Sternocostoclavicular Hyperostosis

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Sternocostoclavicular hyperostosis (SCCH) is a very uncommon disease characterized by hyperostosis of the clavicle, upper ribs, sternum and soft tissue ossification, particularly of the costoclavicular ligament. Although the cause of this disorder is unknown, clinical, radiological and histological examinations may allow the diagnosis of SCCH. This paper presents a case of SCCH and a brief review of the literature.

Key Words: Bones, sclerosis, clavicle, hyperostosis

Sternocostoclavicular hyperostosis is an uncommon syndrome and since first reported in 1974, additional cases have been reported in Japan (Sonozaki et al. 1979). Other conditions presenting sclerotic changes of the clavicle have been reported (Franquet et al. 1985; Jurik et al. 1985; Jurik et al. 1986). To our knowledge, there have been no reports of sternocostoclavicular hyperostosis in Korean literature. It is necessary to recognize and identify the clinical and radiological manifestations for differentiation of SCCH from other ossific diathesis conditions to permit appropriate management.

CASE REPORT

A 64 year old man was admitted to the hospital with a complaint of painful swelling in the right clavicle. He had been well until 5 years earlier, when he experienced a drilling pain in his right upper chest. Thereafter, he felt intermittent pain in his right upper chest, which was aggravated by his work activity (writing). However, he performed ordinary daily activities without any specific treatment. Forty days before admission, the pain in the right upper chest increased and the swelling in the right clavicular area was noticed. Upon physical examination, a protruding, tender and hard mass was observed over the medial aspect of the right clavicle. There was moderate restriction of motion of the neck. There were no signs of venous congestion of the right upper extremity, and no lesions were found on the skin of both hands and feet.

Upon laboratory examination, the hematocrit was 36.3 percent and the white-cell count was 8500, with 61 percent neutrophils, 32 percent lymphocytes, and 7 percent monocytes. The erythrocyte sedimentation rate (ESR) was 28 mm per hour. The prothrombin time was 12.0 seconds, with 100 percent of normal; the partial thromboplastin time was 29.6 seconds, with a control ranging from 29 to 42 seconds. The glucose level was 100 mg per 100 ml and the blood urea nitrogen was 12 mg per 100 ml. The sodium level was 135 m mol, the potassium 4.1 m mol, the chloride 105 m mol, and the carbon dioxide 26 m mol per liter. The serum aspartate aminotransferase (SGOT) was 20 IU, and the alkaline phosphatase was 120 IU per liter. The serologic test for syphilis was negative.

Plain films and subsequent tomography showed expansion and diffuse sclerotic density of the medial two-thirds of the right clavicle, and almost complete fusion of the medial one-third of the right clavicle and the right first rib. The right sternoclavicular joint space was obliterated, likely due to ankylosis. Early ossification of the costochondral junction in both first ribs bilaterally and slight sclerosis of the manubriosternal junction was observed (Fig. 1).

A bone scan with 99m-Tc-MDP showed hot take-up on both clavicles, the sternum and multiple regions of the costal cartilages (Fig. 2).

A partial excision of the cortical bone and curettage of the abnormal medullary sponge bone were performed at the medial one-third of the right clavicle.
Fig. 1. a) Radiograph of the clavicles showing hyperostosis and ossification of the costo-clavicular ligament.
   b) Tomogram through the manubrium. Almost complete obliteration and sclerosis of the right sternoclavicular joint, and amorphous sclerosis in the manubriosternal junction.
Fig. 2. Anterior scan of the chest. Multiple areas of hot uptake on both clavicles, sternum, and costal cartilages.

Fig. 3. Sections obtained from a biopsy of the right clavicle (H & E stain, X200) show newly formed woven bone resorbed by osteoclasts and bordered by periosteal connective tissue fiber.
Sternocostoclavicular Hyperostosis

cle. There was no pus discharged. The results from the bacteriological studies were negative.

The histologic examination of the biopsy specimen from the right clavicle revealed newly formed woven bone, resorbed by osteoclasts and bordered by periosteal connective tissue, with infiltration of lymphocytes and plasma cells between collagen bundles (Fig. 3).

**DISCUSSION**

Sternocostoclavicular hyperostosis is thought to be a multiple skeletal disorder. However, the pathogenesis of this condition is still unclear although many attempts have been made to find infective organisms (Gerster et al. 1985).

SCCH typically affects men more often than women ranging in age from 30-50 years. Painful swelling in the sternoclavicular area is the most common presenting symptom, aggravated by cold, dampness, and recurrent infections. Venous congestion of the upper extremities may be associated with the occlusion of the subclavian vein due to either mechanical compression by the thickened bone of the clavicles or thrombotic obstruction (Jirik et al. 1982; Koehler et al. 1977). Pustulosis palmans and plantaris can be evident in approximately 60% of the cases of SCCH (Sonozaki et al. 1981; Gerster et al. 1985). In our case, there was no evidence of edema of the upper extremities or of pustulous lesions in the palm and sole.

Laboratory tests generally are unremarkable, except for a mild elevation of ESR, an increase in the α- and β-globulins, the persence of c-reactive protein and a slight increase in alkaline phosphatase (Gerster et al. 1985).

Pathological examinations reveal bony resorption and subperiosteal bone remodelling about the clavicle, sternum and upper rib, although bacteriological studies are generally negative (Resnick et al. 1981).

Radiographic features of SCCH are hyperostosis of the clavicle, synostosis of the sternoclavicular joints, ossification of the costo-clavicular ligaments, hyperostosis of the first ribs, and occasionally enlargement of the sternum and upper ribs. In our case, a tomogram clearly revealed the alterations in the sternoclavicular joints and the sternum which was overlapped by the spine in conventional radiograph. According to the classification by Sonozaki et al., our patient had stage 3 disease.

The radiographic appearance must be characteristic for the diagnosis of the sternocostoclavicular hyperostosis. Although the radiographic features of Paget's disease are similar to SCCH, it does not commonly involve only the clavicles or the sternum and the sternoclavicular joint. The laboratory tests commonly show an elevation of serum alkaline phosphatase. Osteoelastic and Friedreich's disease show increased bone density affecting the medial end of the clavicle (Franquet et al. 1985). However, they do not involve the sterno-clavicular joint and a unilateral involvement of the clavicle is common in the former (Franquet et al. 1985; Brower et al. 1974). The chronic recurrent multifocal osteomyelitis (CRMO) affecting the clavicles may be difficult to differentiate from SCCH in radiographic features. However, CRMO is common in children and adolescents and affects other sites of the skeleton (Jirik et al. 1986).

Associated conditions which include ankylosing spondylitis and diffuse idiopathic skeletal hyperostosis (DISH) have been reported (Sonozaki et al. 1981). As compared to these conditions of ossifying diathesis, serologic tests for the histocompatibility antigen B-27 have been reported to be negative in SCCH. The involvement of other parts of the skeleton, in addition to the upper portion of the chest, has been reported (Gerster et al. 1985).

**REFERENCES**


