

## Case Report

# SAPHO: an Unusual Cause of Pulmonary Nodules

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### ABSTRACT

SAPHO is a clinical syndrome comprising of synovitis, acne, pustulosis, hyperostosis and osteitis. We report a case of SAPHO presenting in a child referred for investigations of persistent radiological abnormalities in

the chest. Pulmonary involvement in SAPHO has been reported rarely, is asymptomatic and may be more common than has been described.

KEYWORDS : child, pulmonary nodules, lung disease, SAPHO

### INTRODUCTION

SAPHO is a clinical syndrome comprising of synovitis, acne, pustulosis, hyperostosis and osteitis. Pulmonary involvement has been described rarely but this cause of multiple nodular lesions on chest radiography should be differentiated from other possible diagnoses as it follows a benign course and requires no specific therapy. Here we present a case of SAPHO presenting in a child referred for investigation of persistent radiological abnormalities in the chest.

### Case presentation

A ten-year-old boy was assessed for persistent lung X-ray abnormalities eleven months after a primary illness with myalgia and an erythematous rash with pustular eruptions on the soles of both feet. He made an initial recovery but two months later he developed a widespread, papular eruption with pustule formation. This was most florid on the exposed areas of the limbs and was associated with arthralgia, abdominal pain and muscle weakness. He was investigated for an infective cause of this illness but cultures and serology were not positive. During the ensuing six months the rash relapsed and remitted.

Four months after his initial symptoms, he developed swelling of the left angle of his mandible. A dental abscess was diagnosed and he had extraction of a primary molar. Cytological examination of material from the site demonstrated non-specific, chronic inflammatory changes. One month later he developed another swelling over the

medial end of his left clavicle. This persisted for one month and resolved spontaneously. Chest X-rays taken six weeks apart showed multiple lesions in the left lower lobe and costophrenic angle. He had no respiratory symptoms at this time but the radiological lesions persisted for six months and he was referred to us for advice.

Examination demonstrated a swelling over the left sternoclavicular joint (Fig. 1) and scaly papules with occasional pustules distributed mainly on his trunk but with peripheral lesions (Fig. 2). Biopsy of a skin lesion demonstrated mild, superficial, perivascular dermatitis. He had no pulmonary symptoms or signs and his pulmonary function was normal. Computed tomography of the chest (Fig. 3) showed areas of density at both lung bases that were consistent with areas of radiological abnormalities on plain chest X-ray (Fig. 4). There was no evidence of interstitial lung disease. The lesions were judged to be previous areas of infarction with subsequent residual fibrosis. Extensive investigations, including autoimmune profile, atypical pneumonia screen, Borrelia titers, T and B cell subsets, neutrophil function, antiHib and Tetanus titer and Mantoux test did not demonstrate any evidence of autoimmune disease, immune deficiency or infection. His lung function tests were normal and bone scan showed areas in his right clavicle consistent with osteitis.

We believe that the combination of osteitis of the clavicle and plantar pustulosis in our patient is consistent with SAPHO syndrome.

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Fig. 1: Swelling over the left sternoclavicular joint

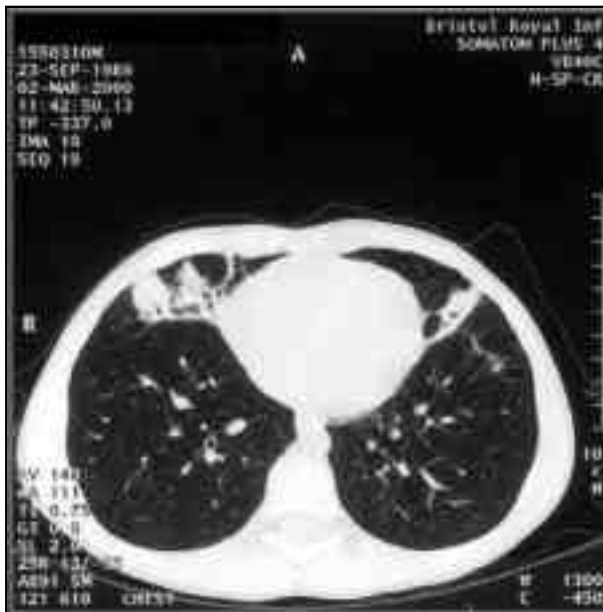


Fig. 3: CT chest showing areas of reported residual scarring

## DISCUSSION

SAPHO was first described in 1987<sup>[1]</sup> and is characterized by variable bony involvement with or without a concurrent pustular dermatosis. Affected bones include the chest wall, sacroiliac joints and long bones. Bony changes include hyperostosis, aseptic osteomyelitis, and arthritis. Skin involvement is also variable and includes one or more of the following: palmoplantar pustulosis, acne conglobata or fulminans, hidradenitis suppurativa, pustular psoriasis and dissecting cellulitis of the scalp.

Benhamou and others described four major diagnostic criteria to establish the diagnosis<sup>[2]</sup>. Any one of the following is regarded as sufficient to diagnose SAPHO:

- Acne conglobata, acne fulminans, or hidradenitis suppurativa with osteoarticular manifestations



Fig. 2: Scaly papules with occasional pustules on the trunk



Fig. 4: Bilateral linear densities at lung bases.

- Palmoplantar pustulosis with osteoarticular manifestations
- Axial or appendicular hyperostosis with or without a sterile pustular dermatosis
- Chronic recurrent multifocal osteomyelitis involving the axial or appendicular skeleton with or without a pustular dermatosis

SAPHO has been described mainly in children but also young to middle-aged adults. Most of the published cases have been reported from Japan or Northern and Western Europe. Discrepancies in the reported prevalence may relate to under recognition or under-reporting of the condition although it has been suggested that ethnic immunogenetic variation may account for some of the geographical heterogeneity in prevalence<sup>[3]</sup>. The cause of SAPHO remains unknown. An infectious aetiology has been suggested and *Propiono-bacterium acnes* has been isolated from the lesions<sup>[4]</sup>. *P. acnes* is an anaerobic, Gram positive bacillus that has been implicated in severe cases of acne but it is a common skin saprophyte and its presence does not necessarily imply causation.

The course of SAPHO is usually relapsing and remitting. Bone lesions may persist for many years and hyperostosis remains stable on sequential radiological examinations. Pulmonary involvement in SAPHO has been reported rarely<sup>[5-7]</sup> although it may be more common than previously described, as it appears to remain asymptomatic and detection relies on fortuitous discovery of the associated radiological abnormalities. The major implication of making this diagnosis is to differentiate the pulmonary lesions from other potential causes and avoid the need for unnecessary investigations or treatment. The presence of pulmonary lesions in SAPHO does not appear to alter the benign, albeit protracted, nature of the condition. However, the

involvement of the lungs in this condition is still poorly understood and there may be a case for pulmonary assessment of all patients newly diagnosed with SAPHO.

## CONCLUSION

SAPHO is an important clinical entity that may be confused with other less benign conditions affecting the skin, bones and lungs of children and young adults. The history and pattern of skin disease is often the key to the diagnosis and early recognition of this syndrome can prevent unnecessary and potentially hazardous investigations and treatment.

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