Case Report: Effect of Ultrasound Therapy on the Adult-Onset Still's Disease: A Case Study

Manal Youssef[1]

1. Department of Internal Medicine, Cairo University Hospitals, Giza, Egypt.

Use your device to scan and read the article online

Citation Youssef M. Effect of Ultrasound Therapy on the Adult-Onset Still's Disease: A Case Study. Physical Treatments. 2020; 10(1):49-54.

doi: http://dx.doi.org/10.32598/ptj.10.1.?

Article info:
Received: 12 Jun 2019
Accepted: 11 Sep 2019
Available Online: 01 Jan 2020

Keywords:
Physical therapy, Pain, Range of motion, Physical function, Adult-onset still's disease, Ultrasound

ABSTRACT

Adult Onset Still's Disease (AOSD) is a rare systemic inflammatory disease of unrecognized etiology and pathogenesis that presents in 5% to 10% of patients as Fever of Unknown Origin (FUO) accompanied by systemic manifestations. We reported a compelling case of a 39-year-old African male who presented with a one-month duration FUO along with skin rash, sore throat, and arthralgia. After an extensive workup, potential differential diagnoses were ruled out, and the patient was diagnosed with AOSD based on the Yamaguchi criteria.

After the application of Ultrasound (US) and exercises on both ankles as well as left knee, left elbow, and left wrist, there was an improvement of Visual Analogue Scale by 66.6%, Five Repetition Sit to Stand Test (FRSTS) improved 50%, Timed Up and Go (TUG) test improved 87.5%, and there was also an improvement in the 36-Item Short Form Survey (SF-36), and muscle testing. This result was due to the anti-inflammatory effect of the US, leading to decrease pain and improve function in the individuals.

*** Corresponding Author:**
Manal Youssef, PhD.
Address: *Department of Internal Medicine, Cairo University Hospitals, Giza, Egypt.*
Phone: *+20 (100) 7734694*
E-mail: *manal.kamel@cu.edu.eg*

Highlights

- Joint pain occurs in two-thirds of patients, which starts concomitantly with fever.
- The treatment of AOSD is mainly centered on the use of non-steroidal anti-inflammatory drugs.
- Ultrasonic enhance soft tissue healing decreases the inflammatory response, increases blood flow, increases metabolic activity, and decreases pain.
- US and exercises for two weeks decrease pain and increase physical performance on patient with AOSD.

Plain Language Summary

Adult Onset Still's Disease (AOSD) is a rare systemic inflammatory disorder of unknown etiology. It is an immunologic disorder affects women slightly more than men. Its prevalence is estimated to be 1.5 cases per 100 000 to 1.5 cases per 1000 000. Joint pain occurs in two-thirds of patients. The arthralgia starts concomitantly with fever, involves any joint, and may migrate at the beginning and become more stable during the disease. A poor prognosis is associated in AOSD patients with poly articular onset. There is a strong causal relationship between joint damage and subsequent disability. Moreover, AOSD patients may experience several severe complications associated with a decrease in life expectancy. The treatment of AOSD is mainly centered on the use of non-steroidal anti-inflammatory drugs, steroids, and disease-modifying ant rheumatic drugs. Ultrasound is based on the application of high frequency sound waves to the tissues of the body in order to obtain mechanical or thermal effects. These effects aim to enhance soft tissue healing, decrease the inflammatory response, increase blood flow, increase metabolic activity, and decrease pain. Moreover, there is some evidence that ultrasonic energy stimulates the repair of joint cartilage in animal models of cartilage injury. Low-intensity pulsed ultrasound may exert a protective effect on articular cartilage by increasing proteoglycan synthesis and counteracting the catabolic activity of pro-inflammatory cytokines, together with positive effects also by inhibiting subchondral bone sclerosis, particularly in early osteoarthritis stages. Low-intensity pulsed ultrasound may also stimulate chondrocyte proliferation and matrix production, with dose-dependent effects and greater attenuation of cartilage degeneration in the early osteoarthritis phases.

