A rare case of the SAPHO syndrome with pulmonary complications
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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 tract 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 metaphysodiaphyseal osteitis (Figure 1).
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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).

**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 [5].
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 [1].
Schilling [1] 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 [8,9] 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 [1]: psoriasis, cellulites of the scalp, pyoderma
gangrenosum, hyadrinitis 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 [1,4,7,9,10]. 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 [11,12]. 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
[1] in both clavicles, who found the lesions in more less the
same sites as reported here. Chamot [2] has described a
sterno-costo-clavicular hyperostosis typically affecting both
clavicles and Quelquejay [1] a left clavicular lytic lesion,
both external and internal. Launomier [1] 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 [13]. Solitary bony lesions have also been reported
[1]. The SAPHO syndrome is therefore a chronic illness with
an evolving course of relapses and remissions [14] 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 [1]. 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 [1], 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 [15,16]. In spite of the many clinical
presentations, the use of Kahn's criteria [17], 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 [1] and Schilling [1]. The symptomatic
treatment of the SAPHO syndrome is primarily the use of
non-steroidal anti inflammatories [15,18]. 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 [1]. The
management of a patient with this syndrome is
multidisciplinary in nature [11,12] and requires a better
diagnostic approach, rigorous observations to avoid
unnecessary repeat investigations and intensive treatments
that are often without good basis.

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