

## THERAPEUTIC CHALLENGES AND COMPLICATIONS IN A CASE OF ARTHRITIS MUTILANS

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### Abstract

The most dramatic and severe form of arthritis associated with psoriasis is arthritis mutilans (AM), which is a rare disorder, affecting very few patients with psoriasis. AM has a predilection for the small joints of the hands and feet. It is generally characterized by seronegative degenerative joint disease, leading to osteolytic changes in the carpal and digital bones. The bone and joint lesions rapidly and progressively cause bone lysis and joint ankylosis with loss of digits, soft-tissue deformities, telescoping of fingers and toes and the hallmark "la main en lorgnette" deformity (opera-glass hand). (1)

Arthritis mutilans is characterized by an asymmetric pattern of peripheral joint involvement, with a predilection for the interphalangeal and metacarpophalangeal joints of the hand and small joints of the feet. (2) Characteristic features of AM are severe deformity of the hands, foreshortened fingers with excessive skin folds, hypermobile joints and digits that can be elongated by traction. (3) Radiologically, AM is characterized by severe resorption of the joint with an attendant loss of function, sometimes to a dramatic degree. (4)

We present the case of a male patient who with psoriatic lesions onset from almost four decades, followed by involvement of hand and feet joint with important deformities. The treatment was difficult due to important comorbidities.

**Keywords:** arthritis mutilans, psoriatic arthropathy, anti TNF- $\alpha$  therapy

### INTRODUCTION

Psoriatic arthritis mutilans is the most severe form of the five clinical presentations of psoriatic arthritis (PsA) described by Moll and Wright (5). The clinical manifestation of the condition is shortening of a digit due to gross osteolysis, resulting in the so-called "opera glass finger", "telescopic finger", or "doigt en lorgnette". Previously published studies on this condition have used different definitions of psoriatic arthritis mutilans, as no international classification criteria are yet to be found in the literature. However, radiographic features have been found to be more sensitive than clinical findings (6). Recently, attempts have been made on the initiative of the Group for Research and Assessment of Psoriasis and Psoriatic Arthritis (GRAPPA) in order to develop a

consensus definition of psoriatic arthritis mutilans, but these have not yet been globally accepted (8).

The reported prevalence of psoriatic arthritis mutilans varies considerably, in the range of 1-21% of patients with PsA (9). In the Nordic countries, the prevalence was found to be only about four cases per 1,000,000 inhabitants (3.7; 95% CI 2.8-4.6) in a multinational population-based study recently published by the Nordic Psoriatic arthritis mutilans Study Group (10).

Radiographic evaluation is helpful and conventional radiographs are the most commonly used radiological tool. The radiographic features of PsA are well known, including joint destruction, inter-joint space narrowing, bony proliferation, periostitis, osteolysis, including pencil-in-cup deformity, and ankylosis. Any or all of these radiological changes may

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be present in one and the same patient. Involvement of the distal interphalangeal joints of the fingers is typical (11). Osteolysis is the defining feature of arthritis mutilans and there is consensus (according to GRAPPA) that involvement of a single joint is sufficient to establish the diagnosis of Psoriatic arthritis mutilans. (12,13)

## CASE REPORT

P.C., male, 57 years of age, ex smoker, ex alcohol consumer, who used to work with vinyl chloride and ammonium, living in the countryside, comes to our hospital accusing inflammatory pain of all small joints of the hands and feet, elbows, swelling of the 3rd IP joint of his right foot and mechanical pain in the lower back.

The patient describes psoriatic lesions on elbows and knees since 1977, treated topically. In 1995 he was diagnosed with erythrodermic psoriasis and treated with PUVA and retinoids. He had a weak therapeutic response and hepatic cytolysis due to medication.

In 1998 he was also diagnosed with “grand mal” epilepsy, treated with Carbamazepine. In 2001 he presented with knee, ankle, MTP and then PIP and DIP joints arthritis. He was diagnosed with psoriatic arthritis and began Methotrexate 10 mg/week, stopped after 5 months due again to increased level of hepatic enzymes. He was switched to Sulfasalazine 2 g/day, with minor response, than to Cyclosporine for 3 months, with no effect.

In 2008, he was diagnosed with liver cirrhosis (chronic alcohol consumer, multiple drug toxicity, AgHBs positivity). He began treatment with Entecavir for 6 months, with HBV-DNA negative after treatment. During this period, he received no treatment for psoriatic arthritis, so when he came back for rheumatologic evaluation he had a swollen joint count (SJC) of 7 and a tender joint count (TJC) of 22, BSA>10%, PASI>10, high inflammatory syndrome (ESR=61 mm/h, CRP=21.29 mg/l) and also anti-CCP antibodies of 150.87 u/ml and rheumatoid factor of 128.5UI/ml. Clinical examination revealed impressive deformities of hands and feet (arthritis mutilans). (Figure 1).

