Rheumatoid Arthritis-Associated Swan Neck Deformity Improved with Adalimumab Therapy

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Rheumatoid arthritis (RA) primarily affects the synovial joints; it is a systemic autoimmune inflammatory disorder. Swan neck deformity (SND) is an articular manifestation of RA; its association with seropositive RA is well documented.

We present a thirty-nine-year-old Bahraini male with seropositive RA-associated SND. At the time of diagnosis in 2015, the patient presented with morning stiffness, 3rd and 4th metacarpophalangeal (MCP) joints pain, tenderness of the left hand and bilateral 3rd and 4th proximal interphalangeal (PIP) joint pain and swelling for two months. Laboratory investigations revealed positive rheumatoid factor (RF) and C-reactive protein (CRP) only. The patient was diagnosed as seropositive RA. The patient was treated with weekly 12.5 mg methotrexate (MTX) injection and folic acid 5 mg. The patient was lost for follow-up for two years and returned with hand deformity, prominent SND of 3rd and 4th fingers bilaterally. Laboratory investigations revealed: antinuclear antibody (ANA) (1:160), RF (64 IU), anti-cyclic citrullinated peptide (anti-CCP) (356) and CRP (13). The patient was started on Humira (Adalimumab) injection 40 mg SC every two weeks plus MTX 10 mg weekly. Humira therapy dramatically improved laboratory and clinical signs of SND.

In RA-associated SND, early detection and initiation of Humira biological therapy are essential in stopping the progression of the disease.

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Rheumatoid arthritis (RA) is a systematic autoimmune inflammatory disease characterized by progressive and destructive polyarthritis; the bone and cartilage of joints are often destroyed and the tendons and ligaments are weakened. The onset of RA disease is usually from 35-60 years with a female predominance; the female to male ratio is 3:1[1,2]. Besides clinical features, laboratory investigations are essential for the diagnosis of RA. Investigations include C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), rheumatoid factor (RF) and anti-cyclic citrullinated peptide (anti-CCP). The European League against Rheumatism (EULAR) recommends conventional radiography, ultrasound or MRI to confirm the diagnosis of RA[3].

There are several rheumatoid arthritis-related complications, articular (joints) and extra-articular (eye, skin, blood, lung, heart, etc.). One of the most common articular complications is hand deformity. The majority of patients with RA develop metacarpophalangeal joints (MCP) pathology and tenosynovitis[4]. SND is one of the most common hand deformities which leads to functional disability in patients[5]. SND is the hyperextension of the proximal interphalangeal joint (PIP) with hyperflexion of distal interphalangeal joint (DIP). The cause of SND includes intrinsic or extrinsic mechanisms or a combination of both. In RA, the intrinsic mechanism plays the main role for SND; irritation from inflammation leads to reflex spasm of adjacent MCP joints. Extrinsic causes with wrist flexion contracture and primary distal interphalangeal joint disease are sometimes present in RA[6]. Hand deformity is more common in a seropositive patient, resulting in pain and functional disability[7].

The early phase of RA is important because the inflammation is at its peak. The rate of erosion, number of inflamed joints and rate of bone loss are also at their peak during this time. The patient is bound to deteriorate if left untreated during the early phase of RA; suppressive treatment is of great benefit at this time[8]. Non-steroidal anti-inflammatory (NSAIDs), corticosteroids, and disease-modifying antirheumatic drugs (DMARDs) are used in the treatment of RA. DMARDs have the capacity to reduce the activity and improve the radiographic outcomes in RA patients[8]. DMARDs was subdivided into two classes: non-biologic/traditional, such as methotrexate and biologic, such as anti-TNF[4].

The American College of Rheumatology (ACR) recommends the use of biologic DMARDs early in the disease combined with methotrexate in a patient with active RA. Health-related quality of life improved with adalimumab[9-11]. A study revealed that treatment with ADA+MTX was significantly superior to

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methotrexate alone[12]. Surgery is necessary if patients develop severe deformities with no passive range of movement of the joint[13].

The aim of this presentation is to report a case of an improved swan neck deformity after the use of biologic DMARDs in RA.

THE CASE

A thirty-nine-year-old Bahraini male presented with a history of morning stiffness, 3rd and 4th metacarpophalangeal (MCP) joints pain and tenderness of the left hand, bilateral 3rd and 4th proximal interphalangeal (PIP) joint pain and mild swelling for three months duration. He reported right shoulder and knee pain for one year. There was a family history of rheumatoid arthritis (his mother). His mother had severe disease with hand deformities.

