

Case Report On Systemic Sclerosis Treated With Pulse Therapy

1] Ms. Sarika Khadse,

Vice Principal, Florence Nightingale Training College of Nursing, SawangiMeghe Wardha.Email:
sarikaselsurkar@gmail.com, 9503807766

2) Ms. Deepali Ghungrud,

Nursing Tutor, Florence Nightingale Training College of Nursing, Sawangimeghe WardhaEmail:
ghungruddeepali@gmail.com

3) Roshan Umate,

Research Scientist, Jawaharlal Nehru Medical College, Datta Meghe Institute of Medical Sciences,
Sawangi, Wardha, Maharashtra.

4) Vaishnavi Kantode,

Department of Medical-Surgical Nursing, Smt. Radhikabai Meghe Memorial College of Nursing, Datta
Meghe Institute of Medical Sciences, Sawangi, Wardha, Maharashtra.

ABSTRACT:

Introduction: Systemic sclerosis (SS) is an autoimmune disorder. It is a rare chronic disease that caused by abnormal connective tissue growth. This connective tissue provides strength and structure to the organs and muscles. In systemic sclerosis, the tissue becomes thick and stiff that may cause swelling and pain. Systemic sclerosis is also known as scleroderma. Pulse therapies are used for inflammatory and autoimmune conditions. Dexamethasone Pulse therapy is the technique of administering suprapharmacologic doses of medications, especially corticosteroids, alternatingly to improve the therapeutic impact and reduce the side effects of the drug.

Patient's medical history: The patient has come to skin OPD with the chief complaint of darkening and tightening of the skin, which was gradually in onset and progressive in nature. It started in the right upper limb and neck, chest, and then lower limb for seven years. The patient had a history of joint pain in the wrist and knee for 7 years, a history of abdominal pain and bloating for six months, and also a history of body pain and generalized weakness for six months. Another complaint of the patient was a headache and difficulties in opening the mouth—no history of hypertension, diabetes, tuberculosis, etc.

The Main diagnosis, Treatment, and outcomes: On physical examination the general condition was moderate. BP was 122/82 mmHg, and her pulse was 81/bpm. Patients treated with Antibiotics, antacid, pulse therapy, steroids and also provide multivitamins supplements.

Nursing care: IV fluids were given, vital signs were taken every six-hourly, and medication was given according to the doctor's orders.

Conclusion: Treatment and care of systemic sclerosis on time will help prevent further morbidity.

Keywords: Systemic sclerosis, Dexamethasone, pulse therapy

INTRODUCTION:

Systemic sclerosis is a chronic autoimmune condition that affects the multiple systems of our body. It is found rarely. The most prominent feature of systemic sclerosis is extracellular matrix components are deposited excessively in many organs and tissue. Damage to Blood vessels, inflammation, and the development of particular autoantibodies are all signs of systemic sclerosis. In this condition, internal organs and skins are affected, for example, kidneys, musculoskeletal system, heart, lungs, and the gastrointestinal tract. The main symptom of systemic sclerosis is Skin sclerosis.¹ Systemic sclerosis is an uncommon illness of connective tissue having the potential to impact many organs in the body. Through pathogenic processes, the disease process can cause regulation of the body's normal functioning, resulting in vascular, fibrotic, and immunological problems. This study was meant to analyze the coetaneous and systemic clinical manifestations and histological aspects of systemic sclerosis and to connect the severity of the cutaneous manifestations with the level of histopathological change in the form of fibrosis.² There are two categories of Systemic sclerosis is subcategories based on the level of involvement of skin confined systemic sclerosis and diffuses systemic sclerosis.³ In contrast to individuals with fibrosis of the

