

A Case Report of Systemic Lupus Erythematosus with Secondary Sjögren's Syndrome

Dr. A Harischandra Prasad¹, Dr. Shubham Sabne², Dr. Ravindra Varma³, Dr. Jagruti Palav⁴,
Dr. Rajarathinam Raja⁵, Dr. Sachin Bangar⁶, Dr. Sandeep Holkar⁷, Dr. A Harischandra Prasad⁸

¹Junior Resident III, Department of Medicine, Dr V M GMC Solapur
Corresponding Author Email: [harishhhhh110\[at\]gmail.com](mailto:harishhhhh110[at]gmail.com)
Mobile No. 7396180094

²Junior Resident III, Department of Medicine, Dr V M GMC Solapur
Email: [shubhamsabne1030\[at\]gmail.com](mailto:shubhamsabne1030[at]gmail.com)

³Junior Resident III, Department of Medicine, Dr V M GMC Solapur
Email: [ravivarma8043\[at\]gmail.com](mailto:ravivarma8043[at]gmail.com)

⁴Junior Resident II, Department of Medicine, Dr V M GMC Solapur
Email: [jagruti.palav95\[at\]gmail.com](mailto:jagruti.palav95[at]gmail.com)

⁵Junior Resident II, Department of Medicine, Dr V M GMC Solapur
Email: [rajarathinam1403\[at\]gmail.com](mailto:rajarathinam1403[at]gmail.com)

⁶Associate Professor, Department of Medicine, Dr V M GMC Solapur

⁷Assistant Professor, Department of Medicine, Dr V M GMC Solapur

⁸Junior Resident III, Department of Medicine, Dr V M GMC Solapur

Abstract: Systemic lupus erythematosus (SLE) is an autoimmune disease in which organs and cells undergo damage initially mediated by tissue-binding auto antibodies and immune complexes. Most of the patients are women of childbearing age. SLE is 5.5-6.5 times more prevalent in women than men Sjogren's is also an Autoimmune disease characterized by lymphocytic infiltration of exocrine glands resulting in Xerostomia, Dry eyes and profound B cell hyperactivity. Females are more affected than males in this condition with a ratio (Female: Male) of 10-20: 1. With advancement in diagnostic modalities there has been a drastic change in understanding the disease pathology and to find treatment for better outcome for the patients.

Keywords: SLE, systemic lupus erythematosus, Sjogren's syndrome pericardial effusion, Anti nuclear antibodies, schirmer's test, pericardiocentesis

1. Introduction

Systemic lupus erythematosus and sjogrens are autoimmune diseases involving multiple organ systems. Females are more commonly affected than males. The diagnosis of SLE is based on clinical features and presence of autoantibodies. We present you a case of a 16 year old female patient who was diagnosed with Systemic lupus erythematosus with Sjogren's syndrome having pericardial effusion.

2. Case Study

A 16 years old female came to a tertiary care hospital with complaints of Breathlessness, fever since 8 days. On presentation to casualty of our hospital, patient was found to have Breathlessness. There was no History of any Blunt trauma, injury; No H/o PND, palpitation or syncope; No H/o Headache seizures or FND; No H/o Trauma, LOC, ENT bleed; No H/o Nausea vomiting abdominal pain or No H/o Cough, Hemoptysis, Hematemesis and No H/o DM, HTN, COPD, Asthma, TB

Past History: Patient had history of fever 5 months back which was continuous and subsided on taking medications. had developed rash gradually over the face 20 days after fever subsided. Patient also gave history of progressive hair-loss with pigmented lesions over the scalp. Patient gave history of generalized swelling on both upper limbs and lower limbs; joint pains since 1 month and Amenorrhea since 5 months.

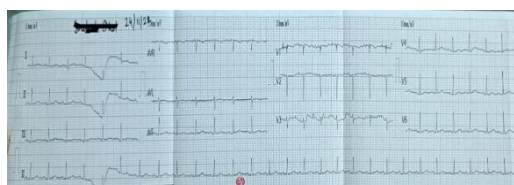
There was No Surgical history. Patient had taken vaccination upto the date. Patient lives in Nuclear family, Non consanguineous marriage, Parents had 3 children 1 Male and 2 females. First female child 18 years old and had pthisis bulbi with Mental retardation. Male child is normal. She is the middle child. Patient consumed mixed diet and had no addictions. Patient was admitted in Medical intensive care unit for the same.