1. Introduction

Adult Onset Still's Disease (AOSD) is a rare systemic inflammatory disorder of unrecognized etiology. It is an immunologic disorder that affects women slightly more than men. Its prevalence is estimated to be 1.5 cases per 100000 to 1.5 cases per 1000000. The disease characteristically affects younger people, with three-quarters of the patients reporting disease onset between 16 and 35 years of age. It is characterized by high spiking fever, arthritis, maculopapular rash, sore throat lymphadenopathy, myalgia, and leukocytosis. Sometimes, the liver, lung, heart, gastrointestinal tract, and the central nervous system might be involved [1].

There is a bimodal age distribution, with one peak between the ages of 15 and 25 years and the second peak between the ages of 35 and 45 years. The clinical course of AOSD could be divided into monophasic, intermittent, and chronic patterns. Systemic manifestations predominate in the two former forms while articular involvement predominates in patients with chronic form.

Compared with clinical examination, Ultrasonography (US) and Magnetic Resonance Imaging (MRI) strongly correlate in terms of detecting superficial erosive bone processes and are more sensitive than conventional radiographic and clinical assessments [2].

There is a strong causal relationship between joint damage and subsequent disability. Moreover, AOSD patients may experience several severe complications associated with decreased life expectancy [3].

Joint pain occurs in two-thirds of AOSD patients. The arthralgia starts concomitantly with fever, involves any joint, and might migrate at the beginning and become more stable during the disease course. A poor prognosis is associated with AOSD patients with polyarticular onset, proximal joint arthritis, prior episode in childhood, and the requirement of systemic steroids for >2 years.

An intra-articular steroid injection can be used in the treatment of the chronic articular pattern of AOSD [4, 5].

The disease pattern of AOSD patients could be divided into 3 distinct types:

1. Monocyclic or self-limiting pattern, which has a single episode of systemic disease of variable duration, followed by complete remission;

2. Polycyclic or intermittent pattern, where 2 or more episodes of systemic disease are separated by symptom-free remission period lasting for a minimum of 2 months;

3.Chronic articular pattern, i.e., characterized by the severe articular manifestations causing joint destruction [5].

Those patients with a chronic articular pattern or a polyarticular onset and course were at higher risks to develop disabling arthritis. Joint involvement at diagnosis might affect functional prognosis through destructions resembling those of rheumatoid arthritis [6, 7].

The treatment of AOSD mainly involves using non-steroidal anti-inflammatory drugs, steroids, and disease-modifying antirheumatic drugs. The effect of corticosteroid injections lasts from 1 to 4 weeks. The prolonged concentration of the corticosteroid in the synovial fluid confers the maximum anti-inflammatory effect locally while minimizing the risk of systemic effects. This duration, together with the possible deleterious effects of large doses, supports the empirical 3-month rule (time-lapse from each injection), i.e., for steroids [8].

AOSD is characterized by a repetitive inflammatory response of the articular cartilage due to the focal loss or erosion of the articular cartilage and hypertrophy of osteoblastic activity or a reparative bone response, known as osteophytotic. Both defining characteristics result in a joint space narrowing or subchondral sclerosis, leading to pain, immobility, and often disability. The symptoms of OA, such as pain and joint stiffness, as well as muscle weakness, are serious risk factors for mobility limitation. They might lead to a reduced quality of life in the affected population. Therefore, Range of Motion (ROM) exercises and stretching have been proposed in this respect, because of their benefits in modulating pain, increasing ROM, reducing soft tissue inflammation, improving repair, extensibility, or stability in the contractile and non-contractile tissues, facilitating movement, and improving function [9].

US works based on the application of high-frequency sound waves to the tissues of the body to obtain mechanical or thermal effects. These effects aim to enhance soft tissue healing, decrease the inflammatory response, increase blood flow, increase metabolic activity, and decrease pain. Moreover, evidence suggested that ultrasonic energy stimulates the repair of joint cartilage in the animal models of cartilage injury [10, 11].