trunk and proximal portions of the limbs, patients with impacting fibrosis of the skin in sacral regions—face and limbs (distal to the knees and elbows)—are classified as having lcSSc.⁴ Pasricha et al. at the All India Institutes of Medical Sciences (AIIMS), New Delhi, created the dexamethasone cyclophosphamide pulse treatment, which was first used to treat pemphigus in 1986. Intravenous Dexamethasone pulse therapy has been used since 1998. Apart from, systemic sclerosis, Dexamethasone pulse therapy was also found to be effective in pemphigoid disorders, pemphigus disorders, systemic lupus erythematosus, etc.⁵ Various medications are used in the Treatment of systemic sclerosis, including corticosteroids, D-penicillamine, colchicines, azathioprine, and cyclosporin-A with or without physiotherapy. None of these measures were able to halt the progress of the disease. Several skin conditions have been treated successfully with corticosteroid pulse therapy (intermittent high dosage), which has fewer or no side effects than continuous daily corticosteroid medication.⁶

Case Presentation:

A 35 year's old female patient reported in skin ward with remote history of joint pain in the wrist and knee for seven years, a history of abdominal pain and bloating for six months, and also a history of body ache and generalized weakness for six months. Her present complaint was darkening and tightening of the skin, which was gradually in onset and progressive its started from right upper limb (Figure 1) and neck, chest and then lower limb since seven years, patient also complaint of bluish discoloration of fingers (Figure 2) 7 years back. Another history of the patient was pruritis, joint pain in the wrist and knee, oral ulcers present, headache, and difficulties in opening the mouth—no history of hypertension, diabetes, tuberculosis, etc. The doctor diagnosed this systemic sclerosis after a physical examination and investigation.



Figure No-1 systemic sclerosis in right
Upper limb



Figure No- 2 - systemic sclerosis on palm in left hand and bluish discoloration on fingers.

The patient's primary concern and clinical features: The Patient was received into Skin OPD with the chief complaint of darkening and tightening of the skin, which was gradually in onset and progressive in nature it started from the right upper limb and neck, chest, and then lower limb, abdominal pain and bloating, joint pain in wrist and knee, oral ulcers present, headache, difficulties in opening the mouth. No history of diabetes mellitus, hypertension, tuberculosis, etc.

Patient's previous medical and surgical history and family and psychosocial history: a patient suffering from Systemic sclerosis, the patient not having any type of last medical and previous surgical history patient belong, joint family, he was oriented date, time and place, and she has maintained an excellent relationship with his family. He kept his personal hygiene in check. There is no genetic history of hypertension, asthma, TB, or diabetes in the patient's family.

Habit: The patient does not have any negative habits, such as chewing tobacco or smoking, and only watches TV, reads the newspaper, and sleeping also.

Patient present medical:- Patient admitted in rural Hospital with the chief complaint of darkening and tightening of the skin which was gradually in onset and progressive in nature its started from right upper limb and neck, chest and then lower limb, abdominal pain and bloating, joint pain in wrist and knee, oral ulcers present, headache, difficulties in opening the mouth. The doctor diagnosed this case with systemic sclerosis after physical examination and investigation.

Present surgical history: No history of current surgery

CLINICAL FINDINGS:

The patient was awake and aware of the date, time, location, and person. She had kept his body build is good and maintained good self-cleanliness.

The client was conscious and aware of the date, time, place, and person. Pallor, Icterus, clubbing, edema, etc., not present inpatient. Her BP was 122/82 mmHg, and her pulse was 81/bpm.

Cutaneous Examination: Tight, shiny skin over the face with the inability to pinch the skin with decreased forehead wrinkling. Parrot break like nose with loss of nasolabial fold—difficulty in mouth opening.

DIAGNOSTIC ASSESSMENT:

Diagnostic methods: Patient history collection and physical examination is done.

BLOOD INVESTIGATION

Name of investigation	Patient value
Hemoglobin -	11
TLC-	6900
RBS-	163
Urine <rm -	oxalate crystal present
KFT:	Urea-34, creat-0.7, Na-137, K- 4.1
SGPT -	26
SGOT-	23
ALP -	70

Challenges in diagnosing:

During the diagnostic evaluation, there were no difficulties.

