onset of symptoms before patients presented for thorough clinical testing. In our patients, the periods of clinical and radiological observation ranged from four to 16 years.

As Koehler et al did not describe the disease in Europe until 1975, and as the earlier Japanese studies were not known to them, a definitive diagnosis of the disease was not made before 1975. Even after 1975, due to the rarity of sternocostoclavicular hyperostosis, it took an average of 7-2 years after the completion of the initial radiological studies before sternocostoclavicular hyperostosis was diagnosed or even suspected. Before this, incorrect diagnoses such as tuberculosis of the bone, osteomyelitis, or cancer had influenced the therapeutic and diagnostic approach in three of our patients.

SYMPTOMS AND CLINICAL FINDINGS

Only one of our patients, who was in an early stage of sternocostoclavicular hyperostosis, was totally asymptomatic. The changes of the clavicles were a chance finding on a chest radiograph. For several years all the other patients had had intermittent episodes of pain over the sternum and the sternocostoclavicular joints when moving or on application of pressure. Sometimes the pain would radiate into the arms, shoulders, or the thorax. Four patients reported additional episodes of spontaneous pain, which typically occurred during damp or cold weather, or at night. Three patients described their pain as constant. One woman had an aggravation of her symptoms during her menstrual period. Characteristically, the pain occurred intermittently and was accompanied by local swelling and reddening over the sternum and the sternoclavicular joints, and an increase in the erythrocyte sedimentation rate. Six patients had a loss of mobility within the shoulder girdle; retroflexion and abduction were typically affected. As a result, these patients had difficulties when putting on a coat or when driving a car. One of the patients with constant pain was so incapacitated that he took early retirement. Another patient was temporarily forced to stop work and had to change jobs. Two patients developed a thoracic inlet syndrome due to occlusion of both subclavian veins. Finally, we noted dermatological changes in four of the 12 patients (table 1).

HAEMATOLOGY AND SEROLOGY

Eleven of the 12 patients showed a persistent increase in their erythrocyte sedimentation rates ranging from 15/35 to 110/143 mm according to Westergren. In all patients an aggravation of symptoms was accompanied by an increase in the erythrocyte sedimentation rate. In six patients we could show increased levels of C reactive protein or α globulin, or both. These

<table>
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<tr>
<th>Patient No</th>
<th>Case histories available (years)</th>
<th>Age when first symptoms appeared (years)</th>
<th>Mobility of shoulders</th>
<th>Pain in the sternum region</th>
<th>Cutaneous manifestation</th>
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<td>30</td>
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</table>

↓ = decreased, n = normal.
findings were, however, non-specific and irregular, and not clearly correlated with the course of the disease. Despite marked radiological changes, alkaline phosphatase levels were generally not increased or only minimally so. In no patient could we show rheumatoid factors, antinuclear antibodies, or antibodies to DNA. One patient was positive for the HLA-B27 antigen; this was assumed to be a chance finding.

HISTOLOGY
The biopsy samples taken from the sternoclavicular joints of six patients showed an increased osseous turnover characterised by an increase in the number of osteoblasts and osteoclasts, a thickening of the trabeculae of the cancellous bone, and formation of osteoid and fibrous tissue occasionally accompanied by a mild accumulation of granulation tissue and round cell infiltration.

In a few patients we noted proliferation of the capillaries. In patient 4 this proliferation extended from the bone marrow into the cartilage, leading to a cystic destruction of the cartilage. In patient 8 we noted a dense array of thickened trabeculae, relatively little osteoid, and an absence of osteoblasts. Clinically and radiologically this patient was in the final stage of sternocostoclavicular hyperostosis (stage III).

There was a correlation between the histology and disease activity in that the active phases were invariably accompanied by an increase in the number of osteoclasts and osteoblasts, round cell infiltrates, proliferation of capillaries and the formation of fibrous tissue. The histological work up of all patients gave a diagnosis of a non-specific osteosclerosis, which was tentatively attributed to ‘post-osteoarthritic changes’, ‘non-specific chronic inflammation with formation of scars’, ‘myelosclerosis’, or ‘post-trauma’.

RADIOMORPHOLOGY AND STAGING
The staging of the sternal manifestations of sternocostoclavicular hyperostosis is based on the work of Sonozaki et al.10 We have modified the basic staging system of Sonozaki et al to accommodate extrasternal manifestations. If there are extrasternal manifestations, the subscript E is placed behind the roman numeral indicating the stage. The subscript R is used to indicate involvement of more than the first ribs (table 2).

Stage I
Based on our own results and a review of previously reported work, we conclude that sternocostoclavicular hyperostosis usually begins with an ossifying costoclavicular tendo-ositis accompanied by a sternoclavicular arthritis which, at least initially, is non-destructive.