Low-Intensity Pulsed Ultrasound (LIPUS) may exert a protective effect on articular cartilage. Such effect occurs by increasing proteoglycan synthesis and counteracting the catabolic activity of proinflammatory cytokines, positive effects, and by inhibiting subchondral bone sclerosis, particularly in early OA stages. LIPUS may also stimulate chondrocyte proliferation and matrix production, with dose-dependent effects and greater attenuation of cartilage degeneration in the early OA phases [12, 13].

2. Case Report

We presented the case of a 39-year-old male of AOSD. He was diagnosed with AOSD two years ago. He was hospitalized and treated by intraarticular corticosteroids and discharged and rejoined his work in a factory (remission); after another two months, he experienced a relapse and remained the same (remission and relapse phases) for two years. He was hypertensive and consumed concor 5 mg, and he was not diagnosed with diabetes. At that time, he was on a full dose of steroids 60 mg daily. He developed activity, and accordingly, he was arranged for rituximab. The patient presented with arthralgia in the left wrist, left knee, left elbow, and both ankles.

The high-resolution US revealed inactive synovial hypertrophy of mild to a moderate degree in the left wrist. The left knee sonography revealed early degenerative changes with minimal to mild effusion. He was treated by intraarticular corticosteroid injection; however, he developed the sensitivity to corticosteroid injection; he was unable to take the full dose of injection because of severe pain. Subsequently, he was referred to physical therapy due to complaining of the arthralgia of joints. Musculoskeletal examination indicated tenderness and hotness in the bilateral ankles and left knee left elbow. The left wrist of the patient with a limitation of active and passive ROM due to swelling. of the patient's daily living activities was affected by arthralgia and myalgia. Muscle testing revealed mild muscle weakness of both lower limbs.

The following tests were performed at the beginning of the treatment and after two weeks:

1. Visual Analogue Scale (VAS) for pain (range 0-100 mm) was reported every day;

2. Five Repetition Sit to Stand Test (FRSTST) was used as a measure of functional lower limb strength;

3. Timed Up and Go (TUG) test was conducted in seconds as a measure of mobility. The time was taken from the rise from the chair, walking for 3 m, turning, walking back to the chair, and then sitting down.

4. Muscle testing of lower and upper limbs;

5. The 36-Item Short Form Survey (SF-36).

3. Intervention

Therapeutic US (model B-619B, made in China) at doses of (1.0 W/cm^2 at 1 MHz for 5minutes was applied for each joint daily, for two weeks. This practice was followed by static and strengthening exercises for each joint; each exercise was repeated 8-15 times. After two weeks of the application of US and exercises, there were improvements in all parameters: VAS improved by 66.6%, FRSTS improved by 50%, TUG enhanced by 87.5%, and there was also an improvement in the scores of SF-36 and muscle testing.

All measurements were performed before the treatment and after two weeks of intervention completion. All procedures were performed according to the Helsinki declaration of 1975 and its modifications. Informed consent was obtained from the patient for the publication of this case report.

4. Discussion

The present study aimed to measure the effect of US and exercises for two weeks on pain and the physical performance of a patient with AOSD. There were improvements in VAS by 66.6%, FRSTS by 50%, TUG by 87.5%, and there were improvements in the scores of SF-36 and muscle testing.

The American College of Rheumatology (ACR), 2000, recommendations for hip and knee Osteoarthritis (OA) were as follows: non-pharmacological modalities strongly recommended for managing knee OA were aerobic, aquatic, and resistance exercises, as well as weight loss for overweight patients. Manual therapy, walking aids, thermal agents, Tai Chi, self-management programs, and psychosocial interventions were also suggested in this regard. Therapeutic exercise programs provide a short-term benefit, i.e., sustained for at least 2-6 months after the treatment cessation, with a magnitude of the effect being moderate (immediate) to small (2-6 months); however, they are comparable with estimates reported for non-steroids anti-inflammatory drugs. Muscular training may improve joint stability and potentially slows down joint degradation; it also prevents stiffness and articular worsening. Among patients suffering from knee OA, isometric quadriceps exercises have been supported to increase hyaluronic acid molecular weight and synovial fluid viscosity [4].

