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Archith Boloor

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9

CHAPTER

Rheumatology and Connective Tissue Disorder Diseases

Differences between inflammatory and non-inflammatory arthritis discussed in **Table 9.1**.

Causes of acute and chronic arthritis is listed in **Box 9.1**.

TABLE 9.1: Inflammatory versus noninflammatory arthritis.

Features	Inflammatory (rheumatoid arthritis)	Non-inflammatory (osteoarthritis)
Age of onset	Usually 20–40 years but may begin at any age	Most commonly over 50 years of age
Speed of onset	Rapid over weeks to months	Slow; over years
Systemic symptoms	Fatigue, low-grade fever, anorexia Extra-articular manifestations: Rheumatoid nodules, Sjögren's syndrome, Felty syndrome	No systemic symptoms
Joint affection	Symmetrical	Asymmetrical
Joint symptoms	Painful, swollen stiff joints and muscle aches	Joints painful without swelling
Joints involved	Primarily affects small joints [metacarpophalangeal (MCP) and proximal interphalangeal (PIP)]	Affects large weight bearing joints (hip, knee or the spine). Affects PIP and distal interphalangeal (DIP) joints
Stiffness	Morning stiffness for > 1 h. Stiffness occurs after periods of rest/inactivity (the so-called gel phenomenon)	Morning stiffness for < 30 min. Stiffness is generally mild and occurs after periods of activity
Relation of movement with pain	Movement or mild-to-moderate activity decreases pain	Movement increases the pain (worsens with activity) and improves with rest
Examination of joint	Swollen, red, warm, tender and painful	Swollen, cool and hard on palpation. When severely inflamed (as in acute gout or septic arthritis), can have erythema of the overlying skin
Radiological findings	Bony erosion, soft tissue swelling, angular deformities, periarticular osteopaenia	Loss of joints space and articular cartilage, routine wear and tear, osteophytes

Continued

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Continued

Features	Inflammatory (rheumatoid arthritis)	Non-inflammatory (osteoarthritis)
Laboratory findings		
Rehumatoid factor (RF), antinuclear antibody	Positive	Negative
Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP)	Both are often raised	Usually normal but transient elevation of ESR may occur due to synovitis
White blood cell (WBC) count in the synovial fluid	WBC count is > 2,000/mm^3 in septic arthritis and not in rheumatoid arthritis (RA)	< 200/mm^3

Box 9.1: Causes of acute and chronic arthritis.

Actue monoarthritis
- **Inflammatory:** Crystal disease (e.g. gout), infectious disease, spondyloarthropathy, rheumatoid arthritis (RA)
- **Mechanical/inflammatory:** Trauma, avascular necrosis

Acute polyarthritis
- **Infectious:** Bacterial, HIV
- **Non-infectious:** Rheumatoid arthritis, spondyloarthropathy, other connective tissue diseases, crystal (gout), sarcoidosis, malignancy, leukaemia, sickle cell anaemia

Chronic monoarthritis
- **Inflammatory:** Crystal disease, infectious disease (e.g. tuberculosis, fungal), spondyloarthropathy, RA
- **Non-inflammatory:** Osteoarthritis, avascular necrosis, neuropathic arthropathy, villonodular synovitis

Chronic polyarthritis
- **Inflammatory:** Rheumatoid arthritis, spondyloarthropathy, other connective tissue diseases
- **Mechanical:** Osteoarthritis
- **Crystal:** Gout
- **Metabolic:** Infiltrative, metabolic, hypothyroidism
- **Malignancy**

RHEUMATOID ARTHRITIS

- Females:Males (3:1)
- Fourth and fifth decades of life
- Association with human leucocyte antigen **(HLA)-DR4** in 70% of patients

Clinical Features

- Fatigue, anorexia, weakness and vague musculoskeletal symptoms
- Joint involvement is usually **symmetric**.
- The metacarpophalangeal (MCP) and proximal interphalangeal (PIP) joints of the hands, wrists, knees, and the metatarsophalangeal (MTP) and PIP joints of the feet are the most common joints involved.
- '**Morning stiffness**' lasting > 1 hour is a characteristic feature.

