From ankylosis to pencil-in-cup deformity in psoriatic arthritis: a case report

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ABSTRACT

A 56-year-old woman with psoriatic arthritis is presented whose initially ankylosed digit was later found to develop pencil-in-cup change. The patient was treated over that period with etanercept and had no signs of active arthritis. The possible mechanisms for these changes are discussed.

Introduction

Psoriatic arthritis (PsA) is a chronic systemic inflammatory disorder characterized by involvement of peripheral joints, axial joints and peri-articular structures as well as skin and nails. The multi-faceted character of the disease is reflected in the different radiographic features that are seen in patients with PsA (1). Radiographic imaging of PsA joints can reveal features of bone resorption in the form of large eccentric erosions and pencil-in-cup deformities along with features of bone formation such as periostitis and ankylosis (2). The mechanism for these radiographic changes is unclear. We describe radiographic progression from total joint ankylosis to pencil-in-cup deformity in a psoriatic arthritis patient.

Case report

A 56-year-old female suffering from PsA had been followed at the University of Toronto PsA clinic since September 1997. She presented for medical attention in 1995, with pain and swelling in several fingers and toes and the diagnosis was thought to be rheumatoid arthritis. She was treated but failed several non-steroidal anti-inflammatory medications and later sulfasalazine and intramuscular gold. However, at her first visit to our clinic she was noted to have an asymmetric arthritis with dactylitis, cervical and lumbar spine limitation of movement, as well as psoriasis in the umbilical area and the hair line. She was diagnosed with psoriatic arthritis on the basis of an inflammatory arthritis with psoriasis, and fulfilled CASPAR criteria by having psoriasis, dactylitis, and negative rheumatoid factor. Treatment with methotrexate was initiated in 1999 first orally and later by subcutaneous injections to a maximal dose of 25 mg per week. While on that medication she only achieved partial improvement of her symptoms. Etanercept was introduced on August 2002 by subcutaneous injections at a dose of 25 mg twice a week. At that time she had 2 actively inflamed joints, active tendonitis and chronic dactylitis in the 2nd right finger. She also had accumulated joint damage with 13 clinically damaged joints, and limited range of motion in her cervical and lumbar spine. Radiographs showed numerous erosive changes mainly in the fingers and toes including some joints with total destruction as well as ankylosis. Erosive changes were also noted in the spine with grade 3 sacroiliitis bilaterally. Inflammatory markers throughout all her disease course were within the normal limits. Within 6 months of initiation of etanercept therapy, she had marked improvement in her symptoms in both axial and peripheral joints and on physical examination no signs of active arthritis were noted. Plain radiographs of the hands from April 2004 showed ankylosis of the 2nd proximal interphalangeal (PIP) and 2nd distal interphalangeal (DIP) joints and large marginal erosions in the 2nd metacarophalangeal (MCP) joint (Fig. 1). The patient remained in remission without any significant change in physical examination findings until December 2005. At that visit a new flail joint was found on the right 2nd DIP joint. The patient denied any joint pain, and no signs of active inflammation were noted in that joint. On questioning she recalled minor trauma to that finger several weeks prior. Radiographs of the hand showed separation of the proximal and distal phalangeal margins in the right 2nd DIP joint, that were previously ankylosed. There were, however, bony bridges that connected the two bones. No interval changes were detected in the PIP and MCP joints (Fig. 2). The patient continued treatment with etanercept and remained in remission. The next set of radiographs, obtained in December 2006, showed total separation of the two phalanges with the appearance resembling a pencil-in-cup deformity. Again, no interval changes were noted in the PIP and MCP joints in that finger (Fig. 3). The patient has remained in remission through her

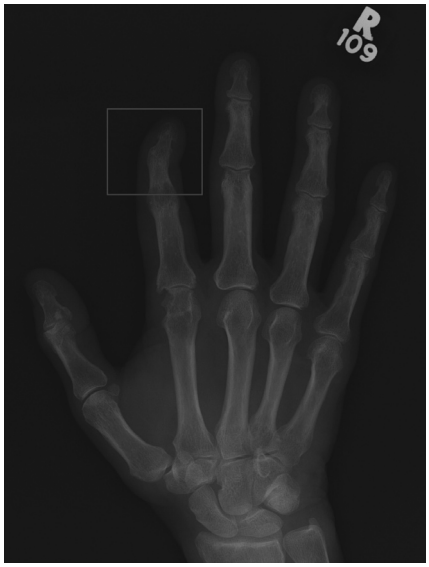

Fig. 1. April 2004 – Total ankylosis of DIP joint in the 2nd finger.