Musculoskeletal examination revealed severe tenderness and mild swellings of PIP of middle and ring fingers bilaterally, more prominent on the left hand with a restricted range of movement of both hands. Investigations revealed normal TWBC, high CRP (7), positive RF (64 IU), severely low vitamin D (13 nmol/l) and normal serum calcium. Other investigations were normal, including anti-CCP, ANA, ENA, ds-DNA, C3, C4, LFT, RFT and chest X-ray. The patient refused MRI of his hands due to machine phobia. The patient was diagnosed with RA according to the 2010 ACR/EULAR RA classification criteria, which stipulate that the patient should have ≥6 points out of possible 10 as diagnostic of RA[14]. Our patient had 7 points: joint involvement 3 points, serology 2 points, acute-phase reactants 1 point and duration of symptoms 1 point. The patient was categorized as low disease activity as his Disease Activity Score-28 for Rheumatoid Arthritis (DAS28) was 3.17[15]. The treatment was initiated with intramuscular (IM) methotrexate (MTX) injection 15 mg weekly and oral folic acid 5 mg weekly plus a single IM injection of vitamin D3 of 600.000 IU. The patient refused to take any oral medications of MTX or hydroxychloroquine (HCQ) and preferred to take injections. The patient had significant improvement of clinical symptoms and laboratory findings in 6 months. Disease activity was estimated as DAS28 remission, his DAS28 was 2.46 (DAS28 was <2.6)[15]. During the follow-up period of 8 months the patient was stable (DAS28 was 1.21).

The patient was lost to follow-up for two years, after which, he presented with deformity of his index, middle and ring fingers on both hands, see figure 1. He also reported a frequent absence from work due to generalized fatigue and joints pain.

Figure 1: Right Hand before Adalimumab Therapy

Physical examination revealed SN appearance on the 2nd, 3rd and 4th fingers bilaterally. The lab result revealed positive ANA, high anti-CCP (356), high RF (70), normal LFT, normal chest X-ray, normal ds-DNA, C3 and C4 and negative ENAs. MRI was refused and ultrasound (US) was performed, see figures 2 and 3. The left-hand US revealed a ganglion cyst arising from the right dorsal scapholunate joint, it measured 1.3 x 0.4 x 1.5 cm. A focus of ossification within the first MCP was also detected. No active synovitis or tenosynovitis was seen.

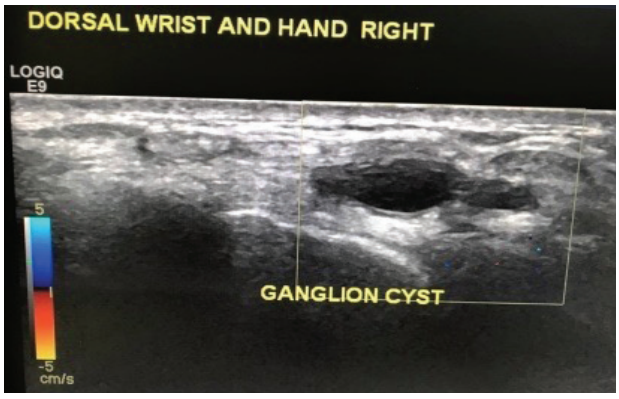

Figure 2: Ultrasound Showing Ganglion Cyst on the Dorsum of the Right Wrist

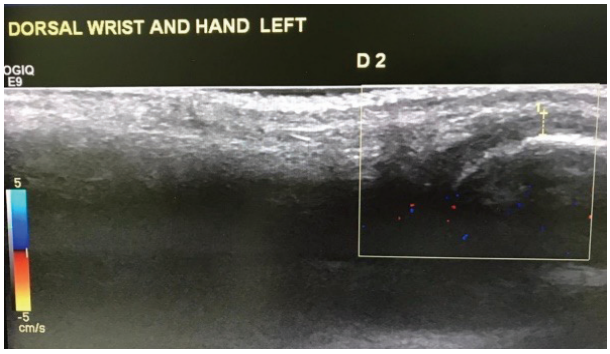

Figure 3: Ultrasound of the Dorsum of the Left Wrist

The patient was started on Humira injection 40 mg SC every two weeks and MTX 10 mg every week. His articular symptoms (pain and swellings) and laboratory inflammatory findings resolved immediately. Gradually his SND improved and eventually resolved, see figure 4. MTX was stopped in June 2018 and the patient was compliant with his therapy since then.