Deformities Seen

- 'Spindling' of the fingers
- 'Swan-neck' deformity
- 'Boutonniere' deformity
- 'Z' deformity
- 'Broadening' of the forefoot
- Hallux valgus deformity

Extra-articular Manifestations

- Rheumatoid nodules
- **Rheumatoid vasculitis:** Polyneuropathy and mononeuritis multiplex
- **Pleuropulmonary manifestations:** Effusion, interstitial fibrosis and nodules (Caplan syndrome)
- Premature atherosclerosis
- **Neurological manifestations:** Carpal and tarsal tunnel syndromes, atlanto-axial subluxation
- **Ophthalmological manifestations:** Scleritis, episcleritis and scleromalacia perforans
- **Felty's syndrome:** Splenomegaly and granulocytopenia with rheumatoid arthritis (RA)

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- Osteoporosis
- **Haematological manifestations:** Anaemia and thrombocytosis

Diagnosis: ACR/EULAR Criteria

- **American College of Rheumatism/European League** Against Rheumatism (ACR/EULAR) criteria are given in **Table 9.2**.

Investigations

- Markers of acute inflammation: Raised Erythrocyte sedimentation rate (ESR), anaemia, thrombocytosis, increased C-reactive protein (CRP)
- Rheumatoid factor (RF)
- Anti-citrullinated peptide antibody (ACPA)

Management

- **Analgesics:**
  Nonsteroidal anti-inflammatory drugs (NSAIDs) are the first-line drugs.
- **Disease-modifying antirheumatic drugs (DMARDs):**
  - Hydroxychloroquine
  - Oral gold (auranofin)
  - Parenteral (intramuscular) gold
  - D-penicillamine
  - Sulphasalazine
  - Methotrexate
  - Leflunomide
- **Biological response modifiers (or biologicals):**
  - Anti-tumour necrosis factor alpha (TNF-α) inhibitors (infliximab, adalimumab, golimumab and certolizumab) and a soluble receptor fusion protein (etanercept)
  - Other biological include:
    - Rituximab (anti-CD20 antibody that blocks CD20 present on B cells)
    - Tocilizumab [a monoclonal antibody that binds to interleukin 6 (IL-6) receptors]
    - Abatacept (T-cell receptor CTLA-4 that downregulates T cells)
    - Anakinra (IL-1 receptor blocker)
- **Corticosteroids**

TABLE 9.2: ACR/EULAR criteria.

Features	Score
• Joint involvement:	
○ 1 large joint	0
○ 2–10 large joints	1
○ 1–3 small joints	2
○ 4–10 small joints	3
○ > 10 joints	5
• Serology:	
○ Negative rheumatoid factor (RF) and negative anti-citrullinated peptide antibody (ACPA)	0
○ Low-positive RF or low-positive ACPA	2
○ High-positive RF or high-positive ACPA	3
• Acute-phase reactants:	
○ Normal C-reactive protein (CRP) and normal erythrocyte sedimentation rate (ESR)	0
○ Abnormal CRP or abnormal ESR	1
• Duration of symptoms:	
○ Less than 6 weeks	0
○ 6 weeks or more	1

(ACR/EULAR: American College of Rheumatism/European League Against Rheumatism)

ADULT-ONSET STILL'S DISEASE

- A triad of:
  - High-spiking fever
  - Characteristic evanescent skin rash
  - Arthritis/arthralgias
- Investigations show anaemia, leucocytosis, thrombocytosis, raised ESR, elevated liver enzymes.
- Ferritin levels usually higher.
- Treatment is with NSAIDs and steroids. Role of DMARDs in not clear.

SJÖGREN'S SYNDROME

- A triad of:
  - Keratoconjunctivitis sicca
  - Xerostomia
  - Mononuclear cell infiltration of salivary gland

Clinical Features

- Dryness of eyes
- Xerostomia
- Enlargement of the parotid occurs with primary Sjögren's syndrome.

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- Interstitial nephritis or glomerulonephritis
- Cutaneous and systemic vasculitis
- Polyneuropathies or mononeuritis multiplex.

