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Sternocostoclavicular Hyperostosis

Presentation and long-term follow-up of three cases

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ABSTRACT

Sternocostoclavicular hyperostosis is a rare disease characterized by recurrent pain and skeletal swelling in the upper part of the chest. The clinical manifestations are closely linked to pustulosis palmo-plantaris but the etiology is still obscure. We present three cases of sternocostoclavicular hyperostosis with a follow-up period of 9-22 years at our department.

INTRODUCTION

Sternocostoclavicular hyperostosis is a rare ailment which can be characterized by clinical and radiological manifestations in the sternum, clavicles, upper ribs and spine. The earliest literature about the ailment was published in 1970 by Kapert and Campbell (8). Several cases have been monitored and reported, especially from Japan by Ishibashi et al. (7), Sonozaki et al. (18), Kojima (11), Abe et al. (1) and from Sweden by Björksten et al. (3) and Hradil et al. (6).

The presenting and most striking clinical feature is recurrent throbbing pain accompanied by local tumescence, an increase in body temperature and a tenderness in the anterior upper part of the chest. Weather changes and recurrent infections appear to aggravate the clinical picture. The radiological changes range from local to diffuse ossification and hyperostosis and Sonozaki et al. (18) has proposed a radiological classification divided into three stages (Table 1).

Table 1. Stage classification of radiological findings according to Sonozaki et al. (18).

Stage 1	Localized. Ossification is mild and localized to the costoclavicular ligaments. Tomography or computed tomography may or may not show intraosseous new-bone formation.
Stage 2.	Generalized. Ossification is widely spread beyond the margins of the costoclavicular ligaments. The inferior margins of the first ribs appear irregular but no periosteal reaction is visible.
Stage 3.	Hyperostotic. Not only the inferior but also the superior margins of the clavicles show hyperostotic changes and the clavicles appear thickened and tumescent.

In the early stages scintigraphy reveals an appreciable radionuclide uptake. The ailment is not characteristically accompanied by any specific laboratory findings, but later on, there can be slight changes in the erythrocyte sedimentation rate (ESR), white blood count (WBC), C-reactive protein (CRP) and serum alkaline phosphatase (SALP). Tests for rheumatoid factor (RF) and both specific and unspecific immunological tests are usually negative. Histopathology and cytology examinations may reveal non-specific, inflammatory cellular changes compatible with osteomyelitis or Paget's disease of the bone.

CASE REPORTS

Case 1

A 59 year old woman complained of periodically increasing pressure pain in the right sternoclavicular joint in 1969. Initially her pain responded well to Indomethacin. Radiological examinations and laboratory tests, (ESR, WBC, SALP and RF) were normal. There was no increase in body temperature.

She had been treated for recurrent pustulosis palmo-plantaris (PPP) several times from the mid 60's onwards at the Department of Dermatology. An infectious focus was suspected as well as a relationship between the PPP and the sternoclavicular clinical manifestations. Radiological findings, such as a widening of the synchondrosis between the manubrium and the corpus sterni, ossification in front and behind the synchondrosis as well as periarticularly around the sternoclavicular joint and the first costosternal joint were recorded in 1974 four years after the earliest clinical manifestations. The manubrium also manifested stage 2 findings according to Sonozaki et al. (18).

In 1976 a histopathological analysis from the sternoclavicular joint was carried out and revealed an unspecific chronic osteomyelitic reaction with an ESR of 43 mm/h (the highest ever recorded). Bone marrow cytology was normal and there was no evidence of malignancy.

An oral examination in 1979 revealed severe paradontitis and a chronic sinusitis was diagnosed. Haemophilus bacteria were cultured from the epipharynx. She was treated successfully with penicillins, wash-outs and a radical sinus operation coupled with dental extraction was performed in 1981. Since then, she has had no complaints of either inflammatory symptoms in the anterior part of the chest or PPP.

Case 2

A 42 year-old woman presented with a pustulosis palmo-plantaris in 1965. She suffered from recurrent manifestations but all bacterial cultures proved negative. She was referred to our department in 1967 with a tender, warm and hard tumescence in the right sternoclavicular area which was accompanied by throbbing pain. She was afebrile and had an ESR of 12 mm/h. X-ray images demonstrated an expansive osteolytic lesion in the medial third of the right clavicle and a periosteal reaction localized mainly in the posterior aspect. Malignancy (sarcoma or Ewing's tumor) was suspected and a total right clavicleectomy was performed. The histopathological examination showed a picture of unspecific chronic osteomyelitis. There was no evidence of malignancy.

Since 1974 she has had episodes of back aches (cervical, thoracic and/or lumbar) but the first radiological manifestations were revealed in 1983 when unilateral sclerotic lesions in the vertebral bodies of L III, L V and S I were observed. They also demonstrated overbridging sclerotic osteophytes between the left aspects of L III and L IV and the right of L V and S I. There was no evidence of lumbar disc collapse. Her symptoms exacerbated during the night and morning hours but gradually improved towards the afternoon and evening hours. They also responded well to Indomethacin therapy.

In 1975, the same type of symptoms reappeared as in 1967 but this time in the left sternoclavicular area. X-ray images and bacterial cultures from the left sternoclavicular joint were negative. In 1987 a hard tumescence accompanied by a local increase in temperature was observed and X-ray images including tomography demonstrated ossification and hyperostosis, with irregular sclerotic lesions in the sternalmanubrium and osteolysis in the medial end of the left clavicle (stage 3 according to Sonozaki et al.) (18).

