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CASE REPORT

Renal dysfunction due to hydronephrosis by SAPHO syndrome: a case report

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Key Clinical Message

Synovitis, Acne, Pustulosis, Hyperostosis, Osteitis (SAPHO) syndrome shows varied pathological symptoms. This is the first report of hydronephrosis due to the mechanical compression of bilateral ureters as a result of SAPHO syndrome. From our experience, MRI is the most useful imaging examination to check the upper urinary tract in SAPHO syndrome.

Keywords

hydronephrosis, magnetic resonance imaging, renal dysfunction, SAPHO syndrome.

Introduction

The acronym "SAPHO" denotes synovitis-acne-pustulosis-hyperostosis-osteitis, which is characterized by the occurrence of dermatoses and the formation of sterile abscesses, accompanied by sterile osteitis that resembles osteomyelitis as well as articular lesions that cause synostosis. Although this syndrome shows varied pathological signs and symptoms, the diagnosis of SAPHO syndrome was difficult due to its non-specific and wide-spectrum manifestations. Hydronephrosis is a common pathological condition for Urologists. In general, upper tract urothelial cancer, urinary stone, and malignancies in the pelvis are major and important causes of hydronephrosis. However, urologists scarcely ever consider SAPHO syndrome as the cause of hydronephrosis. In fact, only a few reports have discussed the association of this syndrome with disorders of the urinary organs [1–3]. This is the first report of a patient with hydronephrosis due to inflammation caused by SAPHO syndrome. We emphasize that discussion and awareness for the usefulness of magnetic resonance imaging (MRI) in the diagnosis are important for the Urologist in this case report.

Case Report

A 60-year-old man was admitted to Nagasaki University Hospital to examine the cause of bilateral hydronephrosis. His chief complaint was contemporary left backache. Abdominal computed tomography (CT) and ultrasonography showed bilateral dilatations from the upper to the pelvic ureters and no definitive location of the obstruction. He had been diagnosed with SAPHO syndrome in 2002 due to symptoms caused by palmoplantar pustulosis and sacroiliac arthritis. From the time of diagnosis, he has been treated with oral azulfidine and predonine. His regular physician judged his current pathological condition as being abate.

A blood test showed a slightly increased white blood cell count (8500/μL) and C-reactive protein level (CRP; 0.77 mg/dL). With regard to renal function, serum creatinine level was elevated at 1.10 mg/dL and the estimated glomerular filtration rate was reduced to 53.97 mL/min/1.73 m^2. In addition, serum level of IgG4 was 70 mg/dl (normal range; 4.8–105 mg/dL). Urinalysis showed no microscopic hematuria or proteinuria. Urine cytology tests, which were repeated 3 times, revealed no signs of

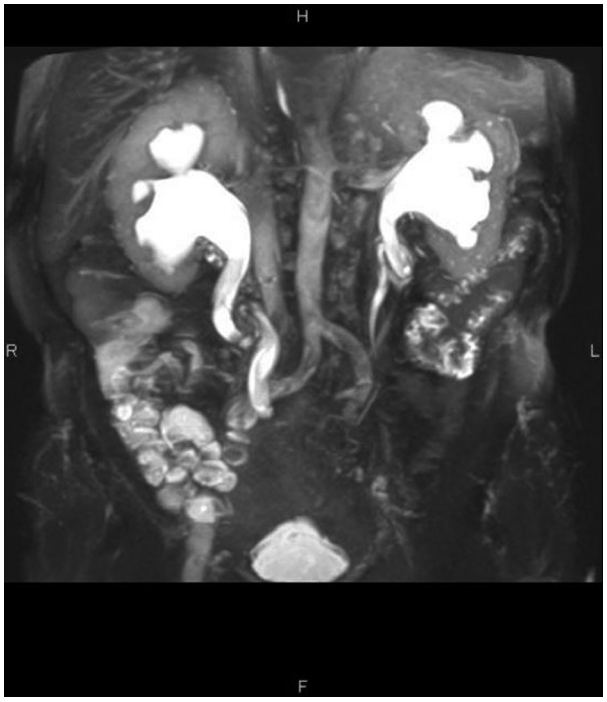

Figure 1. Half-Fourier-acquisition single-shot turbo spin-echo (HASTE) maximum intensity projection (MIP) image showing bilateral hydronephrosis and a hydroureter.

