An emergency in medicine can be defined as a situation that endangers life, or an organ system or quality of life. In that sense SLE itself is an emergency.

Systemic lupus erythematosus (SLE) is an inflammatory autoimmune disorder that affects multiple organ systems and causes significant morbidity and mortality. Patients of SLE are especially prone to develop serious life-threatening complications. We will first look at the disease as a killer and then cover major emergencies. Over the years the rate and trends in mortality of SLE have shown a significant improvement.

The improved survival of a patient with SLE is due to early diagnosis, recognition of milder cases, better or more effective treatment of the disease and infections and overall improved socio-economic conditions, especially in the developed countries. There is a suggestion that the disease tends to be more severe in our part of the world.

Causes of death in SLE patients are - active disease, infection, myocardial infarction, cerebrovascular accidents, ruptured aneurysm, pulmonary embolism, sudden death, renal failure and deaths unrelated to SLE.

SLE manifestations that indicate poor prognosis are anaemia, leucopenia, thrombocytopenia, nephritis, hypertension, cardiac affection, pulmonary involvement and SLE DAI > 20 (A measure of disease activity). Non-SLE related factors also influence the overall prognosis.

These are:

i. Race
   The disease is more severe in African blacks, Chinese and South-east Asians.

ii. Age at onset
   Children carry a poorer prognosis compared to adults.

iii. Socio-economic conditions and health delivery systems

iv. Environmental & geographic influence
Compliance and education

Infection
Infection forms an important emergency in SLE. It is estimated (Western data) that the infection rate in SLE is 59/100 patient year. This figure is likely to be higher in our context. Common sites of infection are urinary tract, lungs, joints, central nervous system, abdomen and skin. Susceptibility to infection is due to immune dysfunction, as manifested by phagocytic dysfunction, lymphopenia (CD4 + lymphocytopenia), decreased cytokine production, decreased immunoglobulin production, impaired functioning of complement system and functional asplenia. Susceptibility to infection is related to disease activity. This is compounded by the adverse effects of glucocorticosteroids and immunosuppressives used to treat the disease. Infections are most common with corticosteroid and cyclophosphamide.

Bacteria account for more than 90% of infections. The bacterial spectrum includes S. pneumoniae, Staph aureus, E. coli, Salmonella, Klebsiella spp, Pseudomonas spp and Mycobacterium tuberculosis. Amongst viral infections, cytomegalovirus, HBV and Herpes zoster are important. Candida, Nocardia and Aspergillus are the common fungal infections. Strongyloides stercoralis, Pneumocystis carinii and Toxoplasma gondii are some of the parasitic infections encountered.

Amongst the bacteria, Salmonella bacteraemia with extraintestinal manifestations is somewhat specific. The extraintestinal manifestations include urinary tract infection, mycotic aneurysms, arthritis, pericarditis, osteomyelitis and soft tissue abscesses. Mortality is high (25%). Extrapulmonary manifestations of Nocardia are common and these carry high mortality (35%). CNS affection has almost 100% mortality. PCP infection is characteristic of immunosuppressed patients. Mortality is around 12.5%. Invasive fungal infections are uncommon. There are many issues related to infection. Infection exacerbates disease activity. It also mimics disease ares and often distinction between the two becomes difficult. Unless both the disease and infection are treated vigorously and simultaneously, the outcome is unfavourable. Often multiple organisms and multiple sites are involved. Early diagnosis of infection is important. There is a case for prophylaxis against common infections.

Respiratory Emergencies

A) Upper airways obstruction due to
   Laryngeal inflammation
   Epiglottitis
   Vocal cord paralysis
   Retropharyngeal abscess
   These need early recognition and urgent intubation. SLE patients are more susceptible to develop post intubation stricture

B) Lungs
   Lupus pneumonias
   Pulmonary haemorrhage
   Pulmonary hypertension
   Shrinking lung syndrome
   Pulmonary embolism
   Lupus pneumonitis is uncommon. A high index of suspicion is necessary. It should be suspected after excluding infection as the cause of pneumonia. Culture, BAL and even tissue biopsy may be required. Lupus pneumonitis presents with cough, dyspnoea and haemoptysis. The white cell count is normal and cultures are negative. Other features of active lupus are usually present. Most patients need ventilatory support. Secondary infections are common. Treatment is with high dose glucocorticosteroids ± antibiotics ± immunosuppressive therapy. Mortality is high (upto 50%).
Alveolar haemorrhage though rare is a serious complication of SLE. Cough, dyspnoea and haemoptysis are the presenting symptoms. The spectrum of haemorrhage varies from mild or even no haemoptysis to catastrophic life threatening haemorrhage. Haemoptysis, falling haematocrit and pulmonary infiltrates form the classical triad of pulmonary haemorrhage. Respiratory failure with mechanical ventilatory support may be required. Most patients have features of active lupus, most commonly nephritis. The differential diagnosis includes infection and lupus pneumonias. Other diagnostic considerations are Wegener’s granulomatosis and Goodpasture’s syndrome. Secondary infection may complicate the picture. The management is similar to that of lupus pneumonitis. Pulmonary embolism is usually seen in the context of antiphospholipid syndrome. The diagnostic and therapeutic approach is as per the standard guide-lines. Pulmonary hypertension presents with progressive dyspnoea and dry cough. Diagnosis is by exclusion of lung parenchymal and cardiac causes. 2D Echo is usually adequate to confirm the diagnosis. The condition carries a poor prognosis. Treatment includes calcium channel blockers, anticoagulation, prostacyclin or its analogues and oxygen. Newer therapies like bosentan and sildenafil are under investigation. Acute reversible hypoxaemia and shrinking lung syndrome may be occasionally encountered.

