Sternocostoclavicular Hyperostosis: An Insufficiently Recognized Clinical Entity

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ABSTRACT: A 79-year-old male chronic hemodialysis patient with no history of central venous catheterization was referred to our hospital with progressive swelling of the left upper limb ipsilateral to a forearm arteriovenous fistula. Radiological assessments revealed marked hyperostosis in the ribs, sternum, and clavicles with well-developed ossification of the sternocostoclavicular ligaments. Such characteristic structural abnormalities and our failure to identify the left subclavian vein with contrast material despite the abundant dilated collaterals in the left shoulder area encouraged us to diagnose our patient with sternocostoclavicular hyperostosis (SCCH) complicated by central vein obstruction. The structural impact of the sternocostoclavicular region as a potential risk for inducing central vein obstruction and the diagnostic concerns of SCCH in this patient are also discussed.

KEYWORDS: sternocostoclavicular hyperostosis, central vein obstruction, hemodialysis, vascular access, SAPHO syndrome

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Introduction

Sternocostoclavicular hyperostosis (SCCH) is a chronic ossifying diathesis affecting mostly juxtasternal structures.[1] It may be accompanied by painful swelling of the upper anterior chest wall with or without erythematous appearance, whereas some subsets of patients may manifest limited shoulder mobility as the condition progresses.[2,3] Despite a steady but modest accumulation of reports on SCCH, the low level of awareness regarding this clinical entity may often lead to a delay in its diagnosis in ordinary clinical practice.[3] In this report, we describe one such case in a male chronic hemodialysis (HD) patient with progressive swelling of the arm ipsilateral to a vascular access site for HD. During the diagnostic investigation of the disease, external compression of the left subclavian vein resulting from concurrent SCCH was fortuitously discovered.

Case Report

A 79-year-old male chronic HD patient with no history of central venous catheterization was referred to our hospital with progressive swelling of the left arm ipsilateral to a forearm radiocephalic arteriovenous fistula (AVF). He had been undergoing HD treatment for 3 years because of chronic renal failure due to hypertensive nephrosclerosis, and the AVF had been surgically created as a permanent vascular access just before initiating periodic HD. A conventional chest radiograph obtained at that time showed marked hyperostosis of the left clavicle (Figure 1A); however, this distinct finding was overlooked and had never been thoroughly investigated.

His medications included benidipine hydrochloride 8 mg/day and furosemide 40 mg/day. Regarding his other medical history, he underwent treatment for hyperuricemia and hyperphosphatemia, which had been treated with allopurinol 100 mg/day, calcium carbonate 1000 mg/day, and lanthanum carbonate 750 mg/day, thereby leading to serum levels of uric acid and phosphorus (Pi) levels ranging from 4.0 to 5.5 mg/dL (reference range: 3.8-6.6 mg/dL) and 3.5 to 5.2 mg/dL (reference range: 2.4-4.6 mg/dL), respectively. He had been administered oral alfacalcidol 0.25 µg/day as well for concurrent secondary hyperparathyroidism, and thus, his serum levels of intact parathyroid hormone and alkaline phosphatase had been favorably maintained at approximately 200 pg/mL (reference range: 10-65 pg/mL) and 200 U/L (reference range: 107-330 U/L). He never had any AVF complications requiring radiological intervention, and there was no history of trauma, erythema, fever, or skin lesions. During the 6 months prior to this referral, the symptoms in his arm had slowly but gradually worsened. The findings on a systemic evaluation were unremarkable, and he did not have any pain in the back, buttocks, or mandible. However, a careful medical interview revealed that he had occasionally experienced painful swelling only in the left sternocostoclavicular region over the past 5 years, although he had not sought any medical advice due to the temporary nature of the aching. No lymphoadenopathy was noted in the cervical, axillary, or inguinal region, and a palmoplantar examination did not reveal any cutaneous abnormalities. The findings from a laboratory evaluation were as