The reasoning for diagnosis:

The investigation is completed.

Prognosis:

The prognosis for this instance was favorable.

THERAPEUTIC INTERVENTION:

A case of Systemic sclerosis treated with Pulse therapy, antibiotics, and Multivitamins. The patient completed 27 cycles of half DP pulse on 7/05/2022.

Medication: Tab. Cetirizine OD, Tab. Pan 40, OD, Inj. Dexamethasone 50mg given in 300ml 5% dextrose for pulse dexta therapy, Cap MVBC in OD, Inj. Nifedipine in BD.

Nursing perspective: IV fluids were provided to maintain fluid and electrolyte imbalance. Pulse therapy as per doctor's order, checked the temperature, respiration, pulses, and BP every hour. Intake and output chart monitoring/hourly.

FOLLOW-UP AND OUTCOMES:

Advised the patient to exercise every day and avoid a high-cholesterol diet and eat a healthy diet. Regular checkups, good personal hygiene, and following doctor's orders for medicine are all essential.

OUTCOMES:

Despite all of the care, the patient's progress is good, and he has been recommended to avoid strenuous work and to stay in bed completely.

Intervention adherence and tolerability:

Patients took all prescribed medications regularly, ate a healthy diet, and did not require any intervention. The patient responded well to the treatment.

DISCUSSION:

Steroid pulse therapy is used in many autoimmune disorders like lupus erythematosus, rheumatoid arthritis, pyoderma gangrenous, and bullous pemphigoid. By Pai et al., this therapy was used on only a few clients, and they reported some beneficial effects of steroid pulse therapy in systemic sclerosis. In the same way, Masood et al. have reported preliminary results of dexamethasone therapy in systemic sclerosis. Only 25 patients were included in their study. They were used 50 mg dexamethasone and dextrose 5% intravenously for three days per month for 12-18 pulses only.⁸⁻¹⁶ In their study, ten patients were completing pulse therapy, from and the clients reported to be an improvement in Raynaud's phenomenon. Ulcers were found in 8 patients out of 9 patients .sclerosis improved markedly in 3 patients, moderate in 6 patients, and mild in one patient, and this was assessed by observation visually, when did palpation, and on pre-and post-treatment biopsies skin.¹⁷⁻²⁵ In our study, Dexamethasone systemic sclerosis treated with pulse therapy, the patient has come to our department complaining of tightening of the skin in Rt. Upper limb, neck, chest, and lower abdomen and also complained of bluish discoloration of fingers. We treated the client with Dexamethasone 50mg with 5% dextrose and also gave multivitamins to the client. After treatment and care patient's condition improved.²⁶⁻²⁸

CONCLUSION:

This patient has been admitted to the skin ward in Rural Hospital Wardha with the chief complaint of darkening and tightening of the skin, which was gradually in onset and progressive in nature. It started in the right upper limb and neck, chest, and then lower limb, joint pain on the wrist and knee since abdominal pain and bloating, and also a history of body ache and generalized weakness. On admission, her general condition was moderate. After the physical examination and investigation patient was diagnosed with systemic sclerosis. Treatment and care of a Systemic sclerosis with pulse therapy given to the patient on time will help to prevent further morbidity.

REFERENCES:

