Treatment-Refractory Sternocostoclavicular Hyperostosis

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Sternocostoclavicular hyperostosis (SCCH) is an infrequent chronic inflammatory disorder of the axial skeleton of unknown origin. SCCH goes often unrecognized due to a low level of awareness for the disorder. It typically presents with relapsing and remitting pain in the shoulder, neck, and anterior chest wall area with occasional swelling and tenderness of the sternoclavicular area. The diagnosis is confirmed radiologically by sclerosis and hyperostosis of the sternoclavicular joints. There have been several reports in which intravenous bisphosphonates and tumor necrosis factor-inhibitors have shown reasonable efficacy in the treatment of this disorder. We report a patient with a long history of SCCH in whom pamidronate 60 mg intravenously every 3 months for 3 years failed to reduce symptom severity and improve radiologic findings.

Keywords: Bisphosphonates; Pamidronate; Sternocostoclavicular hyperostosis; Tumor necrosis factor-inhibitors

Sternocostoclavicular hyperostosis (SCCH) is a rare chronic inflammatory condition of the axial skeleton that was first described in 1968.¹ SCCH typically presents with intermittent attacks of severe pain, redness, and swelling in the sternoclavicular region often associated with considerable impairment of shoulder girdle movement.² The exact incidence of SCCH is unknown, as many cases go undiagnosed.³ Over the last two decades, there have been several reports in which intravenous bisphosphonates and tumor necrosis factor (TNF)-inhibitors have shown reasonable efficacy in the treatment of this disorder.⁴⁻⁹ Here, we report the clinical, laboratory, and radiologic data of a patient with treatment-refractory SCCH.

Case Report

A Caucasian woman, aged 28 years, was seen in our clinic because of a long history of upper shoulder girdle and anterior chest wall pain dating to age 18. The patient recalls developing intermittent episodes of painful swelling of the sternum, clavicles, and upper ribs that became more persistent with each episode. The pain and swelling did not decrease with physical therapy or multiple analgesics including corticosteroids and ibuprofen.

She carried a medical diagnosis of type I diabetes mellitus, depression and affective disorders, ulcerative colitis with

backwash ileitis, dyslipidemia, hypertension, seizures, and right hemicolectomy for moderately differentiated adenocarcinoma of the ascending colon which was diagnosed at age 28. Medications included alprazolam, duloxetine, mesalamine, insulin, losartan, pravastatin, lamotrigine, and ibuprofen. She had no known allergies. She had a 20 pack per year smoking history until 5 years ago, and she drinks alcohol occasionally. There was no family history of rheumatic diseases. On examination, she was exquisitely tender along the clavicle bilaterally and at the manubriosternal joint and proximal sternum. The appendicular skeleton was without synovitis or effusion. No skin lesions were found.

A chest radiograph showed sclerosis and exuberant enlargement of involved bone (figure 1A). A computed tomography (CT) scan revealed extensive mature ossification of the sternoclavicular joints and first costochondral junctions, extending into the soft tissues, consistent with SCCH (figures 1B and 2A). Axial imaging demonstrated additional findings of diffuse idiopathic skeletal hyperostosis throughout the thoracic and lumbar spine. No sacroiliitis was seen on magnetic resonance imaging (MRI).

The patient had a microcytic anemia with a hemoglobin level of 11.3 g/dL. Erythrocyte sedimentation rate was 31 mm/hr, and C-reactive protein was 3.6 mg/dL. Tests of renal, liver,

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Figure 1. Chest radiograph (A) and computed tomography (CT) scan with 3D reformatted image (B) show marked hyperostosis of bilateral sternocostoclavicular joints (arrows). The ossification extends into the soft tissue with a large bony bridge between the left 1st and 2nd ribs.

Figure 2. Computed tomography (CT) scan of the chest with coronal reformatted images reveals slight progressive mature ossification of the bilateral sternoclavicular joints (arrows) pre- (A) and post- (B) treatment.

thyroid, and parathyroid function were normal, as were blood levels for vitamin D, calcium, phosphorus, retinol, fluoride, creatine kinase, and hepatitis C. Bone turnover markers consisting of serum total alkaline phosphatase and urinary collagen type 1 cross-linked N-telopeptide were normal. Rheumatoid factor and HLA B27 were negative. A dual-energy X-ray absorptiometry scan was normal.

Initial brief (6-month) treatment with infliximab, a TNF-inhibitor agent, was unsuccessful in improving clinical symptoms. Intravenous pamidronate was then administered at a dose of 60 mg given as a 2- to 3-hour infusion every 3 months. There were no relevant adverse events associated with pamidronate treatment. After a possible initial improvement in pain and stiffness following each injection, no clinical benefit ensued by the end of her series of 13 pamidronate injections. Her anemia and raised inflammatory markers persisted after treatment. A repeat chest CT scan obtained at the end of therapy failed to demonstrate radiologic improvement in mature ossification of the bilateral sternoclavicular joints (figure 2B).

Discussion

SCCH is a distinct clinical entity that causes progressive hyperostosis of the sternocostoclavicular joints and eventual soft tissue ossification.^{1,2} Some authors believe SCCH belongs to the spectrum of SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis, and osteitis). SCCH is largely underdiagnosed due to a low level of awareness for the disorder, and therefore it may be more common than currently believed.³ SCCH is a disorder of midlife, with a slight female predilection.¹⁰ The condition is bilateral in most patients. The spine, sacroiliac joints, and peripheral joints may also be affected.³ No specific diagnostic test for SCCH is available; the clinical diagnosis is based on pattern recognition and exclusion. It is important to differentiate this entity from the seronegative spondyloarthropathies, infectious osteomyelitis, bone metastasis, and condensing osteitis of the clavicle.¹¹ We found a unique association with diffuse idiopathic skeletal hyperostosis in our case.

The exact pathophysiology of SCCH remains unclear. A nonspecific chronic sterile inflammation is typically seen on histology. The inflammatory osteitis and localized increased

bone turnover result in thickening of trabecular bone and formation of osteoid tissue, eventually giving the characteristic hyperostotic bone changes in the sternoclavicular area.¹² The common clinical picture is one of relapsing and remitting pain in the shoulder, neck, and anterior chest wall area with occasional swelling and tenderness of the sternoclavicular area.³ Computed tomography (CT) has the ability to detect sclerotic bone trabeculae not obvious on plain radiographs.¹³ Whole-body bone scintigraphy typically demonstrates increased radiotracer activity in the sternal manubrium and in the adjacent clavicles and ribs; a distinctive finding known as the “bullhead” sign.¹³ Results of clinical history, laboratory, and radiologic studies all support the diagnosis of SCCH in the present case.

Treatment first involves non-steroidal anti-inflammatory drugs and glucocorticoids, which are sometimes effective in alleviating pain and local inflammation, but do not prevent radiographic progression and the risk of secondary degenerative changes.¹¹ In advanced stages of the disease, surgical resection of the ossifications may be needed.¹¹ Preliminary experience supports the use of TNF-inhibitors and intravenous bisphosphonates in SCCH.⁴⁻⁹ Ringe and colleagues⁵ administered 2 mg of intravenous ibandronate every 3 months for up to a year to three patients with long-standing refractory SCCH and evaluated clinical, laboratory, and imaging at baseline and at 1 year. There was a prompt persistent pain relief, a gradual decline in inflammatory markers, and improvement in radiographic and scintigraphic findings. We used pamidronate 60 mg intravenously every 3 months for 3 years. Although the treatment was well tolerated, lasting reduction in symptom severity and radiologic improvement were not observed. Further clinical studies are therefore needed to determine the future role of bisphosphonates in SCCH.

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