

A rare case of the SAPHO syndrome with pulmonary complications

G Ngom, A Désiré, J Savadogo, M Fall, O Ndour, I Fall, M Ndoeye

Citation

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Abstract

We report a case of SAPHO syndrome with pulmonary complications. A fifteen-year-old boy with a history of pustulosis presented with chronically swollen legs and left hemi thorax. The clinical presentation, the negative bacteriology, the lesions on radiology and the histology were in favour of the syndrome. Particularities of this case were pulmonary complications, cutaneous fistulas and a tibial fracture observed throughout the clinical course. The outcome was favourable after a treatment of non-steroidal anti-inflammatories.

INTRODUCTION

The SAPHO syndrome, an acronym of synovitis, acne, pustulosis, hyperostosis, osteitis, was first defined by a French rheumatologist in 1987 following a national survey [1]. It is characterised by multiple, chronic bony lesions, a negative bacteriology associated with dermatological lesions and a clinical course of exacerbations and remissions. The pathogenesis is still unknown. Nevertheless, the accompanying inflammation and the favourable response to anti-inflammatory treatment, suggest a primary inflammatory osteopathy- primitive and rheumatologic, that is to say, neither infectious, nor cancerous nor dystrophic [1]. Extra-osseous and extra-cutaneous lesions have been described as part of this syndrome. Of note here are lesions within the digestive track and vascular lesions [2]. Less common is the pulmonary lesion which Kerem first described in Canada in 1989 [3]. We report a case of this syndrome, characterised by the symmetry of the bony lesions and their fistulisation, the presence of a pulmonary lesion and a pathological fracture.

CASE REPORT

KL, aged 15, was brought to our services for the management of a bilateral tibial osteitis, present for the past 8 weeks. The child had been receiving antibiotic treatment in the form of amoxicillin and gentamycin for the past 4 weeks without improvement. The history revealed bilateral pulmonary pustuloses which first appeared 2 years ago but had spontaneously regressed. No family history of the condition was found. Physical examination on admission

revealed a good general state of health, normal temperature, normal height and weight for age and pale conjunctivae. A bilateral, symmetrical inflammatory swelling over the anterior tibial tuberosities was noted. Multiple fistulas productive of bloody serological fluid were present over each swelling. An inflammatory swelling was seen in the anterior thorax, over the medial third of the left clavicle. Auscultation revealed crepitations over both lung bases, predominantly over the right and a systolic murmur. The full blood count revealed a normochromic normocytic anaemia of 6.6g/dL. The C reactive protein and erythrocyte sedimentation rate were normal. A test for sickle cell anaemia was negative. Bacterial cultures from the products of the fistulas were negative. Sputum acid-fast bacilli testing was negative. Standard radiographs of the lower limbs showed bilateral hyperostosis and tibial metaphyseal diaphyseal osteitis (Figure 1).

Figure 1

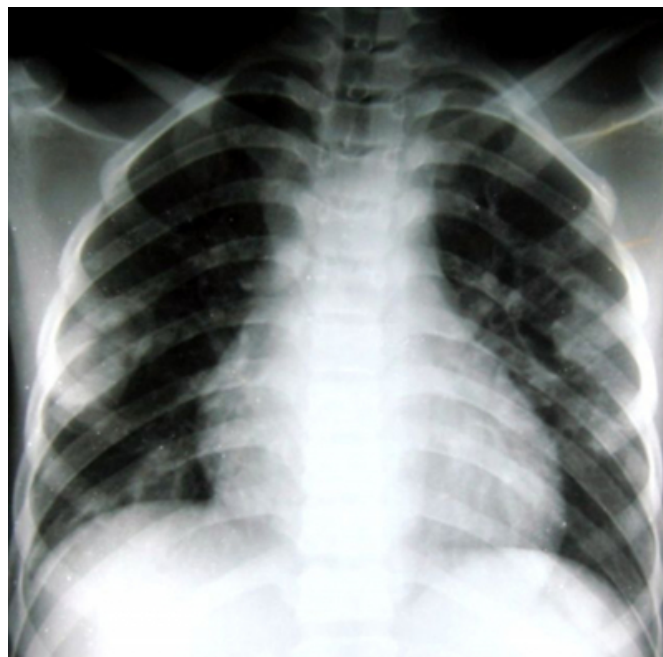
Figure 1: X ray of the right lower limb showing a hyperostosis and a tibial osteitis.



The chest X ray showed a left clavicular hyperostosis and pulmonary cavities associated with a right hilar opacification suggestive of an infectious pneumonia (Figure 2).

Figure 2

Figure 2: Chest X ray showing right clavicular hyperostosis and lung parenchyma opacities.



X rays of the pelvis and spine were normal. A CT scan of the thorax found multiple bullous lesions at the lung bases, more numerous on the right and of varying sizes. The bullae were surrounded by thick walls suggestive of an inflammatory process in evolution. An interstitial opacity was also noted, at the right hilar region with two central bullae (Figure 3).

Figure 3

Figure 3: CT scan of the thorax showing a right hilar opacity and bilateral parenchymal bullae.



The pathology report from a biopsy taken from one of the skin lesions noted a chronic ulceration with an important fibrosis component associated with inflammatory

polymorphs. A biopsy from the clavicle and from the left tibia revealed an area of organising fibrosis with scattered plasma cells. The diagnosis of SAPHO syndrome was made based on Kahn's three criteria. A treatment was administrated including iron for 2 months and non-steroidal anti-inflammatories for 8 weeks. The patient was reviewed three weeks after this admission to hospital and had made a favourable progression: the cutaneous fistulas had closed, swellings of the clavicle and legs had disappeared and the chest X rays were normal. But the X ray of the lower limbs showed a pathological fracture of the left tibia (Figure 4).