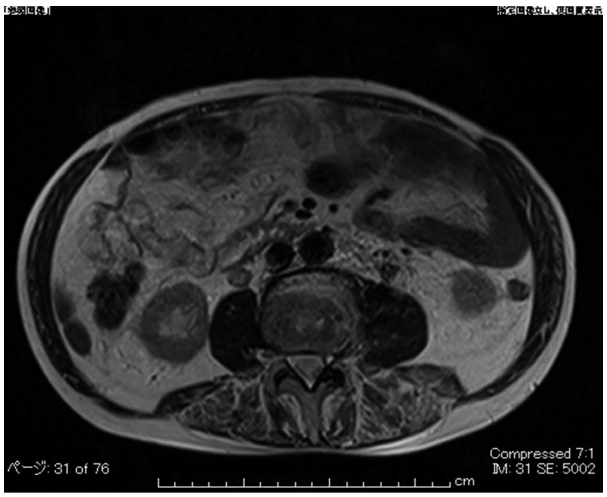

Figure 2. Axial T2-weighted image showing postinflammatory fibrosis around the left ureter at the L3–4 level.

malignancy. From these facts, we recommended that the patient should undergo drip infusion pyelography. However, he decided against this procedure due to adverse events caused by the contrast medium and decreasing renal function. Thus, we planned to perform retrograde pyelography and ureteroscopy to clarify the cause of hydronephrosis. However, MRI was performed prior to these invasive examinations as recommended by radiologists.

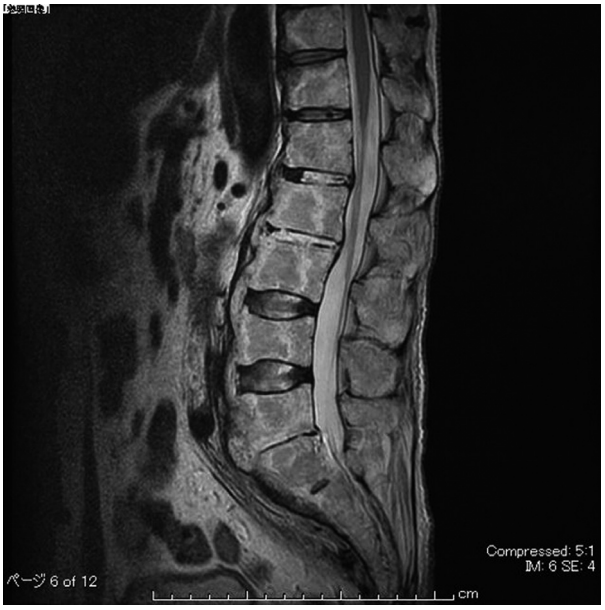

Figure 3. Sagittal T2-weighted image showed the characteristic findings of SAPHO syndrome, such as paravertebral ossification, bony bridges between vertebral bodies, and low signal intensity, and ossification of intervertebral disc.

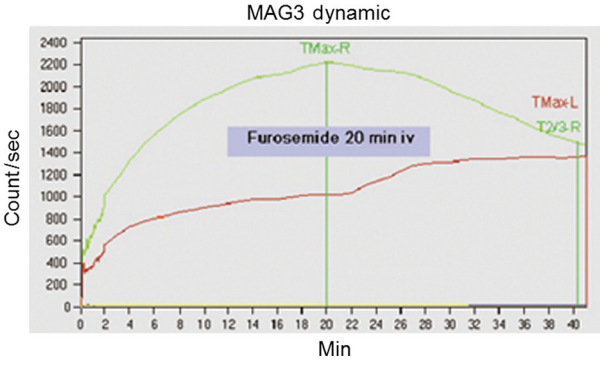

Figure 4. MAG-3 dynamic scintigraphy showed obstructive pattern in bilateral ureters.