On clinical examination patient was conscious, alert, cooperative, poorly built. Patient had BMI of 19.1 and pulse rate of 134/min regularly regular, Low volume with no radio radial or radio femoral delay with all peripheral pulses palpable. Patient had blood pressure of 90/60 mm hg and spo2 of 94% on 4litres of Oxygen and respiratory rate of

38/min, JVP 10 cm of H₂O. There was presence of pallor and bilateral pedal edema in patient. No cyanosis, icterus, clubbing, lymphadenopathy seen. There was presence of Hair-loss along with pigmented lesions over the scalp, Butterfly shaped lesion over the face, oral mucosal ulcers were present. Shirmer's test was positive indicating dryness of eyes; dryness of mouth was present.



Systemic examination: In Cardiovascular examination: Both heart sounds were heard with no murmur. In respiratory system examination: Trachea shifted to left side and Breath sounds absent in Right Mammary, Infra-mammary area, Infra-axillary area, Infra-scapular area. In Neurological examination -No any neurological deficits, Per-abdomen examination: Abdomen is soft and non-tender. CNS examination: HMF normal, cranial nerves normal, sensory and motor system were normal ECG s/o Sinus Tachycardia with electrical alternans. Chest X ray shows Right lower lobe radiological opacity with Cardiomegaly similar to money bag appearance of heart. HRCT s/o pleural effusion of depth 4.4cm on the right side with collapse of underlying lung with shift of mediastinum to left.



2D Echo f/s/o Normal chamber dimension, LA 24mm, AO 20mm, LVID (d) 36mm, LVID (s) 24mm RA4C: 27mm, IVS 10mm, LVPW (s) 10mm RVID 29mm. No MR/TR/PAH, LVEF 60% and Pericardial effusion of depth 3.6cm, RV side: 20mm and RV Apex: 16mm, No RA/RV collapse.

Routine blood investigations were as follows, Hemoglobin was 6.7 mg/dl at admission, MCV 68.5, Platelet count of

346000 and WBC of 4800. Sodium levels of 138meq/L Potassium - 3.8meq/L, Creatinine of 0.7 and urea of 18. Total bilirubin was 0.9 mg/dl of which Direct was 0.5 mg/dl and Indirect was 0.4 mg/dl. Total protein was 6.1 with Albumin of 2.4 and globulin of 3.7. SGOT was 36 and SGPT 22. PTINR was 1.32. HIV, HBsAg, HCV were negative. Direct coombs test positive. IDCT Negative and Reticulocyte count: 0.8%, S. Calcium: 8.4 and C-reactive protein: 38, LDH: 582u/L.

ANA Blot studies: dsDNA/anti Sm/SS-A/SS-B/Ro-52: STRONGLY POSITIVE

Pleural fluid diagnostic tapping has been done. Pleural fluid proteins-2.6 and Sugar: 26mg/dl. Pleural fluid cytology s/o Total cell count: 1950 cells with 70% lymphocytes and 30% neutrophils and no malignant cells seen. Pleural fluid ADA: 12, Pleural fluid CBNAAT was negative. Thyroid function test was normal. Ophthalmology Fundus examination was normal. Urine R/M: Albumin 2+, Sugar Nil, RBC: 25cells, Pus cells: 5. 24-hour urinary protein was 54mg/dl. Spot Urine protein creatinine ratio was 1.5. Her estimated GFR: 108ml/min/1.73m²Sr. FSH: 1.36mIU/ml, LH: 0.23mIU/ml, E2 Estradiol: 6.79pg/ml. Urine pregnancy test negative. FSH, LH and Estradiol decreased s/o Secondary amenorrhea. C3 was 0.6 (normal range 0.9-1.8), C4 was 0.08 (normal range: 0.1-0.4)

Ultrasonography of abdomen and pelvis was suggestive of Mild hepatomegaly, Kidney size normal with maintained CMD. Right ovary: 1.1x1.2cm and left ovary 1.2x1.2cm with Endometrial thickness-4.7mm. gross Pericardial effusion and moderate to gross right sided pleural effusion noted.

After admission into the intensive care unit, Central line was inserted. The Patient was given 2 PCV transfusion and repeat Hemoglobin reveals 8.7gm/dl. An ICD tube has been inserted into the right side of the lung due to massive pleural effusion. Upon insertion guarded approximately 1.8 litres of pleural fluid has been drained. Post ICD insertion Chest X ray has been taken which show partly recovered Lung.



CXR: Massive right sided pleural effusion



CXR: Right sided ICD insitu with Money bag appearance of heart.

Cardiologist opinion has been taken after 2D echo findings and suggested the need of therapeutic pericardiocentesis. Patient's relatives have been counselled and after consent, under aseptic precautions, insertion of pigtail catheter was

made and pericardiocentesis was done. Pigtail catheter was left in place for few days due to ongoing collection and removed after 10 days.



2D Echo s/o Massive pericardial effusion and the other image s/o post pericardiocentesis revealing mild pericardial effusion.

