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Editor
Nancy E. Epstein, MD
Clinical Professor of Neurological Surgery, School of Medicine, State U. of NY at Stony Brook

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Case Report

Destructive spondylodiscitis associated with SAPHO syndrome: A case report and literature review

Shu Takeuchi, Junya Hanakita, Toshiyuki Takahashi, Manabu Minami, Ryo Kanematsu, Izumi Suda, Sho Nakamura

Spinal Disorders Center, Fujieda Heisei Memorial Hospital, Mizukami, Fujieda, Japan.

E-mail: *Shu Takeuchi - shu.tk.0804@gmail.com; Junya Hanakita - heisei.spine-jh@ny.tokai.or.jp; Toshiyuki Takahashi - heisei.t-taka@ny.tokai.or.jp; Manabu Minami - minami@mac.com; Ryo Kanematsu - ryo.knmt@gmail.com; Izumi Suda - izumis306@gmail.com; Sho Nakamura - nsho.1018@gmail.com

***Corresponding author:**
Shu Takeuchi,
Spinal Disorders Center,
Fujieda Heisei Memorial Hospital, Mizukami, Fujieda, Japan.

shu.tk.0804@gmail.com

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ABSTRACT

Background: Synovitis, acne, pustulosis, hyperostosis, and osteitis (SAPHO) syndrome is a rare sterile inflammatory disease characterized by cutaneous and osteoarticular lesions. Associated spinal lesions chronically manifest slight or no neurological symptoms. Only rarely does destructive spondylodiscitis occur.

Case Description: A 62-year-old female with palmoplantar pustulosis presented with a rapidly progressive quadriparesis. When the cervical MR showed destructive spondylodiscitis at the C5–C7 level, the patient underwent anterior debridement followed by posterior reconstruction/fixation. The histopathology showed a nonspecific inflammatory process with vertebral sclerosis consistent with the diagnosis of SAPHO; cultures were negative. Postoperatively, the patient's symptoms improved and SAPHO did not recur.

Conclusion: Destructive spondylodiscitis associated with SAPHO syndrome is uncommon. Early diagnosis and surgical treatment result in the best outcomes.

Keywords: Destructive spondylodiscitis, SAPHO syndrome, Spine surgery

INTRODUCTION

Synovitis, acne, pustulosis, hyperostosis, and osteitis (SAPHO) syndrome, a noninfectious inflammatory disease, can rarely cause destructive spondylodiscitis.[8] Following sternocostoclavicular lesions (65~90%), the spine is the 2nd most commonly affected region (32~52%).[3] Notably, SAPHO syndromes rarely cause spinal deformity, and only rare cases require reconstructive surgery.[1,5,6,8,10] Here, a 62-year-old female with SAPHO syndrome and destructive C5–C7 spondylodiscitis warranted circumferential decompression/fusion.

CASE DESCRIPTION

History and examination

A 62-year-old female presented with progressive shoulder pain and quadriparesis that worsened over a 6 day course in the hospital. On physical examination, she had evidence of the SAPHO syndrome characterized by synovitis, acne, pustulosis, hyperostosis, osteitis, palmoplantar pustulosis, and acne on anterior chest wall [Figures 1a and b]. Labs studies showed just a mild inflammatory reaction; her C-reactive protein was 4.61 mg/dL, the erythrocyte sedimentation rate was 90 mm/H, and her white

blood cell count was 6,260/μ,; she remained afebrile. Blood cultures, the rheumatoid factor, and polymerase chain reactions of the tubercle bacillus were also all negative.

Diagnostic work-up

Plain cervical X-rays showed destruction of the C5–C7 vertebral bodies with kyphosis [Figure 2a]. The CT demonstrated C5–C7 lytic changes with marginal sclerosis, while the MR revealed spondylodiscitis with severe C5–C7 cord compression [Figures 2b-d]. Whole body 18F-fluorodeoxyglucose positron emission tomography (18F-FDG PET/CT) demonstrated increased uptake involving the sternocostoclavicular joint, tonsils, and C5–C7 vertebrae [Figures 3a-c].

Circumferential C5–C7 Surgery

She underwent an anterior C5, C6 corpectomies with C4–C7 fusion (i.e., using iliac crest autograft and a plate) under microscopic visualization. Erosive granulomatous tissue was removed, but no abscess or solid tumor were encountered [Figure 4]. One week later, a posterior fusion from C2 to Th1 was performed with pedicle and lateral mass screws.

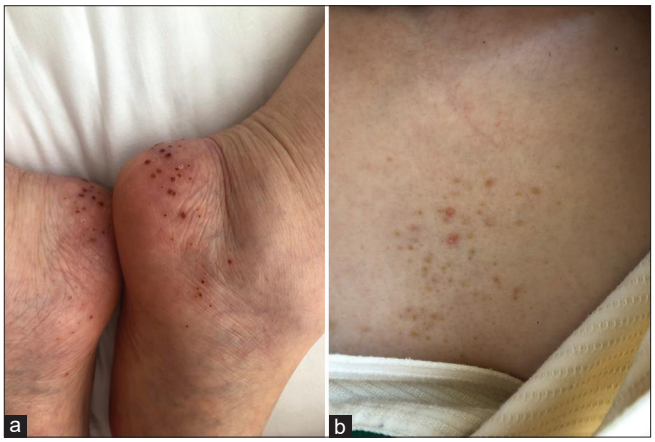

Figure 1: Physical examination revealed pustulosis on the sole of feet (a) and acne on anterior chest (b).