Although MRI of the abdomen confirmed right and left ureter dilations to the S1 and L5 level, respectively (Figs. 1 and 2), no obvious mass was detected in the urinary tract. Moreover, MRI showed prevertebral soft tissue involvement of the bilateral lower ureters from the lower lumbar vertebrae to the sacrum (Fig. 3). Bilateral thickness of the lower thoracic to the sacroiliac joints was also detected. These findings are known to be detected in SAPHO syndrome with considerable frequency [3]. We recommended a biopsy of the prevertebral soft tissue for accurate diagnosis. However, the patient refused as he felt burdened.

We ultimately judged that the bilateral lower ureter stenosis was due to retroperitoneal inflammation caused by SAPHO syndrome. In fact, scintigraphy of bilateral kidney showed the urinary stasis (Fig. 4). Based on these findings and symptoms, we placed a JJ catheter for bilateral ureter, however, there were no abnormal findings in the retrograde pyelography. Thereafter his renal function and backache improved.

Discussion

The acronym SAPHO was coined by Chamot et al. [4] in 1987. This condition is relatively rare and has no validated diagnostic criteria. However, many physicians apply the inclusion and exclusion criteria reported by Benhamou et al. [5]. that include osteoarticular manifestations of acne conglobate, acne fulminans, or hidradenitis suppurativa; osteoarticular manifestations with palmoplantar pustulosis; hyperostosis with or without dermatosis; and chronic recurrent multifocal osteomyelitis, involving the axial or peripheral skeleton with or without dermatosis. Although signs of this syndrome are nonspecific, its major symptoms include chest pain and backache because the anterior chest wall and spine are common sites of involvement in SAPHO syndrome [6]. In the present case, soft tissue inflammation around the hyperostosis and an osteitis lesion in front of the lower vertebrae and sacroiliac articulations were detected. Thus, it is not clear whether bone pain or hydronephrosis caused the backache.

This patient had been undergoing treatment for SAPHO syndrome and currently has no SAPHO-related symptoms. We cannot explain why the bilateral hydronephrosis occurred 11 years after onset of the SAPHO syndrome despite the fact that the condition was well controlled. However, because serum CRP level was stable and he had no new symptoms, we have a negative opinion about the worsening systematic inflammation. Another report stated that approximately 8% of patients with SAPHO syndrome have or develop inflammatory bowel disease [7]. Therefore, the local progression of inflammation in the soft tissue near the ureter is possible. On the other hand, we suspected that cause of hydronephrosis is IgG4-related retroperitoneal fibrosis. However, his serum level of IgG4 was in the normal range. In addition, we could not find the report about the relationship SAPHO syndrome and the IgG4-related disease in English literature. Based on these findings, we suggest that local inflammatory change and its related symptoms should be noted during follow-up in patients with SAPHO syndrome. However, limitation of our opinion should be noted. Briefly, pathological diagnosis was not obtained because biopsy of the prevertebral soft tissue was a big burden for the patient.

Hydronephrosis is often an important sign in the diagnosis of urinary stones and tumors of the upper urinary tract. So, we used various examinations, while assessing for the presence of these diseases. Finally, MRI findings ultimately led to the diagnosis and indicated the cause of bilateral hydronephrosis. MRI is considered an essential imaging examination in patients with SAPHO syndrome, and it is believed to have advantages over CT with regard to diagnosis and follow-up [3]. In addition, there is general agreement that MRI is useful for diagnosing upper urinary tract disease, including urothelial cancer.

SAPHO syndrome is generally considered a rare disease with an estimated prevalence of <1/10,000. However, its incidence is likely underestimated because its diagnosis requires a strong clinical suspicion and detailed examinations [8]. If this patient was not receiving treatment for SAPHO syndrome, we probably would have performed an invasive examination to clarify the cause of hydronephrosis. In addition, it is possible that he would have been diagnosed with retroperitoneal fibrosis if the MRI had not been performed. Although there was a report of renal failure due to SAPHO syndrome, the cause of renal failure was amyloidosis of the kidney [9]. This is the first report of a patient with hydronephrosis and renal dysfunction due to SAPHO syndrome.

Conclusions

We reported a case of bilateral hydronephrosis due to SAPHO syndrome. In our patient, MRI was useful to diagnose this pathological condition.

Consent

Written informed consent was obtained from the patient for publishing this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Conflict of Interest

None declared.

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