1. Sobolewski P, Maślińska M, Wieczorek M, Łagun Z, Malewska A, Roszkiewicz M, et al. Systemic sclerosis – multidisciplinary disease: clinical features and treatment. *Reumatologia* [Internet]. 2019 [cited 2022 Jun 10];57(4):221–33. Available from: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6753596/>
2. Sureshan D, Riyaz N, Thumbayil L. Cross-sectional study on clinical features and histopathology of systemic sclerosis. *JSSTD* [Internet]. 2019 Dec 2 [cited 2022 Jun 10];1(2):77–83. Available from: <https://jsstd.org/cross-sectional-study-on-clinical-features-and-histopathology-of-systemic-sclerosis/>
3. Kowal-Bielecka O, Kuryliszyn-Moskal A. *Twardzinaukładowa*. *Reumatologia*. 2016;(Suppl 1):51–55. [Google Scholar]
4. Barsotti S, Bellando Randone S, Guiducci S, et al. Systemic sclerosis: a critical digest of the recent literature. *Clin Exp Rheumatol*. 2014;32:S194–S205. [PubMed] [Google Scholar]
5. Katakam BK, Kavitha SB, Netha GNR, Shahana M, Sri TS, Vani DS. Prospective Study of Pulse Therapy in Childhood Pemphigus Disorders. *Indian Dermatol Online J* [Internet]. 2018 [cited 2022 Jun 10];9(6):422–5. Available from: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6232984/>
6. Systemic sclerosis treated with dexamethasone pulse [Internet]. *Indian Journal of Dermatology, Venereology and Leprology*. 2002 [cited 2022 Jun 10]. Available from: <https://ijdv.com/systemic-sclerosis-treated-with-dexamethasone-pulse/>
7. Sameem F, Hassan I, Ahmad QM, Khan D, Majeed I, Kamili MA, et al. DEXAMETHASONE PULSE THERAPY IN PATIENTS OF SYSTEMIC SCLEROSIS: IS IT A VIABLE PROPOSITION? A STUDY FROM KASHMIR. *Indian J Dermatol*. 2010;55(4):355–8.