Figure 4

Figure 4: X ray of the left lower limb showing a tibial fracture.



This was treated by three-month plaster cast immobilisation. Further questioning revealed that the patient had undergone excessive physiotherapy at home.

DISCUSSION

The SAPHO syndrome is a rare illness which groups

together a set of illness that are as yet poorly understood [1]. Cases of this syndrome have been described in children [2,3] before this one; it is therefore not an illness exclusively of young adults. Kahn [4] reports extremes of age from 10 to 70 years. The pathogenesis is, as yet, practically unknown [3]. In this patient, as in cases reported by other authors [2,3], no gene has been isolated and the rare positive bacterial culture findings reported in the literature appear to be exceptions [5]. Schilling [2] suggests that the syndrome may be due to an immunopathological process and Bahri [6] implicates the HLA B27 antigen.

Different dermatological lesions have been associated with this illness. In contrast to this, pustulosis, which was present in the history of this patient, is not always implicated. It first appeared more 2 years previously, it was not concomitant with the osseous lesions. Schilling and Bahri [2,6] have previously reported this clinical dissociation between cutaneous and osseous signs. Apart from acne, which not present in this case, other cutaneous lesions have been reported [6]: psoriasis, cellulites of the scalp, pyoderma gangrenosum, hydradinitis suppurativa.

The diagnosis of the SAPHO syndrome is based on clinical findings, X-ray imaging and histology. The diagnosis is straight forward provided the bony and cutaneous lesions are typical in character, complete and found in characteristic sites [1,4,7,8,9,10]. In this case, the tibial and clavicular sites, characteristic of this syndrome, were the key determinants of the diagnosis. However, with bilateral and symmetrical appendicular sequelae associated with visceral complications as reported here, the diagnosis is less obvious [1,11]. Although the multinodal nature of the bony lesions in the SAPHO syndrome has been reported [1], the pathological fracture seen in this case is unique due to its mechanism. It was brought about by intensive massages on a fragile metaphyseal plate and looking like a cancerous or infectious process. The absence of peri-nodal lesions and a negative bacterial culture, however, mean that the exact process is still unclear. The left clavicular swelling we report here, with a hyperostotic lesion on X ray has been reported by Kerem [3] in both clavicles, who found the lesions in more less the same sites as reported here. Chamot [1] has described a sterno-costo-clavicular hyperostosis typically affecting both clavicles and Quelquejay [7] a left clavicular lytic lesion, both external and internal. Laumonier [5] reports the external malleolus and a distal femoral epiphysis being affected. Bony lesions of the SAPHO syndrome, generally multiple, also affect the anterior chest wall, the rib cage, the spine and

the pelvis [6,9]. Solitary bony lesions have also been reported [2]. The SAPHO syndrome is therefore a chronic illness with an evolving course of relapses and remissions [1,7] and its evolution may be exacerbated by complications affecting the viscera. This patient presented with an inflammatory illness of the lungs which was resolved with anti-inflammatory treatment. Imaging therefore plays an important role in the diagnosis of this syndrome [6]. In this case, neither scintigraphy nor MRI was used. The radiological and CT lesions that were found in the lung parenchyma in this case were also observed by Kerem [3], but to a lesser extent. Bullae, more numerous and peripheral in this case, were not exempt from rupturing. Their persistence, after antibiotics were stopped and their subsequent disappearance concomitant with the opacities after the use of non-steroidal anti-inflammatories, confirm their inflammatory nature. The differential diagnosis essentially consists of the incomplete forms of this syndrome with infectious, cancerous or dystrophic pathologies [1,2]. In spite of the many clinical presentations, the use of Kahn's criteria [4], makes the diagnosis easier. The fibrosis and infiltration of plasma cells seen in the biopsy analysis in this case, suggesting a histopathological process of aseptic sclerosis, has also been described by Kerem [3] and Schilling [2]. The symptomatic treatment of the SAPHO syndrome is primarily the use of non-steroidal anti inflammatories [1,2,5]. Other treatments used in the management of this syndrome, are still under discussion and numerous other therapies related to the possible aetiology are still under review [3]. The management of a patient with this syndrome is multidisciplinary in nature [1,2] and requires a better diagnostic approach, rigorous observations to avoid unnecessary repeat investigations and intensive treatments that are often without good basis.

CORRESPONDENCE TO

Gabriel Ngom Postal code : 6863 Dakar-Etoile Phone number : 00221 77 552 00 80 E-mail : gngom2004@yahoo.fr

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Author Information

Gabriel Ngom, MD

Department of Paediatric Surgery, Aristide Le Dantec Hospital

Alumeti Munyali Désiré, MD

Department of Paediatric Surgery, Aristide Le Dantec Hospital

Julien Savadogo, MD

Department of Paediatric Surgery, Aristide Le Dantec Hospital

Mbaye Fall

Department of Paediatric Surgery, Aristide Le Dantec Hospital

Oumar Ndour

Department of Paediatric Surgery, Aristide Le Dantec Hospital

Ibrahima Fall, PhD

Department of Paediatric Surgery, Albert Royer Hospital

Mamadou Ndoye, PhD

Department of Paediatric Surgery, Aristide Le Dantec Hospital