Acquired Hyperostosis Syndrome: Spectrum of Manifestations at the Sternocostoclavicular Region. Radiologic Evaluation of 34 Cases

W. DIHLMANN, S.W. DIHLMANN*

Summary  Thirty-four patients with chest wall hyperostosis, a condition which has been designated by various terms in the literature were evaluated radiologically. We prefer the name acquired hyperostosis syndrome (AHS), which we categorize into the complete, incomplete and possible form. In complete AHS, sternocostoclavicular hyperostosis is associated with axial and/or peripheral (endosteal, periosteal, enthesopathic, metaplastic) hyperostosis and with psoriasiform or acneform dermatosis. In addition, these three manifestations are accompanied by erosive or non-erosive peripheral and/or axial arthritis to a variable degree. Sometimes, concomitant findings which are consistent with ankylosing spondylitis are also to be found in the axial skeleton. AHS is manifested at 11 different sites on the anterior chest wall. Ossification forms of the costal cartilage, inflammatory enthesopathies (three different insertions) and focal hyperostoses as well as processes of remodelling of the ribs, clavicles and sternum which are described in detail have particular diagnostic significance. AHS can start simultaneously at one, two or several sites on the anterior chest wall. Conventional tomography (possibly supplemented by CT) is necessary for early diagnosis and for analysis of the various findings on the anterior chest wall.

Key words: Acquired Hyperostosis Syndrome, Sternocostoclavicular Hyperostosis, Ribs : Hyperostotic Foci, Inflammatory Enthesopathy : Costoclavicularis, Sternalis Superior, Retrosternalis.

INTRODUCTION

In 1967, the Japanese author Sasaki initiated the scientific discussion about a condition which he designated (in translation) as “bilateral clavicular osteomyelitis accompanied by palmar and plantar pustulosis”. Since then, about 350 cases of this condition have been published under various synonyms: sternocostoclavicular hyperostosis (SCCH) (40, 41, 27, 50, 48), inter sternocostoclavicular ossification with three stages (localized, generalized, hyperostotic) (57), pustulotic arthrop-osteitis (58, 59), pustulotic arthro-osteopathy (39), multiple thoracic hyperostoses (with unilateral sacroillitis) (3,4), hyperostotic pulumperisitic spondylarthritis (61), syndrome acne-pustulose hyperostose ostéite (SAPHO) (11), synovitis-acné-pustulos-hyperostosis-osteomyelitis syndrome (Sapho) (2), acquired hyperostosis syndrome (AHS) in its complete, incomplete or possible form (17,18) (Fig. 1 and Table I).

Some other suggested terms such as anterior chest wall syndrome (49) or juxtasternal arthro-osteitis (55,38) should be avoided. The former name characterizes a pain syndrome which can occur after myocardial infarction and also without coronary heart disease (58,8). In addition, costal lesions in the posterior chest wall which can also be detected radiologically have been described in AHS (3). The latter expression was introduced for differential diagnosis according to location and not for delimitation of the entity discussed here, namely SCCH, SAPHO, AHS etc.

Comparative clinical studies (including dermatological and radiological investigations) (5,28,31,35), indicate that sternocostoclavicular hyperostosis and chronic recurrent multifocal osteomyelitis (CRMO) (46,47,56) are related diseases with age-dependent differences. SCCH mainly occurs in middle-aged and elderly adults, CRMO in children, adolescents and young adults. SCCH and CRMO are thus very probably age-related synonyms.
Acquired hyperostosis syndrome

In 34 patients with AHS, the spectrum of manifestation in the anterior chest wall was analysed and the chronological manifestation sequence was determined in progress observations. Our radiological evaluation is a contribution to early and definitive correct diagnosis of episodic unilateral or bilateral pain in the specified region which is often weather-dependent and accompanied by local swelling with or without local erythema and/or local feeling of heat. Hence, our evaluation also contributes to the complex differential diagnosis of diseases in the anterior chest wall (38). Moreover, we would like to show that not only pathological changes of the costoclavicular ligament (57,58) but also certain so far not mentioned hyperostotic foci in the osseous parts of the ribs are of special significance in early diagnosis of AHS.

MATERIALS AND METHODS

Thirty-four patients with AHS including 17 women (mean age 49.4 years, ranging from 24 to 68 years) and 17 men (mean age 47.1 years, ranging from 21 to 81 years) were evaluated. Patients or their history including X-ray films were referred by rheumatologists, orthopaedic surgeons, specialists in internal diseases and radiologists. Dermatological examinations had already been carried out or were suggested by us. The following X-rays (in at least one plane) and/or conventional tomosgrams were evaluated: sternocostoclavicular region comprising follow-up at irregular intervals for up to 14 years in 31 cases. In another patient, only the bone scintigraphy with bone scanning 99m-Tc phosphate complexes provided information on this region. The sternoclavicular region had not been investigated radiologically in one patient with possible and one patient with incomplete AHS, since no symptoms had been specified there. Computer tomograms (CT) of the sternocostoclavicular region were available from five patients.

NOMENCLATURE

For the following reasons, we prefer the disease designation “acquired hyperostosis syndrome” (17,18) for the entity discussed here:

Hyperostosis is a quantitative concept which indicates an increase of bone tissue without considering its cause (62). The hyperostosis can emanate from the endostem of the cancellous and/or cortical bone as well as from the periosteum. Furthermore, it may reflect metaplastic and enthesopathic new bone formation.

The adjective “acquired” indicates that in contrast to other hyperostotic diseases, the syndrome discussed here is not con-