Laboratory Investigations

- Raised ESR, leucopoenia and thrombocytosis.
- Antibodies to Ro/SSA or La/SSB, antinuclear antibody (ANA) or RF are positive

Treatment

- Ocular and mucosal lubricants such as artificial tears, ophthalmologic lubricating ointments and lubricating agents for dryness of mouth
- Cyclosporine emulsion is useful in ocular dryness.
- Pilocarpine and cevimeline
- Hydroxychloroquine

■ ANKYLOSING SPONDYLITIS/ MARIE-STRUMPELL DISEASE

- Seronegative
- Primarily affects the axial skeleton with a predilection for lumbar spine and sacroiliac joints
- About 90% of the affected people carry the histocompatibility antigen HLA-B27.
- Male-to-female ratio of 4:1

Clinical Features

- Low back pain with nocturnal exacerbations
- Low back morning stiffness that improves with activity
- Schober's test positive
- Figure of four tests (Patrick's test) positive
- Enthesitis
- Diminished chest expansion (< 5 cm) and thoracic kyphosis

Extra-articular Manifestations

- Acute anterior uveitis
- Iritis
- Aortic regurgitation and heart failure
- Apical pulmonary fibrosis and cavitation
- Osteoporosis
- Myelopathy secondary to atlanto-axial subluxation and spinal fracture
- Cauda equina syndrome
- Amyloidosis

Investigations

- ESR is raised. Tests for RF are negative.
- HLA-B27 is present in > 90% cases.
- Magnetic resonance imaging (MRI) and bone scan can pick up early sacroiliitis.

Management

- Indomethacin is the most effective drug.
- Sulphasalazine and methotrexate
- Infliximab, etanercept and adalimumab

■ REITER'S SYNDROME

- Classical Reiter's syndrome is a triad of:
  i. Non-specific urethritis
  ii. Conjunctivitis
  iii. Arthritis 2–4 weeks following enteric or urogential infections
- Common enteric pathogens triggering the disease are *Shigella*, *Salmonella*, *Yersinia* and *Campylobacter* species.
- Enthesitis and dactylitis (inflammation of entire digit) are very common.
- Keratoderma blennorrhagica
- Circinate balanitis

Investigations

- Raised ESR
- Anaemia and polymorphonuclear leucocytosis
- HLA-B27 is positive in > 75% of cases.
- Serum tests for RF and ANAs are negative.

Management

- Rest and NSAIDs
- Intra-articular or local steroid injections
- Non-specific urethritis is treated with a short course of tetracycline or ciprofloxacin.
- Biologicals such as infliximab, etanercept, adalimumab and thalidomide in refractory cases.

■ PSORIATIC ARTHRITIS

Types

- Symmetric psoriatic arthritis
- Asymmetric psoriatic arthritis
- Arthritis mutilans
- Psoriatic spondylitis
- Predominant distal interphalangeal arthritis

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Clinical Features

- Involvement of proximal and distal interphalangeal joints with characteristic 'sausage-shaped' digits
- Distal interphalangeal predominant type of psoriatic arthritis
- Characteristic skin lesion of psoriasis may be present.
- Characteristic nail changes of psoriasis are pitting, onycholysis, subungual hyperkeratosis and horizontal ridging.
- Uveitis, unilateral or bilateral, generally chronic

Diagnosis

- Raised ESR and negative tests for RF and ANAs
- Radiological findings are similar to those of RA.
- Radiographs of distal interphalangeal joints may show pencil-in-cup changes because of marked resorption of bone.

Management

- Majority of the patients respond to NSAIDs.
- Other modalities of treatment include:
  - Sulphasalazine
  - Methotrexate
  - Photochemotherapy
  - Infliximab, etanercept and adalimuimab

BEHCET'S DISEASE

Criteria of Behcet's disease are given in **Table 9.3**.

Diagnosis

- Based on clinical features
- Pathergy test

TABLE 9.3: Major and minor criteria of Behcet's disease.

Major criteria	Minor criteria
• Recurrent aphthous stomatitis • Erythema nodosum • Genital ulcerations • Iritis	• Inflammatory arthritis of large joints (non-deforming) • Intestinal ulceration • Meningoencephalitis • Epididymitis • Thrombophlebitis

Treatment

- For mucocutaneous manifestations, local steroids and oral colchicine
- For joint manifestations, NSAIDs are sufficient.
- Aspirin for thrombophlebitis
- Systemic steroids along with cytotoxic drugs (methotrexate, azathioprine, chlorambucil, cyclosporine A or cyclophosphamide) for uveitis

HENOCH–SCHÖNLEIN PURPURA/ ANAPHYLACTOID PURPURA

- Small-vessel vasculitis
- The characteristic features are vasculitic purpura, abdominal pain, haematuria and acute arthritis.

Treatment

- Most patients respond to bed rest and NSAIDs.
- Corticosteroids are indicated in severe systemic disease.

SYSTEMIC LUPUS ERYTHEMATOSUS

- Second and third decades, with a female/male ratio of 9:1.