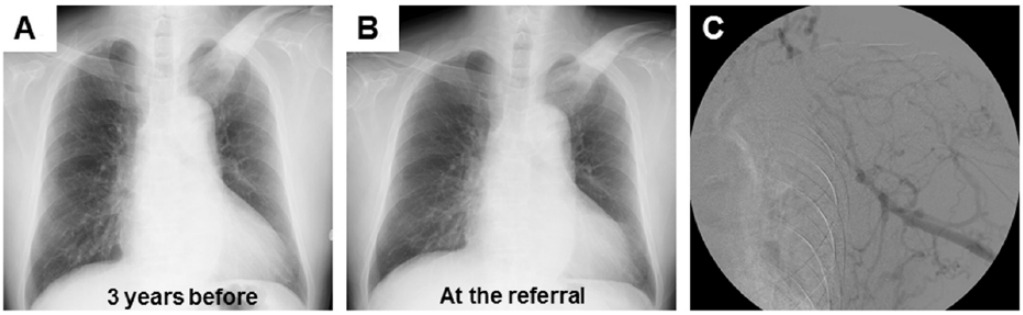

Figure 1. Conventional plain radiographs and left upper limb digital subtraction angiography. (A) A chest radiograph obtained 3 years earlier had already shown the widespread increased radiodensity in the bilateral sternocostoclavicular regions as well as marked left clavicular hyperostosis, neither of which had been investigated. (B) Similar skeletal findings were confirmed at the referral as well. (C) Digital subtraction venogram injected via the drainage vein of the left forearm arteriovenous fistula failed to demonstrate the left subclavian vein despite the abundant dilated collaterals in the left shoulder area.

follows: blood urea nitrogen, 39 mg/dL (reference range: 8-20 mg/dL); serum creatinine, 8.33 mg/dL (reference range: 0.63-1.03 mg/dL); white blood cell count, 6500/μL (reference range: 3900-9800/μL); hemoglobin, 8.6 g/dL (reference range: 13.5-17.6 g/dL); platelet count, 275×10^3/μL (reference range: $130\text{-}369 \times 10^3$/μL); serum albumin, 4.0 g/dL (reference range: 6.9-8.4 g/dL); fibrin/fibrinogen degradation products, 7.7 μg/mL (reference range: 0-5 μg/mL); D-dimer, 3300 μg/L (reference range: 0-1,500 μg/L); C3, 92 mg/dL (reference range: 86-160 mg/dL); C4, 31 mg/dL (reference range: 17-45 mg/dL); and C-reactive protein, 0.21 mg/dL (reference range: 0-0.14 mg/dL).

A chest radiograph showed pronounced hyperostosis of the left clavicle (Figure 1B), whereas digital subtraction angiography (DSA) of the left upper limb revealed failed circulation in the ipsilateral subclavian vein with abundant collaterals (Figure 1C). Diagnostic thoracic computed tomography (CT) revealed marked ossification of the sternocostoclavicular joints with distinct bone overgrowth in the sternum and upper ribs as well as the left clavicle (Figure 2). Despite the injection of a contrast material, the left subclavian vein could not be observed, but we did notice collateralization in the surrounding thoracic region (Figure 2). Based on the results of physical and laboratory assessments as well as the characteristic image findings without suggestive signs of infection or neoplasm, he was diagnosed with SCCH complicated by central vein obstruction.

Discussion

Atraumatic abnormalities in the sternocostoclavicular territory showing swelling, redness, and/or pain in the corresponding regions are occasionally seen in ordinary clinical practice.[2,4,5] A wide variety of underlying causes, including infection, neoplasm, osteitis condensans, osteoarthritis, and chronic osteomyelitis, as well as SCCH, have been described; however, the mild nature of these clinical manifestations and thus little focus given to them by both patients and physicians still lead to the underdiagnosis of these conditions.[2,4-6] In this regard, the lack of reports describing the complications of SCCH even in patients who receive periodic HD is not surprising. To our knowledge, this is the first report of a chronic HD patient concurrently complicated by central vein obstruction and SCCH.

Sternocostoclavicular hyperostosis is a rare disorder that starts with nonspecific inflammation of the sternocostoclavicular ligament and the surrounding soft tissue.[1,2,3,7] The osteoarticular manifestations may have a very slow but variable clinical course characterized by periods of exacerbation and remission, whereas the untreated disease is often associated with marked long-duration morbidity because of a progressive structural abnormality characterized by bone overgrowth as well as secondary degenerative changes with soft-tissue ossification of the sternum, medial clavicle, and upper ribs, either unilaterally or bilaterally.[1-3,5-7] Most of the affected patients were between their 30s and 50s, but a wide range of ages—from 11 years to the late 80s—have been affected, with a slight predominance of female sex.[1,3]

