MANCHESTER 1824
The University of Manchester

The University of Manchester Research

Severe Disability in a Patient With Rheumatoid Arthritis and Sickle Cell Anemia An Underreported, But Yet a Potentially Treatable Combination of Diseases

Link to publication record in Manchester Research Explorer

Citation for published version (APA):
Leung, M. H. K., Hughes, M., Lane, J., Basu, S., Ryan, K., & Jones, A. (2015). Severe Disability in a Patient With Rheumatoid Arthritis and Sickle Cell Anemia An Underreported, But Yet a Potentially Treatable Combination of Diseases. *Journal of Clinical Rheumatology*, *21*(8), 458-459.

Published in:
Journal of Clinical Rheumatology

Citing this paper
Please note that where the full-text provided on Manchester Research Explorer is the Author Accepted Manuscript or Proof version this may differ from the final Published version. If citing, it is advised that you check and use the publisher's definitive version.

General rights
Copyright and moral rights for the publications made accessible in the Research Explorer are retained by the authors and/or other copyright owners and it is a condition of accessing publications that users recognise and abide by the legal requirements associated with these rights.

Takedown policy
If you believe that this document breaches copyright please refer to the University of Manchester's Takedown Procedures [http://man.ac.uk/04Y6Bo] or contact uml.scholarlycommunications@manchester.ac.uk providing relevant details, so we can investigate your claim.

OPEN ACCESS

Download date:29. Nov. 2023

Severe Disability in a Patient With Rheumatoid Arthritis and Sickle Cell Anemia

An Underreported, But Yet a Potentially Treatable Combination of Diseases

To the Editor:

The coexistence of rheumatoid arthritis (RA) and sickle cell anemia (SCA) has been little reported in the literature.[1–4] We describe such a case and discuss the importance of recognizing the coexisting diseases, the role of musculoskeletal (MSK) imaging, and a coordinated multidisciplinary team (MDT) approach to treatment.

A 20-year-old Nigerian woman with known SCA was admitted with acute polyarticular pain and florid synovitis of the small (in particular the hands) and large joints. On questioning, she disclosed several years' history of inflammatory joint symptoms. More longstanding, she was unable to weight bear without severe pain (in particular of her knees), with a progressive inability to walk and reliance on a wheelchair.

Her RA responded very well to disease-modifying antirheumatic drug therapy (currently methotrexate) and has been exquisitely corticosteroid (oral and intramuscular) responsive, with no precipitation of a painful vaso-occlusive episode. Her upper-limb function has been preserved, allowing her to maintain many of her activities of daily living and quality of life. However, despite intensive MDT input, she remains significantly functionally impaired in the lower limbs, with extensive joint destruction in keeping with SCA, resulting in almost no flexion at the knees. She is currently awaiting bilateral total knee replacements.

We are confident of the coexistence of both SCA and RA in our patient. Sickle cell anemia was confirmed on genetic testing (Hb homozygous SS); although she denied previous "typical" sickle crisis episodes, she did disclose a history of recurrent severe noninflammatory (in particular large) joint pain and was notably short in stature. She had a severe microcytic anemia (hemoglobin = 3.6 g/dL; mean corpuscular volume, 68 fL) requiring urgent transfusion. Synovial fluid (from her knee) was noninflammatory (1+ leucocytes only), and magnetic resonance imaging scan of the knees (Fig.) revealed extensive joint destruction from established SCA-related bone infarction and osteonecrosis. However, rheumatoid factor (385 IU/mL [0–20 IU/mL]), erythrocyte sedimentation rate (128 mm/h