Clinical Features

- Fatigue, malaise, fever, anorexia, nausea and weight loss.

Renal Disease

- Characterised by proteinuria (> 0.5 g/24 h) and/or red cell casts (**Box 9.2**)

Musculoskeletal Features

- Arthralgia and myalgia in most patients
- The classical 'Jaccoud's arthropathy' occurs without any erosion.

Box 9.2: Classification of lupus nephritis.

- Class I: Minimal mesangial lupus nephritis
- Class II: Mesangial proliferative lupus nephritis
- Class III: Focal lupus nephritis
- Class IV: Diffuse lupus nephritis
- Class V: Membranous lupus nephritis
- Class VI: Advanced sclerosing lupus nephritis

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Mucocutaneous Features

- Malar rash
- Discoid rash
- Photosensitivity
- Alopecia
- Painless mouth ulcers

Haematological Features

- Normocytic normochromic anaemia
- Coombs-positive haemolytic anaemia
- Thrombocytopaenia
- Leucopoenia

Cardiovascular Features

- Myocarditis and pericarditis
- Endocarditis (Libman–Sacks endocarditis)

Pulmonary Features

- Pleurisy
- Pleural effusion
- Pneumonitis
- Vanishing lung syndrome
- Pulmonary haemorrhage

Gastrointestinal Features

- Hepatosplenomegaly
- Mesenteric vasculitis

Investigations

Immunological Abnormalities

Autoantibodies in patients with systemic lupus erythematosus (SLE) are given in **Box 9.3**.

Diagnostic Criteria

ACR criteria: Presence of four or more criteria is diagnostic (**Table 9.4**).

Systemic lupus international collaboration clinics (SLICC) classification 2012 criteria are given in **Table 9.5**.

Box 9.3: Autoantibodies in patients with systemic lupus erythematosus (SLE).

- ANA
- Anti-DNA (anti-dsDNA and anti-ssDNA)
- Anti-Sm (Anti-Smith)
- Anti-RNP
- Anti-Ro (SSA)
- Anti-La (SSB)
- Anti-histone
- Anti-cardiolipin (aCL)
- Anti-erythrocyte
- Anti-platelet
- Anti-lymphocyte
- Anti-neuronal

TABLE 9.4: ACR (American College of Rheumatism) criteria.

Criteria:	
1. Malar rash	
2. Discoid rash	
3. Photosensitivity	
4. Oral ulcers	
5. Arthritis	
6. Serositis	Pleuritis or pleural effusion/pericarditis or pericardial effusion
7. Renal disorder	Persistent proteinuria > 0.5 g/day/cellular casts
8. Neurologic disorder	Seizures/psychosis
9. Haematologic disorder	Haemolytic anaemia/ leucopoenia/ lymphopoenia/ thrombocytopaenia
10. Immunologic disorder	Anti-DNA antibody/anti-Sm antibody
11. Antinuclear antibody	

TABLE 9.5: Systemic lupus international collaborating clinics (SLICC) classification 2012 criteria.

Lupus nephritis proved by biopsy and ANA or anti-DNA or at least four criteria (one needs to be immunological)	
Clinical	**Immunological**
• Acute cutaneous LE	• ANA
• Chronic cutaneous LE	• Anti-dsDNA
• Oral ulcer	• Anti-Sm
• Alopecia	• aPL antibodies
• Synovitis	• Low complement
• Serositis	• Direct Coombs' test positive
• Renal	
• Neurologic	
• Haemolytic anaemia	
• Leucopoenia/lymphopaenia	
• Thrombocytopenia	

(ANA: antinuclear antibody; Anti-Sm: autoantibodies against Smith antigen; aPL: antiphospholipid; dsDNA: double stranded deoxyribonucleic acid; LE: lupus erythematosus)

