

## **Brachialgia-Unusual Presentation of SAPHO Syndrome A case report and review of the literature**

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**Abstract:** SAPHO Syndrome is an acronym of Synovitis Acne Pustulosis Hyperostosis and Osteomyelitis. The syndrome is characterized by variable association of atypical osteo-articular manifestations and various chronic dermatological conditions. We present a case of SAPHO syndrome with unusual neurological presentation and diagnostic challenge. To our knowledge there is no report of SAPHO syndrome presented with neurological compressive symptoms and brachialgia. We found Nuclear Bone Scan is diagnostic when an unusual presentation and other diagnostic tools fail. Our case highlights that in SAPHO syndrome, juxta-articular compressive manifestation might be sign of severity. This may suggest the need for stage IV as a supplement to the grading of severity.

**Keywords:** Brachialgia, SAPHO Syndrome, Classification.

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### **INTRODUCTION**

SAPHO Syndrome is a unifying acronym to include various features of closely related disorders, namely Synovitis Acne Pustulosis Hyperostosis and Osteomyelitis. It was initially coined for the constellation of the symptoms in 1987[1]. The syndrome is characterized by variable association of atypical osteo-articular manifestations and various chronic dermatological conditions, in particular palmo-plantar pustulosis and severe acne. It manifests as various skin disorders and osteo-articular inflammation closely mimicking other disease categories. Its natural course is still not well defined[3,4]. Genetic, immunological, and bacterial mechanisms are implicated in the development of the disease pathogenesis. It has various courses from self-limiting in minority of patients to a relapsing-remitting pattern or chronic indolent pattern after an early acute phase. There are studies demonstrating SAPHO syndrome is either under recognised or under reported[2].

Diagnostic problems may arise due to incomplete manifestations of SAPHO syndrome or non-simultaneous manifestation of each component. Furthermore, all components need not be present at same time for the diagnosis[1,5].

Here we present a case report of SAPHO syndrome with unusual presentation and diagnostic challenge. To our knowledge there is no report of SAPHO syndrome presented with neurological compressive symptoms and brachialgia.

### **CASE REPORT**

Mr JN is 46 years old unemployed and teetotaler. He was referred by his doctor after 2 years of intermittent left non dominant shoulder pain which got worse over 6 weeks prior to his visit in our Orthopaedic Clinic. In the previous 9 months he had progressively worsening left shoulder pain and inability to lift his arm above shoulder level. He had paresthesia and episodes of acute electric shock like sensation going down to his left arm. He had no history of trauma. He is ex-taxi driver. He is not keen in any sports activity. He has no paraneoplastic symptoms. No recent history of travel.

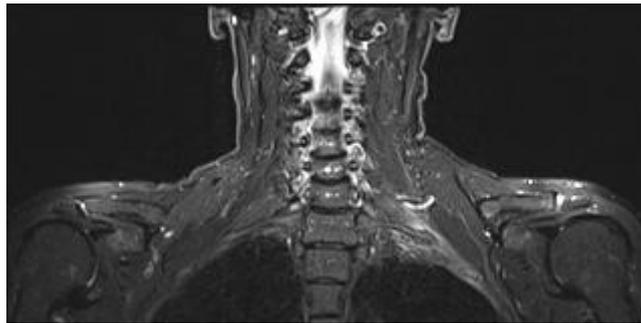
He has history of Fibromyalgia in 1980 and mild Discoid Psoriasis since 2004. In the past 10 years he's had four episodes of spontaneous pneumothoraces treated conservatively. At present, he has no other pertinent past medical and surgical history. He was not on any medications.

Examination shows no obvious swelling and skin colour changes around neck and shoulder. On pressing the supraclavicular fossa, it reproduced numbness and paraesthesia in his arm down to all fingers. His shoulder movements were limited on the extremes due to pain. Neurological examination on his left arm showed reduced sensation to touch and pain on C4 to C8 distribution. He had mild weakness MRC 4+/5 flexors power but normal tone, reflex and coordination. He has normal circulation and no peripheral bruit. Adson & Roos tests were normal. No active skin lesion

noted elsewhere. Examination of neck and elbow was unremarkable.

Investigations showed normal white cell count and differential but persistently moderately elevated CRP and ESR 53.4 and 64 respectively. He had elevated Alkaline phosphates 522 IU/l. The rest of biochemical tests including PSA and Myeloma screening were all normal.

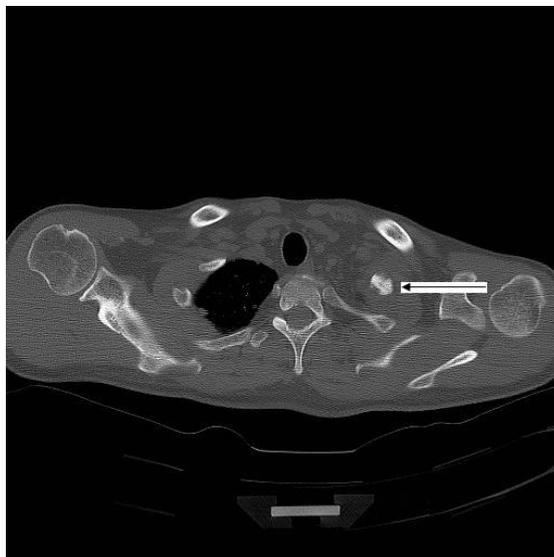
His initial Chest and Left Shoulder radiographs showed no bone or joint abnormality. MR of brachial plexus, cervical spine and shoulder showed no traumatic nerve root injury or any space occupying lesion overlying the distal brachial plexus. There was no evidence of cervical rib either. The cervical spine sequences showed no nerve root compression. On STIR sequences of the thoracic inlet, Figure 1, there was significant marrow oedema noted of the first rib with abnormal signal change around the adjacent brachial plexus at preclavicular area.