8. Kersten BE, den Broeder N, van den Hoogen FHJ, Knaapen-Hans HAK, van den Ende CHM, Vonk MC. Treatment with cyclophosphamide i.v. pulse therapy is an option for effective treatment of skin fibrosis in patients with early systemic sclerosis. *Rheumatology* [Internet]. 2020 Jul 1 [cited 2022 Jun 10];59(7):1550–5. Available from: <https://doi.org/10.1093/rheumatology/kez487>
9. Madke, B., Agrawal, S., Sharma, A., 2020. Using Transparent Gridded Sheets with Pre-Marked Area Zones for Faster Calculation of Scalp Area for Various Purposes. *INDIAN DERMATOLOGY ONLINE JOURNAL* 11, 861–862. https://doi.org/10.4103/idoj.IDOJ_601_19
10. Mandal, S., Mamidipalli, S.S., Mukherjee, B., Suchandra, K.H.H., 2020. Response to letter to the editor: Perspectives, attitude and practice of lithium prescription among psychiatrists in India. *INDIAN JOURNAL OF PSYCHIATRY* 62, 228–229. https://doi.org/10.4103/psychiatry.IndianJPsychiatry_675_19
11. Micah, A.E., Su, Y., Bachmeier, Financing, G.B.D.H., 2020. Health sector spending and spending on HIV/AIDS, tuberculosis, and malaria, and development assistance for health: progress towards Sustainable Development Goal 3. *LANCET* 396, 693–724. [https://doi.org/10.1016/S0140-6736\(20\)30608-5](https://doi.org/10.1016/S0140-6736(20)30608-5)
12. Nagdive, A., Zaman, R.U., Mansharamani, H.D., Behere, P.B., Fernandes, R., 2020. A Study of Perceived Stress & Coping in Interns in a Tertiary Care Hospital in a North Eastern State of India. *JOURNAL OF EVOLUTION OF MEDICAL AND DENTAL SCIENCES-JEMDS* 9, 3950–3955. <https://doi.org/10.14260/jemds/2020/864>
13. Nimbulkar, G., Garacha, V., Shetty, V., Bhor, K., Srivastava, K.C., Shrivastava, D., Sghaireen, M.G., 2020. Microbiological and Clinical evaluation of Neem gel and Chlorhexidine gel on dental plaque and gingivitis in 20-30 years old adults: A Randomized Parallel-Armed, Double-blinded Controlled Trial. *JOURNAL OF PHARMACY AND BIOALLIED SCIENCES* 12, 345–351. https://doi.org/10.4103/jpbs.JPBS_101_20
14. Pasari, A.S., Bhawane, A., Balwani, M.R., Tolani, P., Ramteke, V., Deshpande, N., 2020. Knowledge about COVID-19 and Practices among Hemodialysis Technicians in the COVID-19 Pandemic Era. *INTERNATIONAL JOURNAL OF NEPHROLOGY* 2020. <https://doi.org/10.1155/2020/6710503>
15. Patel, A.B., Bann, C.M., Garces, A.L., Krebs, N.F., Lokangaka, A., Tshefu, A., Bose, C.L., Saleem, S., Goldenberg, R.L., Goudar, S.S., Derman, R.J., Chomba, E., Carlo, W.A., Esamai, F., Liechty, E.A., Koso-Thomas, M., McClure, E.M., Hibberd, P.L., 2020a. Development of the Global Network for Women’s and Children’s Health Research’s socioeconomic status index for use in the network’s sites in low and lower middle-income countries. *REPRODUCTIVE HEALTH* 17. <https://doi.org/10.1186/s12978-020-01034-2>
16. Patel, A.B., Kurhe, K., Prakash, A., Bhargav, S., Parepalli, S., Fogleman, E., V., Moore, J.L., Wallace, D.D., Kulkarni, H., Hibberd, P.L., 2020b. Early identification of preterm neonates at birth with a Tablet App for the Simplified Gestational Age Score (T-SGAS) when ultrasound gestational age dating is unavailable: A validation study. *PLOS ONE* 15. <https://doi.org/10.1371/journal.pone.0238315>
17. Patel, A.B., Simmons, E.M., Rao, S.R., Moore, J., Nolen, T.L., Goldenberg, R.L., Goudar, S.S., Somannavar, M.S., Esamai, F., Nyongesa, P., Garces, A.L., Chomba, E., Mwenechanya, M., Saleem, S., Naqvi, F., Bauserman, M., Bucher, S., Krebs, N.F., Derman, R.J., Carlo, W.A., Koso-ThomasMcClure, M.E.M., Hibberd, P.L., 2020c. Evaluating the effect of care around labor and delivery practices on early neonatal mortality in the Global Network’s Maternal and Newborn Health Registry. *REPRODUCTIVE HEALTH* 17. <https://doi.org/10.1186/s12978-020-01010-w>
18. Patel, K., Yasobant, S., Charan, J., Chaudhari, M., Gaidhane, A., Saxena, D., 2020. Acceptability and Perceptions of Generic Drugs among Patients, Pharmacists, and Physicians. *JOURNAL OF PHARMACEUTICAL RESEARCH INTERNATIONAL* 32, 40–47. <https://doi.org/10.9734/JPRI/2020/v32i3330948>
19. Patel, M.P., Kute, V.B., Goswami, J., Balwani, M.R., 2020. Hospitals May Become “Disease Hotspots” for COVID-19 Amid Shortage of Personal Protective Equipment. *INDIAN JOURNAL OF CRITICAL CARE MEDICINE* 24, 1145–1146. <https://doi.org/10.5005/jp-journals-10071-23645>
20. Patel, S.A., Patel, A.S., Fulzele, P.R., Mohod, S.C., Chandak, M., Patel, S.S., 2020. Evaluation of the role of propolis and a new herbal ointment in promoting healing of traumatic oral ulcers: An animal experimental study. *CONTEMPORARY CLINICAL DENTISTRY* 11, 121–125. https://doi.org/10.4103/ccd.ccd_128_19
21. Patil, B., Chandak, A., 2020. Comparative Study of 0.2% Ropivacaine & 0.2% Ropivacaine with 0.5 mcg/mL Dexmedetomidine in Epidural Labour Analgesia. *JOURNAL OF EVOLUTION OF MEDICAL AND DENTAL SCIENCES-JEMDS* 9, 1091–1096. <https://doi.org/10.14260/jemds/2020/235>