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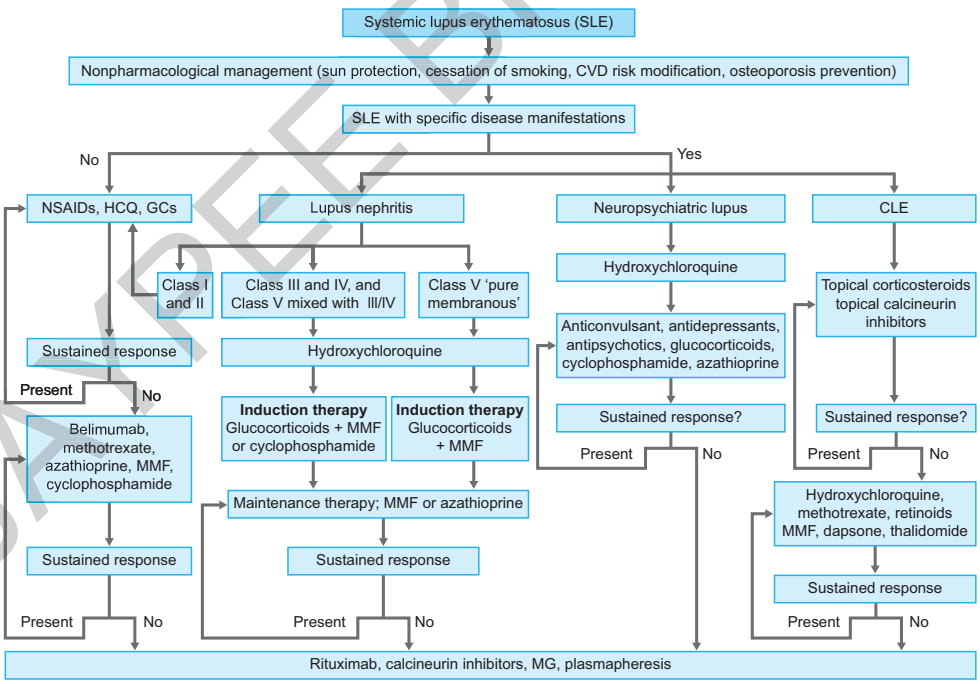
Management

- NSAIDs
- Skin lesions and arthritis—hydroxychloroquine
- Photosensitive skin lesions—sun-screen lotions
- Patients with severe symptoms or life-threatening features—corticosteroids
- Acutely ill patients and patients with proliferative glomerulonephritis
  - 'Pulses' of methylprednisolone for 3 days (1 g/day) followed by oral steroids
- Azathioprine, methotrexate, cyclophosphamide and mycophenolate mofetil

Treatment algorithm for SLE is shown in **Flowchart 9.1**.

DRUG-INDUCED LUPUS

- Drugs implicated:
  - Antiarrhythmics- procainamide, diltiazem, disopyramide, and propafenone
  - Antihypertensive- hydralazine, methyl dopa, angiotensin-converting enzyme inhibitors, beta-blockers
  - Antithyroid drugs- propyl-thiouracil
  - Antipsychotics- chlorpromazine and lithium
  - Anticonvulsants- carbamazepine and phenytoin)
  - Antibiotics -isoniazid, minocycline
  - Antirheumatic -sulphasalazine
  - Diuretic- hydrochlorothiazide
  - Antihyperlipidemics- lovastatin and simvastatin).
  - Biologic agents- Interferons and TNF inhibitors.
- Less female predilection than SLE
- Kidneys or brain involvement rare, less cutaneous involvement
- ANAs positive; antihistone antibodies positive (95%); autoantibodies to dsDNA absent
- C3 levels normal



(CLE: cutaneous lupus erythematosus; GCs: glucocorticoids; HCQ: hydroxychloroquine; MMF: mycophenolate mofetil; NSAIDs: non-steroidal anti-inflammatory drugs; CVD: cardiovascular disease)

Flowchart 9.1: Treatment algorithm for systemic lupus erythematous.

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ANTIPHOSPHOLIPID ANTIBODY SYNDROME

Clinical Features

Clinical features of antiphospholipid antibody (APLA) syndrome are given in **Table 9.6**.

Diagnosis

- Anticardiolipin (aCL) antibodies
- Lupus anticoagulant (LA) antibodies
- Anti-β2-glycoprotein I (IgG and/or IgM)

Treatment

Treatment of venous thrombosis:

- Anticoagulation with heparin and oral warfarin
- Steroids are often added to this treatment.
- In severe cases of generalised thrombosis, plasmapheresis or intravenous immunoglobulin may be tried.