In this early stage the radiograph shows early calcification of the costoclavicular ligament in conjunction with an unusual sclerosis of the ligament’s clavicular insertion (fig 1A). At this stage the scintigram already shows a marked osseous turnover in the sternoclavicular joint.

Stage II
As the disease progresses, ossification extends to the medial end of the clavula, the sternoclavicular joint, the manubrium, and the body of the sternum. Usually, at the same time, progressive hyperostotic changes of the ventral parts of the ribs, the cartilage of the ribs, and the sternocostal articulations are noted (fig 2). As a rule, both first ribs are affected, but less commonly there are corresponding changes in ribs 2 to 7 which are either unilateral or bilateral. Interestingly none of our patients showed

<table>
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<th>Patient No</th>
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<th>Sternal manifestations (radiological stage)</th>
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Figure 1: Patient 4. Radiological diagnosis of sternocostoclavicular hyperostosis stage I in 1979. Since 1977 this patient had pains in the left shoulder girdle and permanently increased erythrocyte sedimentation rates. At the time of the first radiographs the patient presented with reddening and swelling of both sternoclavicular joints with pains caused by pressure and limited adduction of the left arm. The initial tomograph (A) shows calcification of the left sternoclavicular ligament and the beginning of sclerosis of the head of the clavicle (arrows). The bone scan (B) shows an accumulation of tracer in both sternoclavicular joints.
progression and radiomorphology

Stage I

Stage I represents the initial active stage of the disease. It is characterised by a pronounced remodelling of the sternoclavicular joints (fig 4) and by a marked destruction of the clavicles, the sternoclavicular joints, and the manubrium of the sternum. The extent of this destruction was not necessarily identical on both sides (figs 3 and 4). There was usually only minor destruction of the clavicular head. Figure 4 shows that the destructive changes sometimes extended to the sternoclavicular and sternocostal articulations.

Sternocostoclavicular hyperostosis remains in this intermediate stage for many years, characterised by a progressive destructive arthritis which in time leads to eburnation, dislocation, and total destruction of the joint. In general, the most pronounced ossifications are found in the region of the costoclavicular ligament.

Stage III

Stage III represents the final inactive stage of the disease. It is characterised by a total ankylosis of the sternoclavicular joints, the sternocostoclavicular hyperostosis, and ossification of the cartilage of the ribs (figs 5 and 6).

EXTRASTERNAL MANIFESTATIONS

Sternocostoclavicular hyperostosis occasionally coincides with sclerotic or partially destructive, or both, changes in the vertebral column, the bony pelvis or the sacroiliac joints. Several cases of sternocostoclavicular hyperostosis in conjunction with a peripheral arthritis have been noted.

Hyperostotic changes in the long tubular bones, often restricted to minimal periosteal reactions and ossification of the interosseous membrane of the lower thigh have been observed. Thirty four per cent of the patients reported by Sonozaki et al in 1981 had changes in the vertebral column and 32% had a peripheral arthritis. The acromioclavicular joint was most commonly affected. Based on radiomorphological criteria, the extrasternal skeletal manifestations of sternocostoclavicular hyperostosis can be divided into four subtypes: type 1, which resembles bacterial spondylitis or osteomyelitis of the pelvis (fig 4); type 2, which simulates ankylosing spondylitis; type 3, which resembles diffuse idiopathic skeletal hyperostosis; and type 4 which resembles sclerosis of the vertebrae or the bony pelvis, or both, without proliferative or destructive changes (figs 7 and 8).

The peripheral joints can be affected by a non-destructive mono- or oligoarthritis. In addition, unusual areas of sclerosis next to the sacroiliac joint, a sacroileitis resembling ankylosing spondylitis in conjunction with sclerosis, and erosions of the ileosacral joint and ossification of the ileosacral ligaments have been observed.
RADIOLOGICAL CHANGES
At the time of the initial diagnosis, most patients were classified as stage II. Despite observation periods of several years, only one patient reached stage III (total ankylosis of the sternoclavicular joints, the sternal synchondrosis, and ossification of the cartilage of the ribs) (table 2). In nine of 12 patients, however, we noted a slow progression of the sternoclavicular changes during the intermediate stage. This generally correlated with an increase in the extent of changes in ribs already affected and an extension of the disease to previously unaffected ribs (fig 2). The changes in the sternoclavicular joints did not represent only eburnation, but rather a progressive joint destruction in combination with an increasing hyperostosis of the sternum and the medial end of the clavicle. In only two patients were the costoclavicular ligaments unaffected. Owing to extension of the inflammatory process to the adjacent vasculature, we noted one case of unilateral and one of bilateral occlusion of the subclavian veins (patients 7 and 8; fig 5).

In five patients we observed extrasternal manifestations of the disease over a period of several years. Patient 3 initially presented with discovertebral destrucltions resembling those found in bacterial spondylitis (fig 4). The same patient also developed episodes of non-destructive monarthritis of the ankle joint associated with proliferation of the periost.