Since it was first described as a separate entity by Sonozaki et al[8] in 1974 and later by Köhler et al[9] in 1975, the cumulative number of publications on SCCH has been slowly growing, and at present, about 120 articles have been published in the international literature.[3] The incidence of the disease remains to be clarified, partly because the current data are largely limited to anecdotal case reports and case series,[6,7] and no cause has been determined despite pathological analyses revealing signs of widespread bone remodeling with fibrosis, resembling chronic osteomyelitis and Paget bone disease.[1,2] It has been reported that approximately 50% of patients with SCCH may have cutaneous complications, such as palmoplantar pustulosis, pustular psoriasis, and psoriasis vulgaris.[2,3,6] At present, SCCH has numerous synonyms, such as sternocostoclavicular syndrome, SAPHO (synovitis, acne, pustulosis, hyperostosis, and osteitis) syndrome, acquired hyperostosis syndrome, and pustular arthroosteitis.[1-3,6,7,10] This varying terminology mirrors the current lack of understanding of this clinical entity, which hinders the formulation of uniform guidelines regarding the classification of SCCH.[6,10] Complementary studies with radiographs and CT are expected to aid in the diagnostic work-up, despite the lack of specific laboratory findings.[1-3,6,7,10]

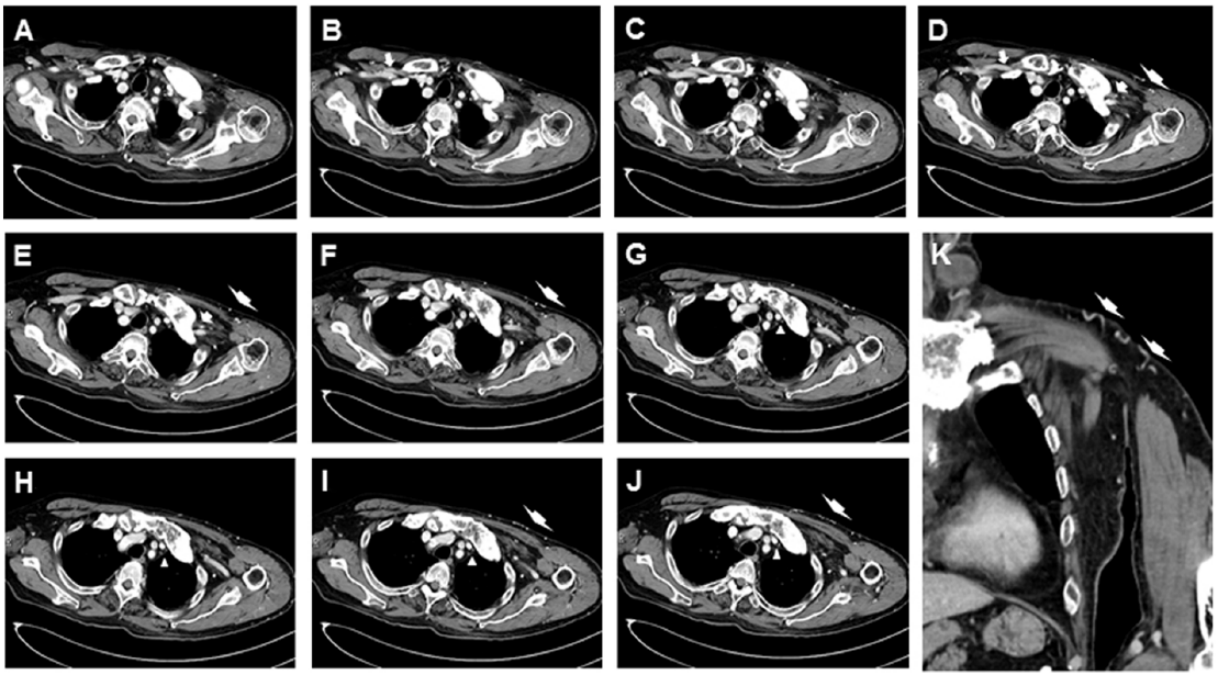

Figure 2. Thoracic computed tomography (CT) with contrast injection. A series of CT scans shown in alphabetical order (A to J) reveals marked hyperostosis in the ribs, sternum, and clavicles with well-developed ossification of the sternocostoclavicular ligaments. Note that the left clavicle and manubrioclavicular joint are predominantly affected. Despite the successful identification of the right subclavian vein (narrow arrows in B to D) and left innominate vein (arrowheads in G to J) with the contrast material, the left subclavian vein could not be visualized sufficiently in the corresponding area (middle arrows in D and E), although we did notice collateralization in the surrounding thoracic region (wide arrows), which was also identified in a coronal reconstructed image (K).