A recent systematic review examined the relationships between physical activity and individual joint structures. According to its results, despite an increase in radiographic osteophytes, there was no increase in joint space narrowing; however, there was evidence of an associated increase in the cartilage volume and a decrease in cartilage defects on magnetic resonance imaging.

Optimizing cartilage health is essential for preventing further degeneration of early OA. Thus, these findings indicated that physical activity might be beneficial, rather than detrimental, to joint health [9, 14]. Evidence suggested that ultrasonic energy stimulates the repair of joint cartilage in animal models of cartilage injury. Therefore, US could be an effective intervention in managing pain and disability in individuals with knee OA [15].

John Z Srbely analyzed the current literature and stated that 6 studies addressing cartilage repair unanimously support the cartilage-enhancing effects of ultrasound [13]. The frequency and intensity output levels used were consistently in the low-intensity ultrasound range (1 MHz, 200-400 mW/cm^2). Treatment frequency and duration varied; however, ranging from 7 minute exposures of pulsed ultrasound, three times weekly for 4 weeks, in comparison with daily continuous-wave exposures lasting one week in duration. Accordingly, optimal exposure parameters must be standardized before optimal treatments, and clinical protocols could be established [13].

There was no direct evidence that any purported clinical benefits of ultrasound are due to altered membrane permeability. These changes include increases in protein synthesis, growth factor production, calcium uptake, fibroblast mobility, and mast cell degranulation. These changes could account for the improved tissue repair, i.e., alleged to follow US therapy [16].

LIPUS may exert a chondroprotective effect on articular cartilage by increasing proteoglycan synthesis and counteracting the catabolic activity of proinflammatory

cytokines; such process could be combined with positive effects induced by inhibiting subchondral bone sclerosis, particularly in early OA stages. LIPUS may also stimulate chondrocyte proliferation and matrix production, with dose-dependent effects and more significant attenuation of cartilage degeneration in the early OA phases [10, 12].

Sánchez et al. found that US could reduce pain by 21%, compared to a control group [17]. Additionally, US applied using low intensity (<1 W/cm^2), pulsed mode, and a therapeutic dose of <150 J/cm^2 could be more effective at reducing pain than US applied using high intensity (1 W/cm^2), continuous mode and a therapeutic dose of >150 J/cm^2 [17].

5. Conclusion

US and exercise are useful interventions in decreasing pain and improving physical function in patients with AOSD.

Ethical Considerations

Compliance with ethical guidelines

All procedures were performed according to the Helsinki declaration of 1975 and its modifications. Informed consent has been obtained from the patient for the publication of this case report.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Conflict of interest

The author declared no conflict of interests.

References:

[1] Jie W, Miaob L, Yankuna S, Honga Y, Zhongxin XU. Demyelinating encephalopathy in adult onset still's disease: Case report and review of the literatures. Clinical Neurol and Neurosurgery. 2013; 115(10):2213-6. [DOI:10.1016/j.clineuro.2013.06.011] [PMID]

[2] Nataraja C, Griffiths H. Atypical cutaneous manifestations in adult onset still's disease. Case Reports in Rheumatology. 2016; 2016:4835147. [DOI:10.1155/2016/4835147] [PMID] [PMCID]

[3] Ruscitti P, Cipriani P, Masedu F, Iacono D, Ciccia F, Liakouli V, et al. Adult-onset still's disease: Evaluation of prognostic tools and validation of the systemic score by analysis of 100 cases from three centers. BMC Medicine. 2016; 14(1):194. [DOI:10.1186/s12916-016-0738-8] [PMID] [PMCID]