Histological diagnosis

The histopathological examination was consistent with SAPHO syndrome. The findings included a sterile inflammatory infiltrate composed of neutrophils and lymphocytes. Within the C5 and C6 vertebral bodies, bony trabeculae and sclerotic changes were observed.

Postoperative course

Postoperatively, her quadriparesis immediately resolved and imaging studies demonstrated decompression of spinal cord with correction of the kyphosis [Figures 5a and b]. She was discharged without any complications; 2-month later, she remained disease-free.

DISCUSSION

SAPHO syndrome is rare nonbacterial inflammatory disease, characterized by cutaneous and osteoarticular lesions, occurring in 1/10,000 in Europe but only in 0.000285/10,000 in Japan.[7] This multifactorial disorder usually occurs in patients ages 30–50.[7,9] Patients usually present with osteoarticular lesions involving the anterior chest wall (65~90%), followed by the spine (32~52%).[3] Radiological findings usually include vertebral body osteosclerosis, hyperostosis, and osteitis, but only rarely, which are associated with destructive lesions.[2,4,11] The differential diagnosis typically includes; infection, osteomyelitis, or bone neoplasms.[3] Acute neurological deficits are only rarely encountered (i.e., rapidly progressive myelopathies).

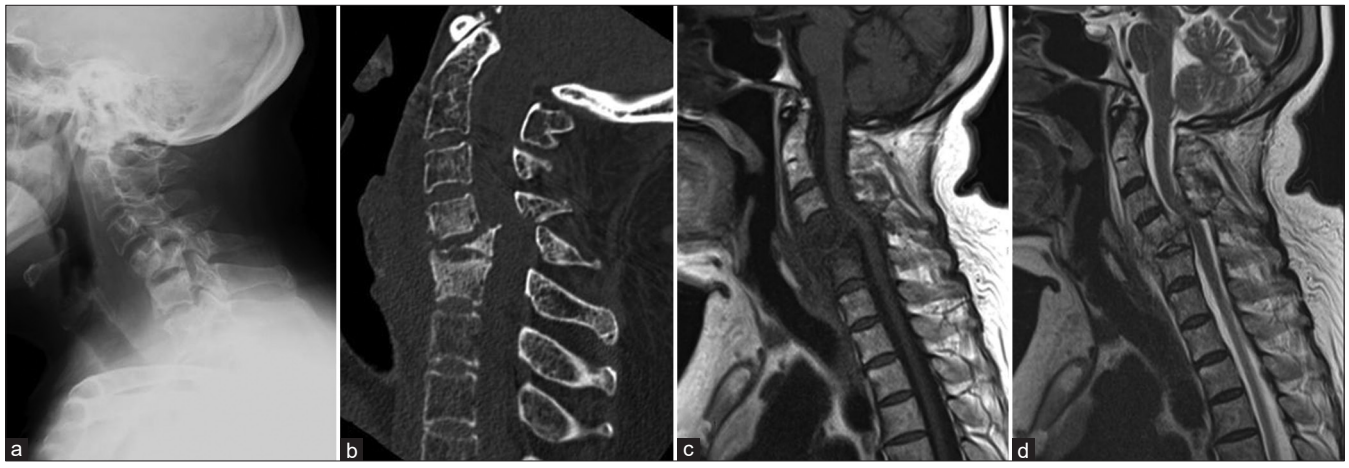

Figure 2: Preoperative images. (a) Cervical plain radiography shows kyphotic deformity of C5–C7 vertebral bodies. (b) Sagittal CT scan demonstrates remarkable destructive change at C6 vertebra. (c and d) Midsagittal MRI of cervical spine shows highly tortuous spinal cord and compressed vertebrae of C5, 6 with T1 hypointense signal, T2 iso-hyperintense signal.

Table 1: Clinical demographics of SAPHO syndrome with destructive spondylodiscitis.

Author and Year	Age, Sex	Level	Symptom	Other lesions	Treatment	Outcome
Deltombe *et al*., 1999	74, M	C6–C7	Cervical pain and progressive limb weakness	Psoriasis of the calf, Sternoclavicular joint	Conservative treatment	Moderate paresis, persistent cervical kyphosis
Takigawa *et al*., 2008	63, F	C4–C7	Cervical pain and progressive limb weakness	PPP	Anterior decompression and anterior-posterior C3–C7 fusion	Slight hypesthesia on right hand
	69, F	T7–T9	Severe back pain	Sternocostoclavicular and sacroiliac joint	Anterior decompressiion and autologous reconstruction	Improved
Nakamura *et al*., 2010	60, F	L4–L5	Back pain. leg numbness and weakness	Sacroiliac joint	Anterior decompression and posterior L4-L5 fusion	Improved
Rekik *et al*., 2015	50, M	T4–T5	Back pain, progressive leg weakness	PPP	Posterior decompression and fusion	Improved
Nakamae *et al*., 2019	73, M	L3	Progressive lower back and leg pain	PPP, Sternocostoclavicular joint	Posterior decompression and L2-L4 fusion	Improved
Funayama *et al*., 2020	22, F	C4–C7	Difficulty looking up	PPP, Sternoclavicular and sacroiliac joints, sternum	Three-stage surgery with C7 resecion and C2-T3 fusion	Improved

