UNIVERSITY OF LEEDS

This is a repository copy of *Arthritis mutilans in systemic sclerosis*.

White Rose Research Online URL for this paper:
http://eprints.whiterose.ac.uk/144920/

Version: Accepted Version

Article:
Abignano, G, Mennillo, GA, Lettieri, G et al. (3 more authors) (2019) Arthritis mutilans in systemic sclerosis. Arthritis and Rheumatology, 71 (1). p. 120. ISSN 2326-5191

https://doi.org/10.1002/art.40723

(c) 2018, American College of Rheumatology. This is the peer reviewed version of the following article: 'Abignano, G, Mennillo, GA, Lettieri, G et al (2019). Arthritis mutilans in systemic sclerosis. Arthritis and Rheumatology, 71 (1). p. 120,' which has been published in final form at [https://doi.org/10.1002/art.40723]. This article may be used for non-commercial purposes in accordance with Wiley Terms and Conditions for Self-Archiving.

Reuse
Items deposited in White Rose Research Online are protected by copyright, with all rights reserved unless indicated otherwise. They may be downloaded and/or printed for private study, or other acts as permitted by national copyright laws. The publisher or other rights holders may allow further reproduction and re-use of the full text version. This is indicated by the licence information on the White Rose Research Online record for the item.

Takedown
If you consider content in White Rose Research Online to be in breach of UK law, please notify us by emailing eprints@whiterose.ac.uk including the URL of the record and the reason for the withdrawal request.

White Rose university consortium
Universities of Leeds, Sheffield & York

eprints@whiterose.ac.uk
https://eprints.whiterose.ac.uk/

Title: Arthritis Mutilans in Systemic Sclerosis

Subtitle: Arthritis Mutilans in SSc

Authors: Giuseppina Abignano MD PhD [1-3], Gianna A. Mennillo MD [1], Giovanni Lettieri MD [4], Angela Padula MD [1], Dennis McGonagle FRCPI PhD [2,3], Salvatore D'Angelo MD PhD [1,5]

Affiliations:

1. Rheumatology Institute of Lucania (IReL), Rheumatology Department of Lucania, San Carlo Hospital of Potenza and Madonna delle Grazie Hospital of Matera, Potenza, Italy
2. Leeds Institute of Rheumatic and Musculoskeletal Medicine, University of Leeds, United Kingdom
3. NIHR Leeds Biomedical Research Centre, Leeds Teaching Hospitals NHS Trust, Leeds, United Kingdom
4. Department of Radiology, San Giovanni di Dio e Ruggi d'Aragona University Hospital, Salerno, Italy
5. Basilicata Ricerca Biomedica (BRB) Foundation, Potenza, Italy

Corresponding Author:

Dr. Giuseppina Abignano

Rheumatology Institute of Lucania (IReL), Rheumatology Department of Lucania, "San Carlo" Hospital, Via Potito Petrone snc, 85100 Potenza, Italy;

email address: g.abignano@hotmail.com; Tel +39 09 7161 3131, Fax +39 09 7161 3036;

Word Count: 297; **Figure**: 2

Funding: No specific funding was received for the reported work.

Competing interests: The authors have no competing interests to declare.

A 40-year-old gentleman was diagnosed with anti-topoisomerase-I antibody positive diffuse cutaneous systemic sclerosis (SSc). Raynaud's phenomenon, diffuse skin sclerosis and severe interstitial lung disease (ILD) were the onset features causing digital ischemia, disability and dyspnea. He subsequently developed inflammatory polyarthritis (IA) with hands and wrists involvement. His brother had psoriasis. Physical examination showed fingertip pitting scars, subcutaneous calcinosis, skin sclerosis with hyper- and hypopigmentation, flexion and extension contractures and shortened fingers (Figure 1.A). Hand/wrist X-ray revealed bilateral destructive arthropathy, acro-osteolysis, calcified deposits consistent with joint and skin calcinosis (Figure 1.B). Joint destruction with diffuse bone loss (left 2^{nd}, 5^{th} and right 5^{th} PIP joints), multiple subluxations (left 1^{st} and 2^{nd}, right 1^{st}, 2^{nd}, 3^{rd} MCP joints), whittling (left 1^{st} MCP joint), pencil-in-cup-like deformity (left 2^{nd} PIP joint and left 5^{th} MCP joint), that are most typical of psoriatic arthritis mutilans (1, 2), were also evident (Figure 1.B).

A previous EUSTAR cross-sectional analysis revealed frequent articular involvement in SSc (3) however severely destructive IA causing finger shortening is not common and should be distinguished by the typical acro-osteolysis secondary to ischemia in order to guide treatment. Since SSc diagnosis, therapy included nifedipine, i.v. prostanoids and bosentan to treat digital ischemia and prevent ulceration and several immunosuppressive drugs to target both the skin involvement and ILD (low dose steroids, cyclophosphamide, azathioprine, mycophenolate). Methotrexate failed to control rapidly progressive articular damage. The patient is currently undergoing rituximab therapy in the attempt to target the SSc-related skin involvement and severe ILD, and to control joint symptoms. An earlier diagnosis of PsA mutilans could have changed the treatment escalation in favor of cytokine inhibition. However, since therapies that reliably target both SSc and PsA are not currently available, treatment choice needs to be driven by the severity and presence of life-threatening manifestations.

References:

1. McGonagle D, Conaghan PG, Emery P. Psoriatic arthritis: a unified concept twenty years on. Arthritis Rheum 1999;42:1080-6.
2. Haddad A, Johnson SR, Somaily M, Fazelzad R, Kron AT, Chau C, et al. Psoriatic Arthritis Mutilans: Clinical and Radiographic Criteria. A Systematic Review. J Rheumatol 2015;42:1432-8.
3. Avouac J, Walker U, Tyndall A, Kahan A, Matucci-Cerinic M, Allanore Y, et al. Characteristics of joint involvement and relationships with systemic inflammation in systemic sclerosis: results from the EULAR Scleroderma Trial and Research Group (EUSTAR) database. J Rheumatol 2010;37:1488–501.