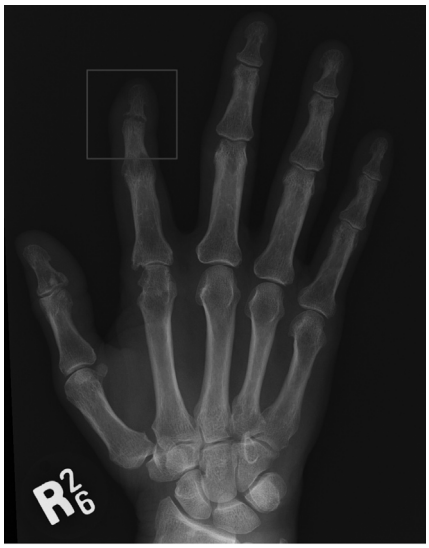

Fig. 2. December 2005 – New flail joint clinically – separation of the proximal and distal phalanges with few bony bridges left in the joint.

last visit in July 2008 and showed no additional radiographic changes in that joint (not shown).

Discussion

Typical radiographic changes among patients with PsA include erosions that may be large and lead to the typical pencil-in-cup deformity (2). Erosions are a common radiographic finding occurring in as many as in 47% of patients with early PsA and 67% of PsA patients at their first visit to the rheumatologic clinic (2-5). Another typical feature is new bone formation, which may

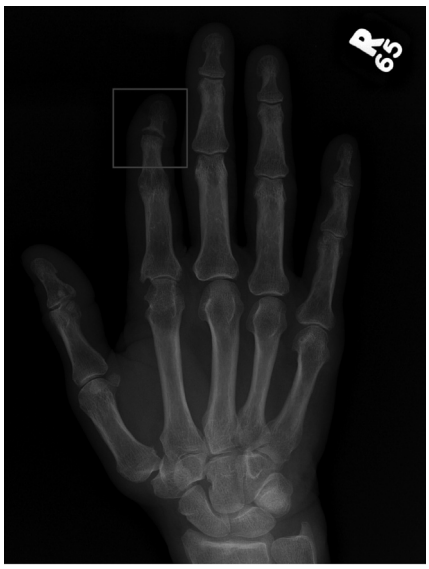

Fig. 3. December 2006 – Pencil-in-cup-like deformity of the DIP joint.

present as either ankylosis or periosteal reaction, which is important in differentiating PsA from other inflammatory arthritides. Fluffy periostitis is included in the CASPAR criteria for the classification of PsA (6). These two seemingly opposing mechanisms, marked erosion and ankylosis, can occur in the same patient and may present in different joints in the same finger (3).

The pathogenesis of these two mechanisms is not yet fully understood. The proinflammatory cytokine TNF-α has been shown to be central molecule in PsA. In the setting of high systemic levels of TNF-α, osteoclast precursors (OCP) are recruited into the joint via blood vessels in the inflamed synovium. These differentiate into active osteoclasts in the synovium, which than transforms into an aggressive and invasive pannus responsible for the erosive manifestations in PsA (7). TNF-α inhibition dramatically reduces OCP levels; this may provide an additional mechanism for the protective effect of TNF blockade on inflammatory bone loss in PsA (8). The mechanism that is responsible for new bone formation in PsA is much less understood. Transforming growth factor (TGF)-β and vascular endothelial growth factor (VEGF) may be pivotal in this process. TGF-β is strongly expressed in synovial tissues isolated from patients with ankylosing spondylitis and synergizes with VEGF to induce bone formation in animal models (9-10). Inflammation may also induce increased signaling in the bone morphogenic proteins pathway, as shown by increase in the downstream metabolite of this intracellular pathway in biopsies taken from the calcaneus in patients with Achilles tendonitis and periostitis (11).

The occurrence of osteolysis while the patient was on etanercept is surprising since the medication inhibits radiographic disease progression as measured by modified Sharp method that includes DIP joints (12). However, specific clinical features of PsA, such as periostitis and pencil-in-cup deformity were similar between treatment and placebo groups following 12 months of treatment (13). It was suggested that this was related to the short duration of follow-up in the study. However, another possible explanation may be the involvement of different cytokines in the pathogenesis of these deformities. Another interesting issue is the sub-clinical nature of the progression of bone resorption over a period of two years. The patient was assessed at regular intervals without detection of any clinical evidence of active inflammation in the affected joint. Buskila *et al*. noted that inflamed joints in patients with PsA were less tender than in rheumatoid arthritis (13). It was suggested that the severity of articular inflammation may be underestimated in PsA patients.

In the case reported here, the patient reported minor trauma that occurred prior to the appearance of the clinical and radiographic changes of joint loosening. While fracture was considered as a possible explanation, the appearance of minor bony bridges (Fig. 2) and the progression to pencil-in-cup like deformity in the late image led us to believe that osteolysis played a role in these changes. Furthermore, local trauma may have played a different role in this case. Trauma may be a trigger for joint inflammation, especially in PsA, where it is thought to result from deep Koebner (14). Up to 24.6% of the patients with PsA reported a traumatic event prior to the diagnosis of PsA (15). In another study, 8% of the patients with PsA compared to 2% of

the patients with trauma in the 3 month before arthritis was noted (16).

In summary we present a patient with PsA with a radiographic progression from ankylosis to pencil-in-cup deformity in a single joint over a period of 2 years while the patient was on an anti-TNF agent. To our knowledge this is the first case reported in the literature. It emphasizes the complicated role that the two opposing mechanisms of bone resorption and bone formation play in the pathogenesis of PsA.

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