The above patient has been treated with higher antibiotics and started on Inj. Cyclophosphamide cycles and on maintenance with inj mycophenolate mofetil. Her skin lesions have been treated with Hydroxychloroquine, prednisolone, flucort cream and anti histaminics. Patient also advised carboxymethyl cellulose eye drops for dry eyes.

3. Discussion

Systemic lupus erythematosus is an autoimmune disease with affecting multiple organs and patients of young age. It includes Haematological, Neuropsychiatric, Mucocutaneous, serosal, musculoskeletal and renal abnormalities.

Etiological factors consist of Over 100 gene loci with polymorphisms (or, rarely, copy numbers or mutations); deficiencies of early complement components C1q, C1r, C1s (>90% risk), C4 (50%), C2 (20%), and TREX1; association with genes on the X-chromosome: Female sex and hormonal

influence (e.g. oestrogen); environmental triggers of SLE (drugs, ultraviolet rays, sun exposure, viral infections like Epstein-Barr virus, smoking, silica exposure, vitamin D deficiency, alfalfa sprouts, and foods containing canavanine).

Antibodies are formed against these antigens in the body leads to formation of antigen antibody complexes and get deposited in various organ systems thus causing type III hypersensitivity reaction. This ultimately damages the involved organs causing variety of symptoms like fever, arthritis, serositis, malar rash, nephritis, CNS complications etc. The antibodies might present in the body at least for 3 years before patient presents with symptoms of SLE. ANA test using immunofluorescent methods are more reliable than ELISA and/or bead assays, which have less specificity.

2019 EULAR/ACR Classification criteria for systemic lupus erythematosus is used for diagnosis of SLE: Positive ANA of at-least 1: 80 is obligatory entry criterion followed by additive weighted criteria in 6 clinical and 3 immunological domains. Score of >10 classifies as SLE.

Treatment: There is no cure for SLE, complete sustained remissions are rare. Aim for LLDAs (Low level disease activity): Mild symptoms on lowest possible doses of medications. It uses SLEDAI-2K: It is a measure of SLE disease activity, Score > 3 reflect clinically active disease. Criteria for LLDAS: (1) a SLEDAI-2K score \leq 4, (2) no new lupus activity since previous visit (3) physician's global assessment scale \leq 1 (score 0-3), (4) current prednisolone dose \leq 7.5mg, (5) well-tolerated stable doses of antimalarials and/or immune suppressives.

Flare up can be predicted by 1) falling complement, 2) increasing proteinuria, 3) Worsening anemia and 4) rising anti DNA titres which has been observed in the patient.

Mainstay of treatment for any inflammatory/organ threatening manifestation of SLE is Inj. MPS 500-1000mg iv daily x 3days (Response within 24hrs) f/by 0.5-1mg/kg of daily prednisone. Doses are tapered as rapidly as clinical situation permits to dose <7.5mg of prednisone/day (look for Infection, Hyperglycemia, Hypertension, Osteoporosis)

Sjögren's syndrome (SS) is a chronic autoimmune inflammatory disorder characterized by lacrimal and salivary gland dysfunction with resultant dryness of the eyes and mouth. The most common diseases associated with SS are rheumatoid arthritis and systemic lupus erythematosus. . SS should be suspected in individuals with persistent symptoms of dry eyes and/or mouth, parotid gland enlargement, an unexplained increase in dental caries, or abnormal results of specific serologic tests (e.g., anti-Ro/SSA antibodies with or without anti-La/SSB antibodies, rheumatoid factor, and hyperglobulinemia) Treatment of Sjogren's syndrome aims to relieve symptoms and Limit the damage from chronic xerostomia and keratoconjunctivitis sicca through substitution or stimulation of impaired secretions. Systemic glucocorticoids and rituximab is also found effective against the disease.

4. Conclusions

Early assessment of disease progression and flare ups is essential for prompt management of the disease. Systemic glucocorticoids and immunomodulators (cyclophosphamide, mycophenolate mofetil, rituximab, etc.) help in prevention and disease progression thus leading to decreased morbidity and mortality. Although recurrent flare ups are the sign of poor response to medications.

References

- [1] <https://www.cdc.gov/lupus/facts/detailed>.
- [2] Angel A. Justiz Vaillant; Amandeep Goyal; Matthew Varacallo, systemin lupus erythematosus, <https://www.ncbi.nlm.nih.gov/books/NBK535405/>
- [3] bevrannahshans, Maureen mcmohan, systemic lupus erythematosus, page no. 2736 harrison's principles of internal medicine 21st edition.
- [4] Haralampos M. Moutsopoulos, Clio P. Mavragani, Sjögren's Syndrome, page no. 2787 harrison's principles of internal medicine 21st edition.
- [5] Kelleys and firestein's textbook of rheumatology 11th edition.