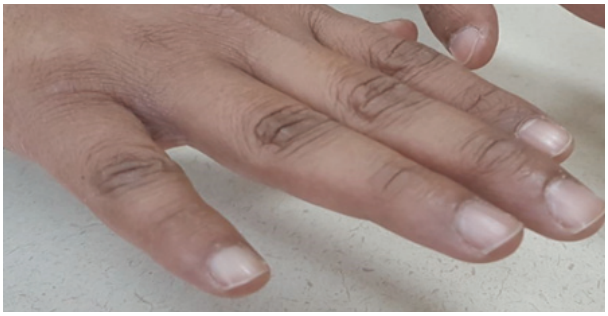

Figure 4: Right Hand after Adalimumab Therapy

In October 2018, MRI of the right hand revealed asymmetry within the first MCP joint with chondral loss and tiny cystic/

erosive changes at the base of the proximal phalanx, as well as, the lateral aspect of the first MCP head, see figures 5 and 6. There was chondral loss involving the central aspect of the fourth MCP head with the irregularity of the base of the proximal phalanx. No significant marrow edema. No change in the size of the ganglion cyst seen previously by US and no active synovitis or tenosynovitis.

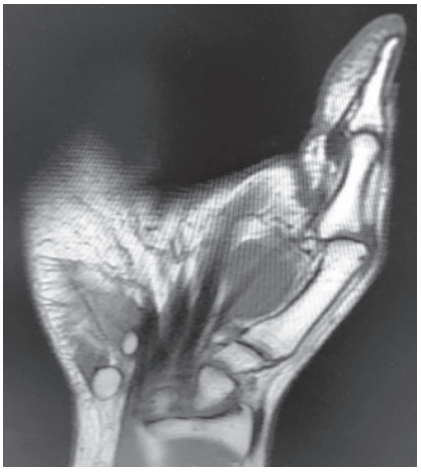

Figure 5: MRI of the Right Hand Showing Asymmetry within the First MCP Joint with Chondral Loss and Tiny Cystic/Erosive Changes at the Base of the Proximal Phalanx, as Well as the Lateral Aspect of the First MCP Head and Ganglion Cyst

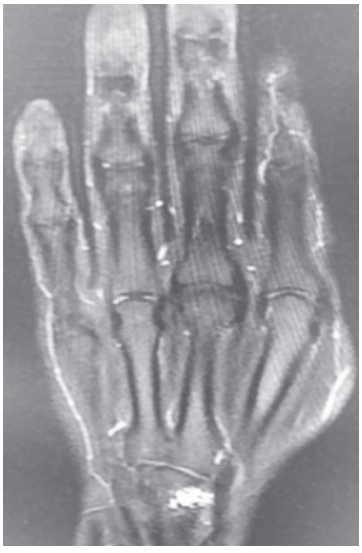

Figure 6: MRI of the Right Hand Showing Ganglion Cyst at the Wrist

DISCUSSION

SND is one of the common manifestations of RA if not treated early. SND could lead to significant impairment of function; therefore, an orthopedic surgeon may be involved to treat the patient surgically[16]. Early treatment with biological DMARDs such as adalimumab (Humira) can prevent the deformity[17].

Our patient was diagnosed early as seropositive RA with active disease and started with the methotrexate; however, he was non-compliant with his medications and the regular follow-up resulting in the delayed proper selection of further treatment such as the biological agents to improve the outcome of the rheumatoid arthritis[18]. Keystone et al found that a 52-week delay of combining adalimumab to MTX led to worse radiographic, functional, and clinical outcomes[11]. Our patient was diagnosed early and was treated with MTX only since the disease was mild and he had low disease activity and both ANA and anti-CCP were negatives. The rapid achievement of clinical response in our patient in the first weeks of therapy with MTX should have been associated with improved long-term outcomes. The initial negative anti-CCP and low positivity of rheumatoid factors tend to be good prognostics and not usually associated with underlying erosion and hand deformity. However, the loss of follow-up and the cessation of the medications were the main factors that led to worse clinical outcomes in our patient.

The clinical improvement in our patient during the first weeks could be because Adalimumab was introduced in less than four years of the disease duration, which is relatively early.

A non-compliant patient is hard to deal with regarding management and outcome. This underlines the importance of clinical assessment, particularly following cessation and then re-continuance of the medication.

CONCLUSION

It is important to educate the patient about the consequences of cessation of the therapy in RA and the positive outcome of early treatment with biologic DMARDs. Any patient with noticeable prognostic factors should be started immediately on biological DMARDs to achieve excellent treatment outcome.

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