**Fig-1: MRI STIR coronal sequence shows oedema in the region of left brachial plexus adjacent to first rib.**

The first rib abnormality was thought to be due to a stress fracture, infection, inflammation (Osteitis) or malignancy. Acute infection was less likely due to low clinical probability and biochemical markers.

On MR image of the left shoulder joint showed features of chronic impingement which would explain to some extent the shoulder symptoms. Further assessment with CT scan showed abnormal texture of the 1st rib on the left side with mixed lytic and sclerotic lesions, figure 2a and 2b.



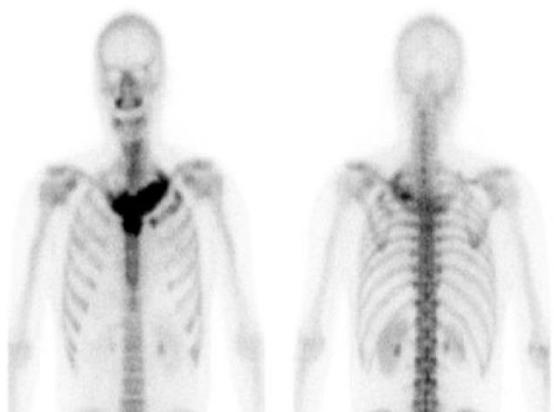
**Fig- 2a: CT showing sclerotic lesion of the 1<sup>st</sup> rib (Arrow).**



**Fig-2b: CT show expansion & sclerosis of medial clavicle and sternum.**

There was no aggressive periosteal reaction or any features of a healing or partially healed fracture. There were further abnormalities noted especially sclerosis and chronic erosive changes in and around both sterno-clavicular joints and in the right first rib. These features did not suggest an acute infection or malignancy but a more acute-on-chronic inflammatory

process. Nuclear bone scan, figure 3 showed the typical “bull’s head” pattern of intensely increased activity in bilateral sternoclavicular joints, 1<sup>st</sup>, 2<sup>nd</sup> rib and medial end of clavicles. No other long bone, spine or other joint was involved. Final diagnosis of SAPHO syndrome was confirmed. Nerve conduction test was not required.



**Fig-3: Tc<sup>99m</sup> Nuclear bone scan shows tracer uptake & expansion of sternum & first ribs, described as “Bull’s head” lesion.**

## DISCUSSION

SAPHO syndrome has been described as constellation of synovitis, acne, pustulosis, Hyperostosis and Osteomyelitis in 1987 by AM Chamot et al[1] after observation of a series of patients. It is described as rare or under diagnosed due to discordance of symptoms or diagnostic features appearing in different time span.

In young or in middle-aged adults the disease predominates in the sternoclavicular region followed by the spine, pelvis and long bones including mandibles. Sternoclavicular region is the most frequent site of the

disease (65–90%) followed by the spine as the second most common site involving 33% of cases[5].

SAPHO syndrome tends to have a protracted with waxing and waning course that vary greatly from patient to patient. Most patients seem to have a fairly good prognosis, but in some, SAPHO syndrome may become severe disease leading to persistent pain and disabling. Because of the rarity of the disease, unknown aetiology and pathomechanism; targeted therapy is still unavailable. Treatment involves symptomatic, antimicrobials, aminobisphosphonates and other disease-modifying drugs (DMARDs) such as

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methotrexate, sulfasalazine, and anti-tumour necrosis factor (anti-TNF) agents[6].

Three stages of SAPHO syndrome has been described[7].

**Stage 1** Disease localised to costoclavicular ligament. It may be a primary enthesopathy.

**Stage 2** Disease progress into arthropathy in the sternoclavicular joint and sclerosis involving the medial end of the clavicle; the first rib and the adjacent sternum with sclerotic hypertrophy of the costal cartilage.

**Stage 3** is a continuum with osteosclerosis, hyperostosis and bone hypertrophy affecting the medial ends of the clavicles, the sternum and upper ribs with arthritis in the adjacent joints.

The radiological features are those of synovitis, arthritis and chronic inflammatory bone disease. The characteristic radiological features are predominated by hyperostosis and osteitis. Nuclear medicine is extremely sensitive in the detection of anterior chest wall lesions. The “bull’s head” pattern of increased activity is a highly specific sign and may even obviate the need for biopsy. CT scanning is the optimal modality to demonstrate the osteoarticular lesions. Targeted scans, particularly with multislice CT and reformatted images, will accurately demonstrate the extent of involvement. MR imaging will demonstrate the changes due to arthritis and bone marrow oedema utilising fat suppressed T2-weighted or STIR sequences.

Our case presented had an unusual presentation. It was diagnostic challenge mimicking various pathologies. His symptoms fall into Stage III and severe form of SAPHO syndrome. He is on Steroids and Methotrexate. His neurologic symptoms started to improve after three weeks of treatment but he is still symptomatic occasionally. He is on a long term follow up.

Meticulous history, physical examination and appropriate investigation is a key for diagnosis of unusual and rare pathologies. Our case confirmed that Nuclear Bone scan is investigation of choice when SAPHO syndrome is suspected.

This case may highlight that in severe cases juxta-articular compressive manifestation might be sign of severity. This may suggest the need for Stage 4, an additional stage, to supplement the grade of severity.

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