Prevention of recurrence of arterial thrombosis: Aspirin

SYSTEMIC SCLEROSIS

- Diffuse cutaneous systemic sclerosis/limited cutaneous systemic sclerosis

Clinical Manifestations

- Raynaud phenomenon
- Diffuse cutaneous scleroderma is characterised by symmetrical skin thickening of proximal and distal extremities, face and trunk.
- Kidney and other systemic involvement
- Limited cutaneous scleroderma is associated with:
  - Skin thickening limited to distal extremities and face
  - CREST syndrome (calcinosis, Raynaud phenomenon, oesophageal dysfunction, sclerodactyly and telangiectasia)
  - Muscle weakness
  - Arthralgia
  - Resorption of terminal phalanges
  - Dysphagia, reflux oesophagitis and sliding hiatus hernia
  - Pulmonary fibrosis

Investigations

- Elevated ESR
- Anaemia
- Anti-topoisomerase I (previously called anti-Scl-70) and anti-centromere antibodies

Treatment

- Control of Raynaud phenomenon
- Management of gastroesophageal reflux disease (GERD)
- D-Penicillamine—reduces skin thickening
- Cyclophosphamide—lung disease and severe skin disease
- Methotrexate—scleroderma overlap syndromes

TABLE 9.6: Clinical features of antiphospholipid antibody (APLA) syndrome.

Organ/system	Manifestations
Arterial/venous	Thrombosis in any artery or vein
Cardiac	Angina, myocardial infarction, valvular vegetations
Haematologic	Thrombocytopaenia (in 40–50%) haemolytic anaemia, disseminated intravascular coagulation
Neurologic	Transient ischaemic attack, stroke, headache, mononeuritis multiplex
Obstetrical	Pregnancy loss, intrauterine growth retardation
Ophthalmologic	Retinal artery/vein thrombosis
Renal	Thrombosis of renal artery/vein, acute renal failure, haematuria
Gastrointestinal (GI)	Budd–Chiari syndrome, intestinal infarction, splenic infarction, pancreatitis
Cutaneous	Ulcers and infarcts in the skin, gangrene
Pulmonary	Pulmonary embolism and pulmonary hypertension

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POLYMYOSITIS AND DERMATOMYOSITIS

- The skeletal muscle is damaged by an inflammatory process dominated by lymphocytic infiltration.
- Polymyositis spares the skin.
- **Dermatomyositis**—associated with a characteristic skin rash.

Clinical Manifestations

- Weakness of the proximal muscles of the lower limbs (hips and thighs)
- Weakness of the proximal muscles of the upper limbs (should girdle muscles)
- The distal muscles are spared.
- Other features are dysphagia, respiratory impairment, myocarditis resulting heart failure and arrhythmias, arthralgia and interstitial lung disease.
- **Characteristic skin changes:**
  - Classic lilac-coloured (heliotrope) rash is seen on the upper eyelids.
  - Gottron's papules
  - Shawl sign or the V sign
  - Mechanic's hand

Diagnosis

- ESR is usually raised.
- ANAs
- Characteristic electromyography (EMG) findings (myopathic pattern) may be present.
- Muscle biopsy shows the typical pathologic changes in myositis.

Management

- Prednisolone
- If no improvement, azathioprine is to be added.

The differences between dermatomyositis, polymyositis and inclusion-body myositis are given in **Table 9.7**.

POLYMYALGIA RHEUMATICA

- Proximal myalgias are characteristic.
- There are chronic, symmetric, proximal muscle aching and stiffness usually involving shoulder, pelvic girdles and neck. Muscles may be tender on palpation.
- Neuropsychiatric manifestations are frequent, particularly depression.
- About 10–30% of these patients have associated temporal arteritis.

Investigations

- Elevated ESR (> 50 mm/h) and CRP, and normocytic normochromic anaemia
- Tests for RF and ANAs may be positive.
- Muscle enzymes are characteristically normal.

Treatment

- Prednisolone

TABLE 9.7: Differences between dermatomyositis, polymyositis and inclusion-body myositis.

Features	Dermatomyositis	Polymyositis	Inclusion-body myositis
Sex	F > M	M = F	M > F
Age of oneset	Any	20 years +	50 years +
Onset	Subacute/acute	Chronic	Chronic
Distribution of weakness	Proximal	Proximal	Proximal + distal + asymmetric (typically quadriceps + finger flexors)
Muscle pain/swelling	In acute cases	No	No
Skin involvement	Often	No	No
Raynaud's arthralgia	Frequent	Infrequent	No
Dysphagia	In severe cases	Infrequent	Occasional
Association with cancer	Up to 20%	Probably no	No
Cardiac involvement	Yes	No	No
Interstitial lung disease	Associated with anti-Jo	Associated with anti-Jo	No

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VASCULITIS

Classification of Vasculitis is described in **Figure 9.1**.