The symptoms could only be contained by continuous antibiotic (Cephalexin) administration during a period of five months. However the pain worsened and a partial medial clavicle resection was performed. The histopathological examination revealed findings compatible with chronic osteomyelitis but no organisms cloud be cultured from the specimen.

She has never been free from symptoms for more than 4 months each year. Her ESR has remained at a relatively high level (25-40 mm/h) since 1986.

She has recently presented with the same inflammatory symptoms again in the proximal right side of the manubrium, incapacitating pains in the left sacroiliac and hip joints, pustulas on her palms as well as decrease in her cervical- and lumbar- spine mobility.

Although multidisciplinary approaches have been implemented, no infectious focus has ever been revealed.

Case 3

In 1980, a 26-year-old woman presented with atypical dermatological manifestations on her right leg and left foot. Several diagnoses such like psoriasis, neurodermatitis, recurrent chronic dermatitis, were postulated but the dermatologists could not arrive at one definitive diagnosis.

In 1986 she was referred to our department because of pains and a tenderness in the left sternoclavicular joint. She had been out of work for more than one year. The pains worsened during the night and morning hours and varied with climatic changes. In 1987 a hard tumescence developed gradually in the sternoclavicular joint.

Scintigraphy revealed a high radionuclide uptake in the manubrium sterni and the sternoclavicular areas. Radiological examination including computerized tomography of the left sternoclavicular joint demonstrated hyperostotic lesions in the clavicle with cortical destruction (stage 3 according to Sonozaki et al.) (18). Histopathological studies of the bone from the area have shown findings compatible with chronic osteomyelitis but a culture from the specimen proved negative. There was no evidence of malignancy.

Table 2. Clinical and laboratory characteristics of the three cases of sternocosto-clavicular hyperostosis.

Case	Sex	Presenting age (years)	Follow-up (years)	Side	ESR	Skin manifestations
1	female	40	19	left	low	PPP
2	"	21	22	bilat	varying	PPP
3	"	26	9	left	high	atypical

DISCUSSION

Our three patients (Table 2) were all females and at the time of the debut of their symptoms aged 21, 26 and 40 years respectively. These facts are not in agreement with the observations of Resnik et al. (17), who stated that this disorder typically affects men or, less frequently, women in the fourth, fifth or sixth decades of life. Our observations are in agreement with those of authors from Japan, such as Chigira et al. (4), Kojima (11) and Sonozaki et al. (18). Our diagnosis of sternocostoclavicular hyperostosis is based on typical clinical symptoms, the histopathological picture of chronic osteomyelitis and the radiological and tomographic features described previously by Björksten et al. (3), Chigira et al. (4), Kapert and Campbell (8), Kato et al. (9), Kawai et al. (10) and Köhler et al. (12). Scintigraphy is an extremely useful tool especially in the early stages when radiological findings are not yet conclusive (16).

Laboratory tests have only been of value for the purpose of differential diagnosis. The ESR which usually varies in inflammatory diseases is relatively unpredictable. It is known that psoriasis can also be accompanied by joint complications, which tend to follow the same irregular chronic pattern characterized by remissions and exacerbations, having an unpredictable onset and duration. In our three cases we found that pustulosis palmo-plantaris (in cases 1 and 2) proceeded the sternocostoclavicular manifestations by 3–5 years, as observed by Enfors and Molin (5), Ishibashi et al. (7) and Kato et al. (9). Case 1 and 3 had atypical psoriasis-like lesions but the dermatologists could not arrive at a single diagnosis. The course and non-infectious genesis of the mentioned dermatological lesions are compatible with the well-established entity of "localized pustular psoriasis" (13). Our three patients could not recall any close relative with psoriasis.

Case 1 did not have any dermatological lesions or pains in the upper part of the chest since 1981 (at the age of 51 years) when the radical removal of a chronic infectious focus was effected. Scintigraphic (Tc 99m) imaging 8 years postoperatively still revealed a striking radionuclide uptake in the sternum and the sternocostoclavicular joints as evidence of a low continuous level of activity. This supports Ono (15) who postulated the possibility of a natural spontaneous remission of the clinical symptoms in elderly patients especially after the sixth decade. A distant infectious focus can function like a trigger mechanism or behave like an exacerbating factor of clinical manifestations in the sternocostoclavicular area (2). This

may also explain why random tonsillectomy as proposed by Ono (14) can relieve clinical symptoms in certain cases. We have not yet localized any infectious foci in case 2 and 3 but the symptoms sometimes appear to come under control during long-term antibiotic therapy (Cephalexin). Treatment with cyclooxygenase inhibitors has proved to be very effective during the peak symptomatic period (4,19).

In summary, sternocostoclavicular hyperostosis is a rare, recurrent disease which is easily confused with other chronic diseases, such as osteomyelitis, Paget's disease, ankylosing spondylitis etc. The diagnosis is based on typical clinical manifestations, radiological findings and negative bacterial cultures. In the acute stages, the patients respond to treatment with cyclooxygenase inhibitors. Systematic broad spectrum antibiotic therapy is sometimes effective in relieving the pain. Clavicle resection does not treat the symptoms radically but offers prolonged pain relief. Our three cases seem to demonstrate a variety of joint and bone complications as sequels to dermatological disease.

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