Cardiovascular Emergencies
Premature coronary artery disease (CAD) with myocardial infarction
Vasculitis
Myocarditis
Pericarditis
Valvular affection
SLE predisposes to accelerated atherosclerosis. In ammation, vascular injury, hypertension, lipid abnormalities (due to nephrotic syndrome and steroids) are major factors involved in atherogenesis. In the West, CAD is the leading cause of late deaths in patients of SLE. In one series, the relative risk of MI was 52 times more in SLE females aged between 35-44 year. CABG is associated with higher complication rates. It is easy to miss the symptoms of angina and myocardial infarction in young females. Preventive strategies need to be followed in all SLE patients. Coronary artery vasculitis is a rare cause of myocardial infarction. Antiphospholipid antibodies cause myocardial infarction with normal coronary arteries. Pericarditis with tamponade is relatively rare as also constrictive pericarditis. Response to steroids is good. Inappropriate tachycardia may be the only manifestation of myocarditis. It responds to steroids. Drugs and viral aetiologies need to be excluded.

Neuro-psychiatric Emergencies
Lupus cerebritis
Cerebrovascular accidents
Aseptic meningitis
Seizures
Transverse myelitis
Psychosis
Organic brain syndrome
Extrapyramidal syndrome
GBS
Stroke is an important neuromanifestation. Any area of the brain can be affected. The relative risk for
all types of strokes is 7.9%. APL syndrome is an important cause. Contrary to the usual belief, cerebral vasculitis is not a common cause of ischaemic cerebrovascular accidents. APL syndrome patients need to be on life long anticoagulation. Cerebral haemorrhage may be due to rupture of aneurysms, (secondary to vasculitis), apart from the usual causes.

Transverse myelitis can present acutely or subacutely. APL syndrome is an important cause, as is, also vasculitis. CSF examination and MR are important diagnostic modalities. Prognosis for recovery is not good. Early institution of high dose steroids along with cyclophosphomide and plasmapheresis may prove beneficial.

Seizures of all types can occur in SLE patients. These often precede other SLE manifestations. Status epilepticus can be a preterminal event. Vasculitis, APL syndrome, embolism and haemorrhage are important causes. Thrombotic thrombocytopenic purpura is a rare cause. Aseptic meningitis needs to be differentiated from other causes of meningitis. CSF usually shows 200-300 cells/mm³, with lymphocytic predominance. NSAIDS especially ibuprofen have been incriminated in some cases. Treatment is with corticosteroids.

Gastrointestinal emergencies
Bowel perforation
Pancreatitis
Bowel ischaemia
Rupture of hepatic artery aneurysm

Patients with active SLE are more likely to develop vasculitis of abdominal vessels. Bowel ischaemia and perforation warrant early diagnosis and emergency laparotomy, while simultaneously continuing treatment for active lupus. Pancreatitis poses difficult therapeutic problem as steroids can cause pancreatitis. However, in most patients, pancreatitis is a manifestation of active lupus and patients need to be treated with steroids, apart from usual measures. Mortality is high.

Haematologic emergencies
Autoimmune haemolytic anaemia
Thrombocytopenia
APL syndrome
Thrombotic thrombocytopenic purpura

These are treated along the standard lines. Haemolytic anaemia and TTP may be the first manifestations of lupus.

Renal
Rapidly progressive glomerulonephritis is an important renal emergency. Early diagnosis with energetic treatment (plasmapheresis, steroid & cyclophosphomide pulses) can prevent development of irreversible renal damage. Endoxan pulse therapy is indicated to treat WHO class III and class IV nephritis.

Pregnancy
Pregnancy poses special problems. Disease flares can occur. There is an increase in pregnancy related complications such as toxaemia. Neonatal lupus, intrauterine growth retardation (IUGR), heart block, abortion and premature delivery are the hazards to the foetus. An active disease is not an indication for MTP. Prednisolone even in large doses is safe as it does not cross placental barrier. For treatment of foetal affection (evolving heart block) dexamethasone or betamethose is to be used (cross placental).
Anti-Ro antibody is the risk marker. Such pregnancies need close monitoring with periodic foetal echocardiograms.

APL antibodies cause repeated abortions in second trimester, toxaemia, IUGR, premature labour & still births. Anticoagulation along with aspirin are indicated throughout the pregnancy.

Conclusion
It is clear that SLE covers the whole gamut of internal medicine. Treating SLE emergencies can be extremely taxing and sometimes frustrating; but the rewards are a million times more satisfying.

References
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