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**Connective Tissue diseases:**

 Are groups of clinico pathological conditions involve connective tissue of most systems of the body, include mainly LE, scleroderma, systemic sclerosis, dermatomyositis, MCTD &others…

**Lupus Erythematosus:**

 is an autoimmune disorder resulting from an interplay of genetic, environmental &hormonal elements with a heterogeneous clinical expression extending from a localized cutaneous form to a life threatening systemic form.

 LE is a spectrum of diseases, in one end of spectrum: DLE is purely cutaneous LE, at the other end :SLE. In the middle of spectrum: sub acute LE, neonatal LE, complement deficiency LE& drug-induced LE .



**Exacerbating factors:**

**1. Ultraviolet light**

**2. Sex hormones**

**3. Stress**

 **Creteria for classification of SLE: SLE criteria ( require 4 out of 11):**

**1.**Malar rash :fixed erythema over malar eminences, sparing nasolabial folds.

**2**.Discoid rash :erythematous raised patches with adherent keratotic scaling &follicular plugging .

**3**.Photosensitivity :skin rash as a result of unusual reaction to sunlight .

**4**.Oral ulcer: usually painless.

**5**.Arthritis:non erosive arthritis involving 2 or more peripheral joints ,characterized by tenderness, swelling or effusion .

**6**.Serositis:pleurisy or pricarditis.

**7**.Renal disorder: persistent proteinuria >0.5g/day or cellular casts.

**8**.Neurological disorder: seizure or psychosis .

**9**.Hematological disorder: Hemolytic anemia ,leucopenia<4000/mm3, lymphopenia <1500/mm3, or thrombocytopenia less than 100.000/mm3.

**10**.Immunological disorder: **anti-DNA, anti-Sm, antiphospholipid antibodies**

 **11.**Abnormal antinuclear Ab titer

**Drug-induced SLE:**

***It is different from idiopathic SLE by* presence of anti-histone antibodies instead of ANA**

***Most commonly implicated drugs*:**

***Procainamide, hydralazine, minocycline, INH, penicillamine and TNF-alpha inhibitors***

**Diagnosis:**

**Hx e.g joint pain and swelling, worsening or appearance of skin lesions with sun exposure, weight loss, fever( which indicates systemic involvement), Hx of drugs**

**Physical exam: scar, follicular plug, dyspigmentation , oral ulcer, pleural or pericardial rub,**

**Investigations: CBC ( decreased WBC, RBC or platelets), ESR(high in SLE), urinalysis ( cast or protein), ANA (sensitive but not specific)**

**If ANA is positive go to specific autoantibodies e.g. anti-dsDNA for SLE**

**Treatment:**

**Topical: Sun protection, topical and intralesional steroids**

**Systemic: Antimalarial e.g. hydroxychloroquine, chloroquine**

**Morphea**

**Affect female more than male**

**Does not affect survival but can cause a disability especially the linear type**

**Fibroblast isolated from morphea lesion produce increased amount of collagen and this is thought to be due to production of IL-4 and TGF-β by T-cells**

**Some believes that Borrelia plays a role**

***Clinical types:***

**1. Plaque-type : present as shiny indurated plaque surrounded by lilac border**

**2. Deep morphea: invlove deep dermis, subcutis +/- fascia**

**3. Generalized morphea: plaques coaleasce affecting the entire trunk except nipple, can involves the extremities, it is disabiling and causing difficulty in breathing. Distinguished from systemic sclerosis by :**

 **(a) Absence of Raynaud's phenomenon,**

 **(b) Absence of internal organ involvement and**

 **(c) Asymmetry of involvement**

**4. Linear morphea: different from plaque morphea by:**

 **(a) Childhood onset,**

 **(b) High ANA titer and**

 **(c) Disabling especially when involve joint or cause atrophy of the whole limb**

**Variants of linear morphea:**

**En coup de sabre type (sword hit): linear morphea of head, can involve muscle, bone and rarely brain causing seizures**

**Parry-Romberg syndrome: hemi facial atrophy including eyes and tongue (the most severe form of linear morphea)**

***Diagnosis:***

**Hx and physical examination e.g. hardenening of skin lesion**

**Investigations e.g. autoantibodies: ANA and anti-ssDNA are commonly seen in linear and generalized types**

**Biopsy: hyalinized and thick collagen bundles**

**Treatment:**

**Topical :**

**Corticosteroid is ineffective**

**Vit D analogues e.g. calcipotriol may be of benefit**

**Systemic :**

**Glucocorticoids, methotrexate , PUVA (psoralen plus UVA)**

**Cutaneous manifestations of scleroderma:**

**1. Hardening of skin (hard to pinch)**

**2. Microstomia (hard to open mouth) with furrowing around mouth**

**3. Beaking of nose**

**4. Loss of facial expression**

**5. Telangiectasia of skin, lip and tongue**

**6. Ulcers and necrosis of finger tips**

**7. Calcinosis cutis (deposition of calcium in skin, subcutaneous tissue and muscle)**

**8. Nail fold telangiectasia**

**Dermatomyositis**

**Classification (1):**

**Polymyositis (muscle only)**

**Amyopathic dermatomyositis ( skin only)**

**Dermatomysitis (skin and muscle)**

**Classification (2):**

**Juveile type: not associated with malignancy but associated with more calcinosis than adult type**

**Adult type: associated with malignancy especially ovarian, lung and breast**

**Clinical features:**

**Cutaneous:**

**Heliotrop rash : violaceous patch and edema around eyes**

**Gottron papules: flat-topped violaceous papules on knuckles**

**Gottron sign: violaceous discoloration of knuckles, elbows and knee**

**Photodistribution of skin manifestations (shawl distribution)**

**Nail fold telengictasia**

**Calcinosis cutis: deposition of calcium in skin, subcutis and muscle**

**Systemic(extracutaneous) :**

**Proximal myopathy (most important): inability to comb, to walk upstairs or to stand from sitting position**

**Lung: interstitial lung disease, restrictive lung disease**

**Heart: conduction defects, arrhythmia**

**Diagnosis:**

**History :. appearance or worsening of skin lesions with sun exposure**

**Physical examination: e.g. nail fold telengictasia, heliotrop rash, etc…**

**inability to walk upstairs, physical exam of muscle strength**

**Investigations :**

Increased muscle enzymes in the serum ex. creatine phosphokinase (CPK) and adolase.

- Increased 24 - hour urinary creatine.

- Electromyography (EMG).

- Magnetic resonance imaging (MRI) or spectroscopy.

- Serum antibodies: **autoantibodies such as ANA, anti-Jo1, anti-Mi2 antibodies**

- Muscle biopsy.

- Searching for internal malignancy in adult dermatomyositis.

**Treatment: the same as cutaneous LE (but less responsive to treatment than cutaneous LE)**

Oral steroids.

- Cytotoxic drugs.

- Antimalarials.

 - Intravenous immunoglobulin.

- Physical therapy.