Arthropathy and cutaneous manifestations in a 28-year-old patient with cystic fibrosis

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ABSTRACT: Arthropathy and Cutaneous Manifestations in a 28-Year-Old Patient with Cystic Fibrosis. S. Lampert, S. Pour Schahin, G.H. Wiest, E.G. Hahn, J.H. Ficker.

We report on a 28-year-old man with known cystic fibrosis who presented with pain and cutaneous nodules in the elbow joints. His symptoms had appeared episodically in the previous months, they were always self-limiting, and independent of pulmonary exacerbations. A radiograph of the joints was unremarkable. These findings fit well with a special form of CF-related arthritis. As in the case of classical CF arthritis, the treatment to be considered is, in particular, symptomatic administration of non-steroidal anti-inflammatory drugs and possibly glucocorticoids. Also under discussion as a further possibility is the use of antibiotics. Our patient has always refused medication. The condition again proved to be self-limiting.

In contrast to the classical form of CF arthritis, the special form is not associated with either joint swelling or local warmth. In the presence of arthritic symptoms in CF patients, consideration must always be given to a hypertrophic pulmonary osteoarthropathy. The latter, however, shows typical radiological changes and is exacerbated by lung infections.

The presence of arthritic pathologies in a patient with CF further underscores the fact that CF is a multiorgan morbid condition.

Keywords: Cystic fibrosis, arthritis, arthropathy, cutaneous manifestations.

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In patients suffering from cystic fibrosis (CF), two different articular pathologies have been described: so-called CF arthritis and hypertrophic pulmonary osteoarthropathy [1]. The incidence of each of these entities in cystic fibrosis patients is around 10% [1, 2]. We report herein on a 28-year-old man with CF who developed a special form of CF-related arthritis.

Case report

A 28-year-old man with cystic fibrosis (ΔF508 homozygote) complained of painful elbow joint symptoms associated with episodic limitation of movement during the previous three months. Neither joint swelling nor erythema was seen, but he developed clearly palpable cutaneous nodules over the joints affected by the attacks. It was not possible to establish any temporal relationship between the self-limiting symptormatology persisting for between 1 and 3 weeks, and pulmonary infections.

In addition to a CF-associated distal intestinal obstruction syndrome (DIOS) two years earlier, the patient has a 10-year history of schizophrenia. The patient's regular medication comprised pancreas enzymes, bronchodilators and neuroleptic agents. The patient smoked (10 py), and his attendance at the CF clinic for routine surveillance examinations was irregular. Physical examination revealed an underweight patient (166 cm, 44 kg) in a reduced state of health. In addition to clubbed fingers and toes, crackles were also heard over both lungs. Cardiac status and abdominal status were both unremarkable. Laboratory investigations showed no elevation of inflammatory parameters or immunological alterations. The chest radiograph revealed typical signs that had remained unchanged since the preceding radiograph. This meant that, despite a diminishment in the parameter forced one second capacity (FEV₁ 16% pred.; VC 27% pred.), his state of health was recorded as poor but stable. As reported by the patient, firm, non-mobile, non-tender nodules were present, bilaterally, in the region of the dorsum of the proximal forearm (fig. 1). Neither erythema nor swelling of the elbow joint was to be seen. Although active mobility was not restricted, the patient experienced pain on passive movement of the respective joints.

At ultrasonography, the nodular changes, measuring up to 25 x 6 mm, presented as echo-poor structures in direct contact with the periosteam. From the radiograph obtained in the region of the cutaneous manifestations (elbow joints; proximal part of the forearm), no pathological intra- or peri-
pathogenetic mechanism is the deposition of immune complexes in the synovia [5]. Furthermore, a triggering role of colonisation of with Pseudomonas aeruginosa deep in the lower airways, for example, has been considered [12]. Unfortunately, since no data on such colonisation in the patient has been provided in previous publications, a relevant comparison is not possible. Nor is the underlying cause of the nodular cutaneous manifestations clear. The prognosis of the disease is usually good.

Proposed treatment comprises the symptomatic administration of non-steroidal anti-inflammatory drugs and, where indicated, glucocorticoids [1]. Furthermore, there are also reports of successful treatment with intravenous antibiotics [6]. Admittedly, we currently have no prospective studies investigating the value of medication even when the untreated symptoms are of limited duration. In our own case, the response of the symptoms to drug treatment could not be investigated, since the patient always refused medication. During the further course of the disease, three more attacks of joint pain associated with nodular cutaneous manifestations occurred.

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Discussion

The clinical picture encompassing nodular cutaneous manifestations and painful joints, and
unremarkable radiographic findings, are readily compatible with a variant of CF-related arthritis [3].
The latter is differentiated from the classical form of CF-related arthritis in which swelling of the
joints and local warming also occur [1] and should therefore probably be termed arthropathy rather
than arthritis. The appearance of this type of arthritis during the course of cystic fibrosis is indepen-
dent of the severity of the pulmonary manifestations [1]. The literature also reports a number of
cases with considerably impaired primary function, as in our own patient [2]. A wide variety of
Cutaneous manifestations is found in up to 40% of
patients with CF-related arthropathies [2]. Such manifesta-
tions may be nodular changes, erythema or
maculopapillary exanthems, which, as in the case of
our own patient, are frequently to be found in
the immediate neighbourhood of the affected
joints [4].

Apart from these conspicuous nodules, our patient differs from most cases previously described
as he was relatively “old”, and showed no signs of
inflammation. All other symptoms and the course
of the disease were identical with those of the case
histories of CF-related arthritis patients in the
literature [1-11].

CF-related arthritis must be distinguished from
hypertrophic pulmonary osteoarthropathy (HPOA).
The latter, also commonly associated with CF and
joint pain, the articular and osseous symptoms typi-
cally increase appreciably in association with lung
infections [1]. In contrast, the differential diagnosis
of the two CF-related arthropathies is facilitated by
the presence in HPOA of radiological changes,
such as thickening of the periostem.

To date, the pathogenesis of CF arthritis has not been unequivocally clarified. A possible