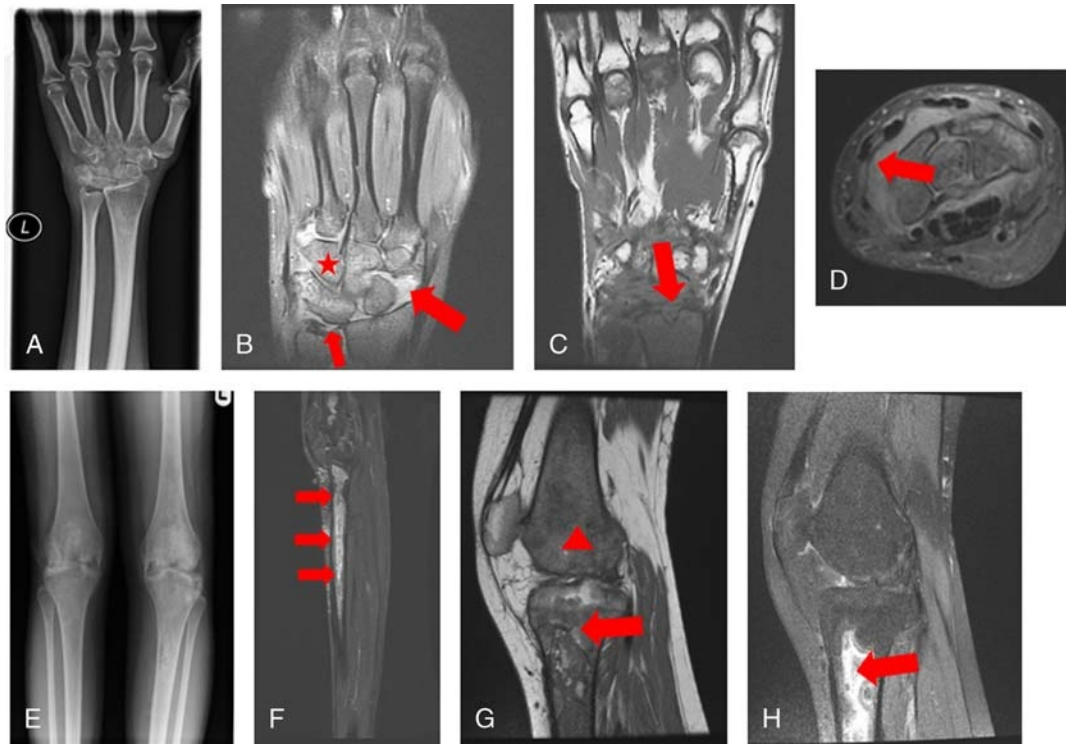

FIGURE. A–D, Hand. A, Radiograph of the left hand with loss of the joint space at the radiocarpal articulation. B, Magnetic resonance imaging coronal fluid-sensitive (short tau inversion recovery [STIR]) image of the left hand that demonstrates marked effusion and synovitis involving the radiocarpal joint (arrow); there is reactive bone marrow edema involving the carpal bones (star), and there is an incidental finding of a triangular fibrocartilage complex tear (small arrow). C, Magnetic resonance imaging T1 image of the left hand that demonstrates a focal erosion involving the distal radius (this was not visualized on the radiograph). D, Magnetic resonance imaging axial fluid-sensitive (STIR) image demonstrating marked effusion and synovitis within the wrist. E–H, Knees. E, Radiograph of both knees, weight-bearing views, demonstrates severe loss of joint space involving the tibofemoral compartments; in addition, in the left proximal tibular shaft, there is sclerosis (arrow). F, Magnetic resonance imaging sagittal fluid image through the left tibia demonstrates extensive serpingeous signal change through the shaft in keeping with bone infarction (multiple arrows). G and H, Sagittal T1 (G) and Sagittal fluid-sensitive (STIR) images (H) demonstrating extensive infarction (arrows) involving the proximal tibia and background degenerative change at the knee joint, with loss of normal marrow signal of the distal femur (triangle) (G).

Copyright © 2015 Wolters Kluwer Health, Inc. All rights reserved.