Patient 2 showed an unchanging sclerosis of four thoracic vertebrae (fig 7). In patient 7 the thoracic spine showed a hyperostotic spondylosis without changes of the radiological signs during follow up. Patient 9 noticed a slowly progressing painless stiffening of his vertebral column, beginning four years after the first sternoclavicular symptoms. Initial radiographs

Figure 4  Patient 3. Beginning of the disease (in 1980) with a painful swelling of the right sternoclavicular joint. Neck aches occurred two years later. The computed tomography examination from 1981 (A and B) shows the destruction of the sternocostoclavicular joints (arrows) and a right sternocostal joint (arrow) accompanied by a soft tissue tumour which was incorrectly diagnosed as a malignant tumour. The radiographic signs of the cervical spine (C) resemble those found in bacterial spondylitis or spondylodiscitis. Twenty-two (D) and 31 (E) months later progressing fusion of the vertebrae were seen. Biopsy samples taken from the sternocostoclavicular joint and the cervical spine only showed tissue from a non-specific chronic inflammation with the formation of scar tissue. No bacteria could be seen. The radiological diagnosis was stage II.
showed typical radiographic signs of a hyperostotic spondylitis, which 10 years later led to a complete fusion of the lumbar spine similar to ankylosing spondylitis. In patient 5 we observed a slowly progressing sclerosis of the bony pelvis without inflammatory changes of the sacroiliac joints (fig 8).

SCINTIGRAPHIC FINDINGS
The sternal and extrasternal manifestations of sternocostoclavicular hyperostosis showed an increase in the accumulation of tracer on the scintigraphs. With the exception of patient 3, however, the extrasternal manifestations were

Figure 6  Patient 9. Stage III sternocostoclavicular hyperostosis. Ankylosis of the sternoclavicular joints. Retrosternal ossifications and soft tissue proliferation (arrows) are seen.

Figure 7  Patient 2. Sternocostoclavicular hyperostosis, showing sclerosis of the vertebrae.
asymptomatic and were detected only from the scintigraphs. Radiographs subsequently taken tentatively established the diagnosis of sternocostoclavicular hyperostosis. The extent of increased tracer accumulation corresponded with the radiological stage of the disease. For instance, stage I patients showed increased tracer activity only in the sternoclavicular joints (fig 1B).

Discussion
Sternocostoclavicular hyperostosis is characterised by endosteal and periosteal bone formation and the proliferation of fibrous tissue that is partially destructive with subsequent ossification occurring in the area of the sternoclavicular joints and in other parts of the weight bearing skeleton. Similar changes have also been described in the long bones. At times sternocostoclavicular hyperostosis is associated with skin symptoms that resemble psoriasis, acne conglobata, or skin changes found with the various forms of seronegative spondylarthritis. At the centre of this yet to be defined syndrome, however, are changes in the sternum and sternoclavicular joints.

The increased erythrocyte sedimentation rate, histological findings and the proliferation of soft tissue in the area of the sternoclavicular joints indicate that sternocostoclavicular hyperostosis represents an enthesopathy—that is, a disease that begins with soft tissue lesions and ends with a destructive sternoclavicular arthritis with reactive low turnover sclerosis as indicated by a normal or minimally increased level of alkaline phosphatase. Although the sternoclavicular ligaments are affected first in most patients this is not uniformly so.

The concept that sternocostoclavicular hyperostosis represents a chronic, aseptic, tendoarthritis-ostitis is further supported by the fact that it is often associated with a spondylodiscitis or hyperostotic spondylitis, or both, in combination with an ossification of the longitudinal ligament or formation of bone structures resembling synodemosphytes. Furthermore, some patients develop a non-destructive peripheral arthritis during the course of the disease. No causative microbial agent has yet been identified. Consequently, we believe that sternocostoclavicular hyperostosis is primarily a rheumatic disease and not a bone disease.

The decisive criterion that distinguishes sternocostoclavicular hyperostosis from other benign hyperostotic processes of the sternoclavicular region is evidence of the retrosternal proliferation of soft tissues. Hence conventional radiology should be supplemented by computed tomography of the upper aperture of the thorax. If there is evidence of major soft tissue destruction, however, a biopsy sample is indicated to rule out cancer. Scintigraphic studies are always indicated when sternocostoclavicular hyperostosis is diagnosed or suspected to detect extrasternal manifestations.

Sternocostoclavicular hyperostosis is characterised by phases of inflammation leading to a progressive hyperostosis and eventual destruction of the sternoclavicular joints. The final stage—that is, the fusion of the sternoclavicular joints, is only reached after several years. At present only symptomatic treatment in the form of corticosteroids and non-steroidal anti-inflammatory drugs and analgesics is available.7-19

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