Technetium bone scintigraphy is another imaging modality of choice for identifying SCCH[1-3,6,7,10]; therefore, diagnosing the disease without performing this procedure, as in the present patient, may be challenging. However, such a diagnostic manner is not exceptional.[11] We feel that the radiographic images obtained in the current patient were quite pathognomonic and diagnostic for SCCH, preventing us from ascribing the sternocostoclavicular manifestations to other infections, neoplasms, osteitis condensans, or osteoarthritis.[2,4-6] Chronic osteomyelitis and Paget bone disease may be somewhat difficult to differentiate from SCCH based on the radiographic features alone.[12] However, chronic osteomyelitis occurs exclusively in children and adolescents and affects other sites of the skeleton,[6,12] and Paget bone disease most commonly involves the axial skeleton and is characterized by elevated levels of serum alkaline phosphatase,[12,13] which was absent in our patient. Finally, we believe it is reasonable to consider that the previous episodes of painful left sternocostoclavicular swelling were probably related to regional inflammatory events mediated by the disease. Needless to say, it is important to note that primary neoplasms of the sternocostoclavicular region, although rare, are much more frequently malignant than benign.[4] Therefore, careful assessments and appropriate vigilance are important, and a biopsy may be required if rapid and further enlargement of the swelling occurs.[4,5]

Central occlusive venous disease is a serious complication in chronic HD patients as it can disrupt the vascular access circuit through venous hypertension and flow reduction, with or without overt arm swelling on the same side of the arteriovenous vascular access site.[14,15] Mechanical damage due to dialysis catheter insertion and continuous catheter movement inside the vasculature, which induces endothelial injuries, neointimal hyperplasia, fibrosis, and thrombophlebitic reaction, has received attention as a potential cause of such pathology in HD patients.[14,16,17] The vascular access–related changes in the flow dynamics in the central veins, which are associated with abnormal shear stress, turbulence, and platelet aggregation, thereby leading to neointimal hyperplasia, may also act as an alternative pathogenic basis for central occlusive venous disease, regardless of the patient's history of central venous catheterization.[14-19]

The details regarding how subclavian vein obstruction occurred in the present patient remain unclear; however, the presence of prominent collateral veins in the surrounding region, which are seen most often with chronic thrombosis,[20] supports the notion that the chronic and extrinsic compression of the left subclavian vein at the level of the costoclavicular space by hyperostotic bone structures, combined with an elevated arterialized venous flow resulting from the AVF placement, may have played a role in accelerating the disease progression. Such a scenario may not be surprising as the link between SCCH and venous thoracic outlet syndrome, which has already been acknowledged anecdotally,[1,2,21–23] may act as the groundwork for the disease. Nevertheless, our experience encourages us to emphasize the structural impact of the sternocostoclavicular region as a potential risk for inducing central

vein obstruction in chronic HD patients, which still seems to be a challenge in the field of nephrology.[16] Indeed, the hyperostotic lesion was completely overlooked in the current patient when the left forearm AVF was created, despite its demonstration on a perioperative chest radiograph.

Author Contributions

TS and TA drafted the manuscript. HK and AK helped with the acquisition of clinical data. DN provided a detailed review of the contents and structure of the manuscript, resulting in significant changes to the original document. All of the authors have read and approved the final manuscript.

Disclosures and Ethics

As a requirement for publication, the authors have provided the publisher with signed confirmation of their compliance with legal and ethical obligations including, but not limited to, the following: authorship and contributorship, conflicts of interest, privacy and confidentiality, and (where applicable) the protection of human and animal research subjects. The authors have read and confirmed their agreement with the ICMJE authorship and conflict of interest criteria. The authors have also confirmed that this manuscript is unique and not under consideration for publication or published in any other journals and that they have permission from the rights holders to reproduce any copyrighted material. Any disclosures are made in this section. The external blind peer reviewers report no conflicts of interest.

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