PPP: Palmoplantar pustulosis

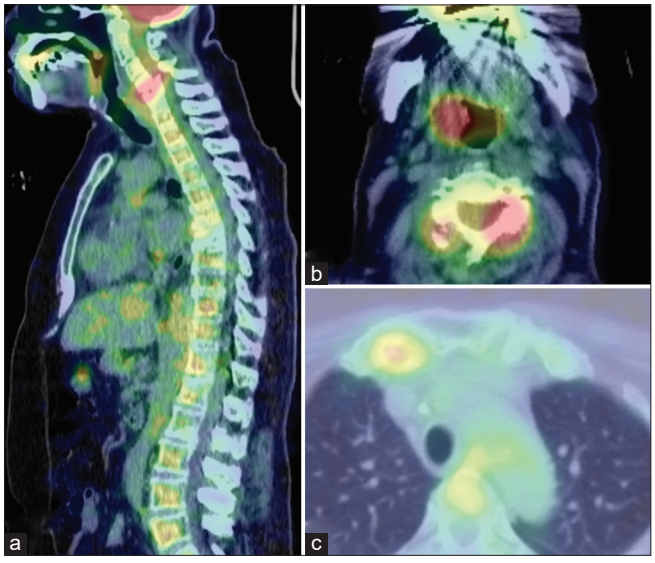

Figure 3: 18F-fluorodeoxyglucose positron emission tomography (18F-FDG PET/CT) shows an abnormally high uptake of cervical spine (a), tonsils (b), and sternocostoclavicular joint (c).

Literature review

From the literature, we identified seven cases of SAPHO syndrome accompanied by destructive spondylodiscitis and neurological deficits [Table 1]. Patients averaged 58.7(22~74) years of age; four were female. Lesions involved the cervical (three cases), followed by the lumbar (two cases), and thoracic spine (two cases). It took an average of 4.2 months to establish the diagnosis. Only one patient was treated conservatively with a cervical orthosis that resulted in a slow recovery. The remaining six patients successfully underwent surgery with complete resolution of their neurological deficits.

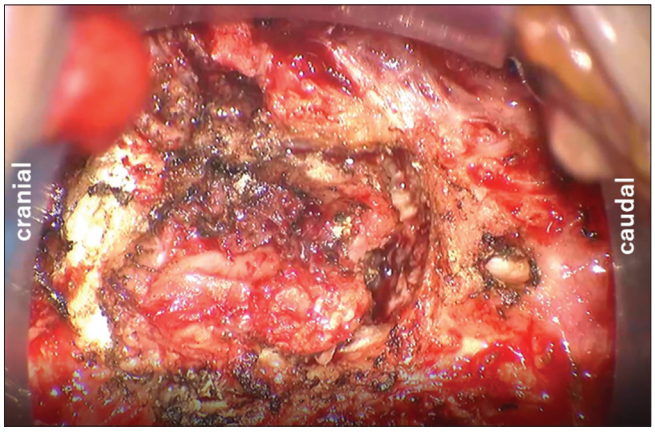

Figure 4: Intraoperative microscopic views. Granulomatous tissue was observed at anterior part of C5,6 vertebrae.

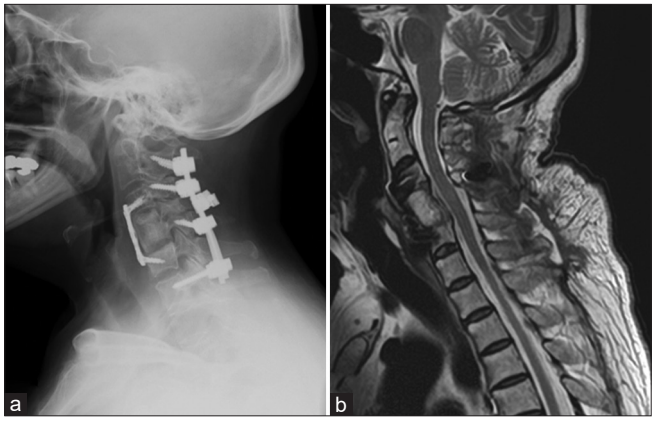

Figure 5: Postoperative images. (a) C5–C6 corpectomy and anterior fixation in the C4–C7 were performed, and posterior instrumentation was fitted from C2 to Th1. The alignment was well corrected. (b) The compression of cervical spinal cord improved.

CONCLUSION

Although SAPHO syndrome is usually chronic, those who rarely develop spinal spondylodiscitis may present with acute neurological deterioration warranting timely operative intervention.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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