EMERGENCIES IN SYSTEMIC RHEUMATIC DISEASES

SYSTEMIC RHEUMATIC DISEASES ARE CHRONIC, INFLAMMATORY, AUTOIMMUNE DISORDERS SUCH AS RHEUMATOID ARTHRITIS, SCLERODERMA OR LUPUS

MANY EXTRA-ARTICULAR MANIFESTATIONS OF RHEUMATIC DISEASES CAN RESULT IN SERIOUS MORBIDITY AND MORTALITY IF NOT RECOGNISED

INFECTION IS THE LEADING CAUSE FOR ICU ADMISSION, FOLLOWED BY RHEUMATIC DISEASE FLARE

AIRWAY EMERGENCIES:

- Critical airway obstruction may develop at many levels
- UPPER AIRWAY OBSTRUCTION:
    - Cricoarytenoid joint arthritis:
        - Occurs in RA, SLE → arthritis or oedema of the cricoarytenoid joints can lead to acute upper airway obstruction
        - Signs/symptoms → throat pain or tenderness over cartilaginous structures which is aggravated by swallowing or speaking
        - Treatment → systemic high-dose steroids and antibiotics if infection is suspected
    - Angioedema:
        - In patients with SLE, which is associated with C1 esterase inhibitor deficiency
    - Subglottic stenosis:
        - Can be observed in Wegener granulomatosis, with inability to clear secretion
        - Often requires surgery
    - Intubation considerations in these patients:
        - Considered difficult and anticipate need for adjunctive techniques
        - RA/ankylosing spondylitis patients develop TMJ dysfunction with ↓d mouth opening and also have high incidence of C1-2 instability → avoid hyperextension
        - Patients with anylosing spondylitis are at high risk for fractures even after minor trauma
        - In scleroderma, involvement of skin and face and neck can be severe → very difficult to intubate

PULMONARY EMERGENCIES:

- Lung involvement may be due to the underlying disease itself or secondary to infection and is a frequent cause of morbidity and death → especially in scleroderma, SLE, RA, Wegener's

- Pulmonary complications of systemic rheumatic disease manifest primarily as interstitial lung disease and vascular disease -→ develop respiratory failure even with minor infection
- Respiratory arrest has been reported in SLE and dermatomyositis due to phrenic nerve involvement, in RA due to cervicomedullary compression and in Sjogren due to hypokalaemia due to distal renal tubular acidosi
- Acute complications → pneumonia or alveolar haemorrhage
- ALVEOLAR HAEMORRHAGE:
    - Uncommon but catastrophic
    - Can be complication of SLE, Wegener, dermatomyositis, scleroderma
    - For improvement in outcome, early recognition and aggressive management are critical
    - Delay in treatment, age >60, ESRF worsen prognosis
    - Symptoms → abrupt onset SOB, fever, cough with rapid progression to respiratory failure
        - Classical triad (HAEMOPTYSIS, PULMONARY INFILTRATES ON CXR, RAPID FALL IN HAEMOGLOBIN) → not always present
            - New lung infiltrates (83-100%) and anaemia (75-100%) appear the most sensitive
    - If symptoms are accompanied by high fever, it is difficult to distinguish between infection and alveolar haemorrhage → but this is crucial as TREATMENT ARE DIAMETRICALLY OPPOSED → immunosuppression for alveolar haemorrhage and antibiotics for infection
    - BAL may aid diagnosis
    - TREATMENT → IMMUNOSUPPRESSION WITH HIGH DOSE STEROID, CYCLOPHOSPHAMIDE, PLASMA EXCHANGE, LOCAL VESSEL EMBOLISATION
- INTERSTITIAL LUNG DISEASE:
    - Leads to fibrosis, pulmonary hypertension and respiratory insufficiency
    - Seen in RA, scleroderma, HSP
    - Produces slowly progressive symptoms but can cause acute respiratory failure in some
- PULMONARY HYPERTENSION:
    - Can be complication of any rheumatic disease but is especially common in scleroderma and SLE due to pulmonary vasculitis and pulmonary embolism

CARDIOVASCULAR EMERGENCIES:

- Heart disease develops through several mechanisms

ACUTE CORONARY SYNDROMES:

- SLE, antiphospholipid, RA promote accelerated coronary atherosclerosis and have increased rate of CV morbidity and mortality
- The risk of MI is pronounced in young women with SLE → premenopausal women have up 50 fold increased risk compared to women without lupus

- Young patients may also get pericarditis
- MANAGEMENT IS THE SAME AS FOR THE GENERAL POPULATION FOR ACS

ACUTE HEART FAILURE:

- May develop as a consequence of myocardial disease, pericardial disease (tamponade or constriction), valvular diseaes (acute regurgitation) and rhythm disturbance
- Acute management does not differ except when a flare of the underlying disease is responsible
- In scleroderma → diastolic dysfunction, malignant hypertension during a renal crisis and decompensated pulmonary HT are important causes of acute heart failure