22. Patil, M., Khatib, M.N., Gaidhane, S., Telrandhe, S., Saxena, D., Unnikrishnan, B., Bhardwaj, P., Gaidhane, A.M., Zahiruddin, Q.S., 2020a. Process of Developing the Online Certification Tests for Research Project Trainees and Assessing the Feasibility for Adaptation in Maternal and Child Health Projects. *JOURNAL OF EVOLUTION OF MEDICAL AND DENTAL SCIENCES-JEMDS* 9, 1446–1449. <https://doi.org/10.14260/jemds/2020/315>
23. Patil, M., Telrandhe, S., Khatib, M.N., Gaidhane, S., Saxena, D., Bhardwaj, P., Unnikrishnan, B., Gaidhane, A.M., Quazi Syed, Z., 2020b. Stimulating Home Environment for Early Childhood Development by Household Play Materials for Under 5 Rural Children in Forest Buffer Zone of Wardha District. *JOURNAL OF EVOLUTION OF MEDICAL AND DENTAL SCIENCES-JEMDS* 9, 1336–1340. <https://doi.org/10.14260/jemds/2020/291>
24. Patil, M., Telrandhe, S., Khatib, M.N., Gaidhane, S., Saxena, D., Saha, S., Bharadwaj, P., Gaidhane, A., Quazi Syed, Z., 2020c. Improvements in developmental outcomes of an 18-month-old child from Rural Wardha - A Case Study. *MEDICAL SCIENCE* 24, 1302–1306.
25. Prasad, N., Bhatt, M., Agarwal, S.K., Kohli, H.S., Gopalakrishnan, N., Fernando, E., Sahay, M., Rajapurkar, M., Chowdhary, A.R., Rathi, M., Jeloka, T., Lobo, V., Singh, S., Bhalla, A.K., Khanna, U., Bansal, S.B., Rai, P.K., Bhawane, A., Anandh, U., Singh, A.K., Shah, B., Gupta, A., Jha, V., 2020. The Adverse Effect of COVID Pandemic on the Care of Patients With Kidney Diseases in India. *KIDNEY INTERNATIONAL REPORTS* 5, 1545–1550. <https://doi.org/10.1016/j.ekir.2020.06.034>
26. Pusdekar, Y., V., Patel, A.B., Kurhe, K.G., Bhargav, S.R., Thorsten, V., Garces, A., Goldenberg, R.L., Goudar, S.S., Saleem, S., Esamai, F., Chomba, E., Bauseman, M., Bose, C.L., Liechty, E.A., Krebs, N.F., Derman, R.J., Carlo, W.A., Koso-Thomas, M., Nolen, T.L., McClure, E.M., Hibberd, P.L., 2020. Rates and risk factors for preterm birth and low birthweight in the global network sites in six low- and low middle-income countries. *REPRODUCTIVE HEALTH* 17. <https://doi.org/10.1186/s12978-020-01029-z>
27. Raisinghani, N., Gupta, A., Kumar, S., Acharya, S., Mahajan, S.N., 2020. Clinical spectrum of patients with Intracranial Hemorrhage at rural teaching hospital. *MEDICAL SCIENCE* 24, 503–508.
28. Ranganathan, K., Kavitha, L., Sharada, P., Bavle, R.M., Rao, R.S., Pattanshetty, S.M., Hazarey, V.K., Madhura, M.G., Nagaraj, T., Lingappa, A., Warnakulasuriya, S., 2020. Intra-observer and inter-observer variability in two grading systems for oral epithelial dysplasia: A multi-centre study in India. *JOURNAL OF ORAL PATHOLOGY & MEDICINE* 49, 948–955. <https://doi.org/10.1111/jop.13056>