Multiple venous thrombosis in SAPHO syndrome

T Kawabata, Y Morita, A Nakatsuka, H Kagawa, M Kawashima, T Sei, M Yamamura, H Makino

A 42 year old Japanese woman presented to our clinic complaining of severe pain in her neck, both shoulders, and lumbar spine. She had had pustulosis palmaris and plantaris from age 16 and recurrent painful swelling of the clavicles, the sternoclavicular joints, and the sternum from age 25. She reported multiple visits to the doctor for similar symptoms, and she was treated with non-steroidal anti-inflammatory drugs.

On initial examination at our clinic, her lumbar and cervical spine mobility was limited considerably. She had an oedematous face and subcutaneous collateral circulation on chest and abdomen. Laboratory testing showed a white cell count of $13.9 \times 10^9/\text{l}$ and a C reactive protein of 142 mg/l (normal <5). Antinuclear antibodies, antineutrophil cytoplasmic antibodies, anticardiolipin antibodies, lupus anticoagulants, and rheumatoid factors were all negative. Prothrombin time and activated partial thromboplastin time were normal. Locus B HLA typing was positive for B48 and B52. She reported that her C reactive protein level was always over 100 mg/l.

Plain radiographs disclosed marked hyperostosis in the medial aspect of both clavicles. There was narrowing and ankylosis in the apophysial joints of the cervical spine, and complete ankylosis in both sacroiliac joints. Contrast enhanced computed tomographic scans demonstrated a soft tissue mass in the upper mediastinum (fig 1A). Both subclavian veins were severely stenotic, and the superior vena cava (SVC) was almost completely occluded (fig 1B). Thrombosis was seen within the left internal jugular vein, and the right internal jugular vein could not be observed (fig 1C). Injection of contrast medium showed a network of collateral veins in the neck, chest, and abdominal wall. A diagnosis of SAPHO syndrome complicated by multiple venous thrombosis and SVC syndrome was made. Prednisolone was given at 10 mg/day, and the pain in her shoulders, neck, and lumbar spine improved. The treatment also included warfarin for anticoagulation.

DISCUSSION

Venous thrombosis complicating SAPHO syndrome is uncommon. In a series of 120 patients with this syndrome, Hayem et al found that only one (0.8%) patient had thrombosis of the subclavian vein. We found eight well documented cases of patients with SAPHO syndrome who developed venous thrombosis. Six of the patients had subclavian vein thrombosis, and only one case had SVC...
obstruction. In one of the eight patients, thrombosis of the iliac vein was found in a patient with lumbar vertebral osteitis and a large tissue mass surrounding the vein. The pathological mechanisms of venous thrombosis are still not clearly understood, but it is suggested that it is caused by vein compression by hyperostosis, or sheathing of the veins by an inflammatory and fibrotic tissue mass.

We have observed a new case of SVC obstruction in SAPHO syndrome, which is of particular interest, as multiple thrombi were observed, not only in the SVC and both subclavian veins but also in the internal jugular vein, which is not directly adjacent to the affected bones and the mediastinal mass. In our case the congestive venous flow may reflect the thrombosis in the internal jugular vein. Van Holsbeeck et al reported a case of a patient with SAPHO syndrome with bilateral subclavian vein obstruction who had a high level of lupus anticoagulant. Although our patient does not have any antiphospholipid antibodies, such factors may be associated with venous thrombosis in this syndrome.

SAPHO syndrome has some features of the spondyloarthropathies. Whether this syndrome is a unique subset of the spondyloarthropathies or is a distinct clinical entity is not well established. Nevertheless, the concept of SAPHO syndrome is useful in defining a subgroup of patients with unique clinical features. Venous thrombosis may lead to serious complications in patients with this syndrome. Our patient further emphasises the importance of recognising this complication in SAPHO syndrome.

A potential pitfall in the use of the Disease Activity Score (DAS28) as the main response criterion in treatment guidelines for patients with rheumatoid arthritis

P V Gardiner, A L Bell, A J Taggart, G Wright, F Kee, A Smyth, R McKane, J Lee, M E Rooney, E Whitehead

The Disease Activity Score\(^1\) is widely used to quantify disease activity and gauge response to treatment. A rather complex calculation conceals the relative contribution of each measure to the composite score. The 28 joint version (DAS28) is used in the British Society for Rheumatology guidelines to determine response to anti-tumour necrosis factor \(\alpha\) (anti-TNF\(\alpha\)) treatment.\(^2\) A reduction in DAS28 of \(\geq 1.2\) is considered significant improvement, as is a reduction in DAS28 score to \(< 3.2\). These figures are important, as under current guidelines clinicians are advised to discontinue anti-TNF treatment if either of these criteria is not achieved at 3 months.

Figure 1  Contrast enhanced computed tomographic scans (A) demonstrating a soft tissue mass in the upper mediastinum (*); (B) showing almost complete occlusion of the SVC (arrow); and (C) thrombosis within the left internal jugular vein (arrow). In (C) The right internal jugular vein cannot be seen.

Y Morita, Division of Nephrology and Rheumatology, Department of Internal Medicine, Kawasaki Medical School, Japan

T Sei, Department of Radiology, Okayama University Graduate School of Medicine and Dentistry, Japan

Correspondence to: Dr Y Morita, Division of Nephrology and Rheumatology, Department of Internal Medicine, Kawasaki Medical School, 577 Matsuhashi, Kurashiki 710–0192, Japan; morita@med.kawasaki-m.ac.jp

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