CARDIAC ARRHYTHMIAS:

- Rhythm and conduction disturbances are common,e specially in RA, systemic sclerosis, SLE and dermatomyositis
- Sudden cardiac death has higher occurrence in patients with systemic rheumatologic diseases → ventricular arrhythmia especially common in scleroderma

MALIGNANT HYPERTENSION:

- Systemic HT may be a clinical manifestation of renal injuries or an adverse effect of treatment
- Severe malignant HT occurs in scleroderma, usually during a "renal crisis"

VASCULAR DISEASE OF THE AORTA/GREAT VESSELS:

- Arterial lesions can be aneurysmal, or occlusive
- Aortitis may lead to aortic aneurysms inthose with GCA, Takayasu arteritis and Behcet disease → aneurysm may dissect or rupture

THROMBOEMBOLISM:

- DVT/superficial thrombophlebitis common in Behcet disease → usually not associated with peripheral embolism and treatment with anticoagulant and fibrinolytics may increase risk of fatal haemorrhage
- Thromboembolic disease also seen in antiphospholipid syndrome
- Vessel thrombosis can be arterial or venous and can affect any vessel

CATASTROPHIC ANTIPHOSPHOLIPID SYNDROME:

- This is an uncommon vaso-occlusive process that leads to MOF and is usually associated with SLE
- Multiple simultaneous venous and/or artieral occlusive thromboses (mainly small vessels) occur and multi-organ failure proceeds rapidly
- In half cases → a precipitating event is identified the kidney is the primary organ involved, followed by lungs, CNS, heart

- LIVEDO RETICULARIS (lacelikel purple discolouration of lower limbs) and thrombocytopaenia are important diagnostic findings
- TREATMENT IS IV HEPARIN, HIGH DOSE STEROIDS, IMMUNOGLOBULIN AND PLASMA EXCHANGE → mortality approaches 50%

NEUROLOGIC EMERGENCIES:

- Can result from accelerated atherosclerosis, direct involvement by theprimary disease, vasculitis, aneurysm, dissection
- GCA has predilection for extracranial vessels, especially vertebrobasilar circulation

SPINAL CORD COMPRESSION:

- OCCURS IN RA AND ANKYLOSING SPONDYLITIS:
    - Atlantoaxial joint preferentially involved in RA and can sublux or dislocate → acute cervical myelopathy, even after minor trauma
        - Patients present with severe neck pain with occipital radiation and paraesthesiae and weakness of upper limbs
        - Can lead to progressive quadriparesis
        - MRI crucial
    - Ankylosing spondylitis leads to rigid spinal column that is easily fractured

OCULAR EMERGENCIES:

- The eye is sensitive barometer for onset or flare of rheumatic diseases
- Sudden and permanent blindness is a concern in HCA → patients report amaurosis fugax, new headache, tender scalp, visual disturbance, jaw/tongue/upper extremity claudication
    - HIGH DOSE STEROIDS ARE MANDATORY, EVEN BEFORE BIOPSY OF THE TEMPORAL ARTERY

RENAL EMERGENCIES:

- Kidney is almost always involved in systemic rheumatic disease and is a major source of morbidity and mortality
- Renal disease is the most common causes of death in scleroderma and a sudden catastrophic form of renal involvement is called SCLERODERMA RENAL CRISIS
    - 75% cases occur in first four years and is characterised by abrupt and rapid deterioration in renal function
    - Classically patients have severe headche, visual disturbance and hypertensive encephalopathy
    - Mainstay of therapy is EFFECTIVE AND PROMPT BLOOD PRESSURE CONTROL→ ACE-Inhibitor is agent of choice
- Distal renal tubular acidosis occurs in 30% of patients with primary Sjogren syndrome → characterised by HYPERCHLORAEMIC, METABOLIC ACIDOSIS WITH HYPOKALAEMIA AND LOW BICARBONATE

GI EMERGENCIES:

- Complications include ischaemia/infarction, perforation, vascular rupture, infection and haemorrhage
- GI haemorrhage is the most common GI manifestation reported
- Chronic or recurrent ischaemia may lead to abdominal wall oedema, strictures and stenosis
- Acute pancreatitis is also common in severe systemic rheumatic diseases

INFECTIOUS EMERGENCIES:

- Rheumatic disease predispose to infection in several ways
    - The disease itself has immunosuppressive effect
    - TREATMENT is immunosuppression
    - The disease can lead to anatomic changes that favour infection
- EMPIRIC ANTIBIOTICS ARE INDICATED UNTIL INFECTION CAN BE EXCLUDED
- Beware opportunistic pathogens due to immunosuppressive (Candida, PCP, Legionella, TB)
- THERE IS A SIGNIFICANT DELAY IN DIAGNOSIS OF SEPTIC ARTHRITIS IN PATIENTS WITH R.A. (often up to three weeks) → associated with mortality of 30% and joint destruction/poor function in survivors.