[4] Senthilve E, Papadakis A, McNamara M, Adebambo I. Adult-Onset Still Disease (AOSD). The Journal of the American Board of Family Medicine. 2010 ;23(3):418-22. [DOI:10.3122/jabfm.2010.03.090157] [PMID]

[5] Gopalarathinam R, Orlowsky E, Kesavalu R, Yelaminchili S. Adult onset still's disease: A review on diagnostic workup and treatment options. Case Report in Rheumatology. 2016; 2016:6502373. [DOI:10.1155/2016/6502373] [PMID] [PMCID]

[6] Cush J, Medger T, Christy W, Herbert D, Coopertein L. Adult onset still's disease: Clinical course and outcome. Arthritis & Rheumatism. 1987; 30(2):186-94. [DOI:10.1002/art.1780300209] [PMID]

[7] Guilpain P and Le Quellec A. About the complexity of adult onset Still's disease … and advances still required for its management. BMC Medicine. 2017; 15(1):5. [DOI:10.1186/s12916-016-0769-1] [PMID] [PMCID]

[8] Kon E, Filardo G, Drobnic M, Madry H, Jelic M, van Dijk N, et al. Non-surgical management of early knee osteoarthritis. Knee Surgery, Sports Traumatology, Arthroscopy. 2012; 20(3):436-49. [DOI:10.1007/s00167-011-1713-8] [PMID]

[9] Bhatia D, Bejarano T, Novo M. Current interventions in the management of knee osteoarthritis. Journal of Pharmacy and Bioallied Sciences. 2013; 5(1):30-8. [DOI:10.4103/0975-7406.106561] [PMID] [PMCID]

[10] Hochberg M, Altman R, April K, Benkhalti M, Guyatt G, McGowan J, et al. American College of Rheumatology 2012 recommendations for the use of non-pharmacologic and pharmacologic therapies in osteoarthritis of the hand, hip, and knee. Arthritis Care & Research (Hoboken). 2012; 64(4):465-74. [DOI:10.1002/acr.21596] [PMID]

[11] Singh K, Sobti V, Roy K. Gross and histomorphological effects of therapeutic ultrasound (1 watt/cm^2) in experimental acute traumatic arthritis in donkeys. Journal of Equine Veterinary Science. 1997; 17(3):150-5. [DOI:10.1016/S0737-0806(97)80307-3]

[12] Knoop J, Dekker J, van der Leeden M, van der Esch M, Thorstensson CA, Gerritsen M, et al. Knee joint stabilization therapy in patients with osteoarthritis of the knee: A randomized, controlled trial. Osteoarthritis and Cartilage. 2013; 21(8):1025-34. [DOI:10.1016/j.joca.2013.05.012] [PMID]

[13] Srbely J. Ultrasound in the management of osteoarthritis: Part I: A review of the current literature. J Canadian Chiropractic Association (CCA). 2008; 52(1):30-7.

[14] Urquhart D, Tobing J, Hanna F, Berry P, Wluka A, Ding C, et al. What is the effect of physical activity on the knee joint? A systematic review. Medicine & Science in Sports & Exercise (MSSE). 2011; 43(3):432-42. [DOI:10.1249/MSS.0b013e3181ef5bf8] [PMID]

[15] Cook S, Salkeld S, Patron L, Doughty E, Jones D. The effect of low-intensity pulsedultrasound on autologous osteochondral plugs in a canine model. The American Journal of Sports Medicine. 2008; 36(9):1733-41. [DOI:10.1177/0363546508316766] [PMID]

[16] Baker K, Robertson V, Duck F. A review of therapeutic ultrasound: Biophysical effects. Physical Therapy. 2001; 81(7):1351-8. [DOI:10.1093/ptj/81.7.1351] [PMID]

[17] Sánchez A, Richardson J, MacIntyre N. Efficacy of ultrasound therapy for the management of knee osteoarthritis: A systematic review with meta-analysis. Osteoarthritis and Cartilage. 2010; 18(9):1117-26. [DOI:10.1016/j.joca.2010.06.010] [PMID]