With these clinical and biological scene, he was considered a candidate for biological therapy, but the Quantiferon Gold TB test was positive, so he received Isoniazid with strict liver enzymes follow-up every 2 weeks and had approval from the pneumolo-



**FIGURE 1.** Important deformities of hands

gist for the biologic therapy. He continued to have undetectable level of HBV-DNA so he got approval for biologic therapy also from gastroenterologist and was started on Infliximab. He got a very good response for 2 years.

In 2013 he had 3 episodes of atrial fibrillation; the first two were converted and he received only clopidogrel, for prevention. He did not receive anticoagulation because of the hepatic involvement and the difficulties in monitoring the INR (living in the countryside). In august 2014 the third episode of atrial fibrillation caused a stroke which lead to hemiplegia. The therapy with Infliximab was interrupted for more than 8 months due to admission in neurologic clinic for rehabilitation and the impossibility to come for reevaluation.



**FIGURE 2.** Extensive psoriatic lesions



**FIGURE 3.** Severe deformities of the feet

In May 2015 he managed to be reevaluated in our department. He was immobilized, completely dependent of a care-taker, presenting extensive psoriatic lesions and painful swelling and deformity of his knees, ankles, PIP and DIP joints, with biological inflammatory syndrome (ESR=59 mm/h, CRP=15.46 mg/dl) and anti-CCP antibodies=273.6 U/ml, with a normal RF and normal hepatic and renal markers. Considering all this, in July this year the therapy was switched to Adalimumab. After 3 administration of

the new biologic the result was spectacular, with a very good cutaneous, as well as joint response (Figure 4).



**FIGURE 4.** Disappearance of psoriatic lesions after taking Infliximab

## CONCLUSIONS

We present the case of a patient with severe rheumatologic involvement and with multiple problems raised by important comorbidities. The biologic treatment managed to control the articular and cutaneous disease despite all the collateral problems. The patient followed two TNF- $\alpha$  blockers, with an actual good response to Adalimumab.

## REFERENCES

1. Moll J.M.H, Wright V. Psoriatic arthritis. *Semin Arthritis Rheum* 1973; 3:55-78.
2. Roberts M.E., Wright V., Hill A.G., Mehra A.C. Psoriatic arthritis. *Ann Rheum Dis* 1976; 35:206-12.
3. Gladman D.D., Brockbank J. Psoriatic arthritis. *Opin Investig Drugs* 2000; 9: 1511-22.
4. Belt E.A., Kaarela K., Kauppi M.J., et al. Assessment of mutilant-like hand deformities in chronic inflammatory joint disease. A radiographic study of 52 patients. *Ann Rheum Dis* 1999; 58:250-2.
5. Larsen A., Dale K., Eek M. Radiographic evaluation of rheumatoid arthritis and related conditions by standard reference films. 1977; 18:481-91.
6. Jones G., Grotty M., Brooks P. Psoriatic arthritis: a quantitative overview of therapeutic options. The psoriatic Arthritis Meta-Analysis Study Group. *Br J Rheumatol* 1997; 36:95-
7. Robert Monger M.D. and Virginia Hrywnak, D.O., *N Engl J Med* 2003; 349:1935
8. Psoriatic Arthritis Mutilans: Clinical and Radiographic Criteria. A Systematic Review. *J Rheumatol.* 2015 Aug; 42(8):1432-8.
9. Gudbjornsson B., Ejstrup L., Gran J.L., et al. Psoriatic arthritis mutilans (PAM) in the Nordic countries: Demographics and disease status. The Nordic PAM study. *Scand J Rheumatol* 2013; 42: 373-378.
10. Tillet W., Jadon D., Shaddic D., et al. Feasibility, reliability and sensitivity to change of four radiographic scoring methods in patients with psoriatic arthritis. *Arthritis Care Resch* 2014; 66: 311-317.
11. Koo T., Nagy Z., Sesztak M., et al. Subsets in psoriatic arthritis by cluster analysis. *Clin Rheumatol* 2001; 20: 36-43.
12. Marsal S., Armadans-Gil L., Martínez M., et al. Clinical, radiographic and HLA associations as markers for different patterns of psoriatic arthritis. *Rheumatology (Oxford)* 1999; 38: 332-337.
13. Bruce I.N., Ho P.Y.P. Clinical features of psoriatic arthritis in *Rheumatology* vol. 2, 6<sup>th</sup> ed, Mosby Elsevier Ltd 2015 pp. 989-996