[0–9]), and CRP (20 mg/L [<10 mg/L]) were elevated; magnetic resonance imaging scan of the wrist (Fig.) confirmed marked synovitis with established erosive disease, and her inflammatory joint disease has responded to immunosuppressive treatment as above, all consistent with RA. In addition, she fulfilled the 2010 American College of Rheumatology/European League Against Rheumatism RA classification criteria.[5]

There are few case reports of coexistent RA (and juvenile idiopathic arthritis) in patients with SCA.[1–4] Common to these is the marked delay in the diagnosis of an inflammatory arthritis, with a severe destructive (and often erosive) arthropathy and associated disability.[1,3,4] The delay in diagnosis may result from both the commonness of MSK symptoms and a lack of awareness of possible RA in patients with SCA.[3,4] The local phagocytosis of sickled erythrocytes has been implicated in the pathogenesis of both SCA-related inflammatory disease including joint effusions.[6,7] Diagnostic imaging (eg, magnetic resonance) may help to distinguish inflammatory joint disease from SCA MSK complications where diagnostic uncertainty exists and to delineate the contribution of the 2 diseases.[3] Of caution, corticosteroid therapy (by any route, including intraarticular) may precipitate a potentially life-threatening acute vaso-occlusive episode.[1,3,8]

In conclusion, our patient highlights the possibility of RA coexisting in patients with SCA, including the potential diagnostic role for MSK imaging. Although she remains significantly functionally impaired in the lower limbs, her upper limb function and quality of life have been maintained by treating her RA with immunosuppressive therapy and by an MDT approach to treatment.

Matthew Hong Kay Leung, MRes
School of Medicine
The University of Manchester
Manchester
United Kingdom

Michael Hughes, MSc, MBBS, BSc(Hons), MRCP
Centre for Musculoskeletal Research
Salford Royal NHS Foundation Trust
The University of Manchester
Manchester Academic Health Science Centre
Manchester
United Kingdom
Michael.Hughes-6@postgrad.manchester.ac.uk

Jayde Lane, BSc(Hons)
Department of Rheumatology
Salford Royal NHS Foundation Trust
Salford
United Kingdom

Subhasis Basu, MBBS, BSc(Hons), FRCR
Department of Musculoskeletal Radiology
Salford Royal NHS Foundation Trust
Salford
United Kingdom

Kate Ryan, MD, FRCP, FRCPath
Department of Haematology
Manchester Royal Infirmary
Central Manchester University Hospitals NHS
Foundation Trust
Manchester
United Kingdom

Anthony Kenneth Peter Jones, MD, FRCP
Human Pain Research Group
Institute of Brain, Behaviour and Mental Health
University of Manchester
Manchester
United Kingdom

The authors declare no conflict of interest.

REFERENCES

1. Gladman DD, Bombardier C. Sickle cell crisis following intraarticular steroid therapy for rheumatoid arthritis. *Arthritis Rheum*. 1987;30:1065–1068.
2. Marino C, McDonald E. Rheumatoid arthritis in a patient with sickle cell disease. *J Rheuumatol*. 1990;17:970–972.
3. Nistala K, Murray KJ. Co-existent sickle cell disease and juvenile rheumatoid arthritis. Two cases with delayed diagnosis and severe destructive arthropathy. *J Rheumatol*. 2001;28:2125–2128.
4. Adelowo O, Edunjobi AS. Juvenile idiopathic arthritis coexisting with sickle cell disease: two case reports. *BMJ Case Rep*. 2011. doi: 10.1136/bcr.10.2011.4889.
5. Aletaha D, Neogi T, Silman AJ, et al. 2010 Rheumatoid arthritis classification criteria: an American College of Rheumatology/European League Against Rheumatism collaborative initiative. *Arthritis Rheum*. 2010;62:2569–2581.
6. Schumacher HR, van Linthoudt D, Manno CS, et al. Diffuse chondrolytic arthritis in sickle cell disease. *J Rheumatol*. 1993;20:385–389.
7. Mann D, Schumacher HR. Pseudoseptic inflammatory knee effusion caused by phagocytosis of sickled erythrocytes after fracture into the knee joint. *Arthritis Rheum*. 1995;38:284–287.
8. Darbari DS, Castro O, Taylor JG 6th, et al. Severe vaso-occlusive episodes associated with use of systemic corticosteroids in patients with sickle cell disease. *J Natl Med Assoc*. 2008;100:948–951.

© 2015 Wolters Kluwer Health, Inc. All rights reserved.

Copyright © 2015 Wolters Kluwer Health, Inc. All rights reserved.