TEMPORAL ARTERITIS, CRANIAL ARTERITIS OR GIANT-CELL ARTERITIS

- Exclusively in people over the age of 55 years
- Often occurs in the carotid arteries and their branches.
- The disease is characterised by the classic complex of:
  - Fever
  - Anaemia
  - High ESR
  - Headache (usually temporal) in an elderly patient
- The temporal artery may become tender, thickened and cord-like or nodular.
- Scalp pain, and claudication of tongue and jaw are frequent.
- Visual symptoms including diplopia and even sudden blindness may result from ischaemic optic neuritis.
- This condition is very sensitive to corticosteroid therapy.

POLYARTERITIS NODOSA

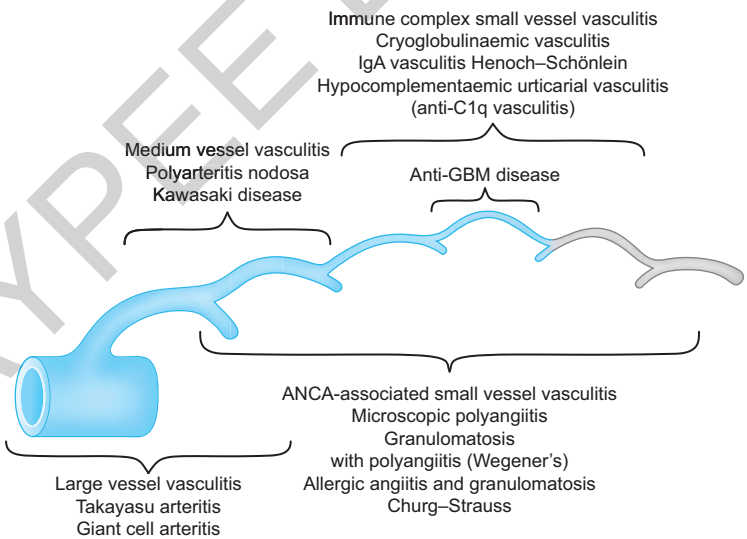
- Necrotising vasculitis of small- and medium-sized muscular arteries
- Kidneys, heart, liver and GI tract are involved.
- It does not involve pulmonary arteries.

Clinical Features

- Predominantly affects males
- Common clinical manifestations are:
  - Hypertension
  - Renal failure
  - Polyarthritis
  - Myalgia
  - Peripheral neuropathy
  - Mononeuritis multiplex

Diagnosis

- Anaemia, raised ESR and neutrophilic leucocytosis
- About 30% are positive for hepatitis B surface antigen (HBsAg).



(ANCA: antineutrophil cytoplasmic antibody; GBM: glomerular basement membrane)

Fig. 9.1: Classification of vasculitis.

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- Perinuclear anti-neutrophil cytoplasmic antibody (p-ANCA) is present in only about 20% of cases.
- Characteristic findings of vasculitis on biopsy material of involved organ confirm the diagnosis.

Treatment

- Corticosteroids, cyclophosphamide and plasma exchange

MICROSCOPIC POLYANGIITIS

- Most common ANCA-associated small-vessel vasculitis
- Characterised by absence of immune deposits in the involved vessels

Clinical Features

- About 40–60 years. More common in males compared to females
- Presentation is by variable combinations or renal involvement.
- Palpable purpura
- Abdominal pain
- Cough and haemoptysis
- Kidney involvement is present in nearly 90% cases.

Diagnosis

- Most patients have positive p-ANCA, although cytoplasmic ANCA (c-ANCA) may also be present in 40% cases.

Treatment

- Combined treatment with cyclophosphamide and corticosteroids has reduced mortality and morbidity significantly.

CHURG–STRAUSS SYNDROME

- Allergic granulomatous angiitis affects small- to medium-sized arteries and veins.

Clinical Features

- History of asthma and allergic rhinitis that is followed by small-vessel vasculitis and granulomatous inflammation
- Common features are:
  - Arthralgia and arthritis
  - Purpura
  - Myocarditis, congestive heart failure, pericariditis and myocardial infarction
  - Abdominal pain, diarrhoea and GI bleed
  - Mononeuritis multiplex
  - Stroke
  - Coronary arteritis and myocarditis are the principal causes of morbidity and mortality.

Investigations

- Eosinophils > 10% in peripheral blood
- Elevated ESR and CRP
- Urine showing RBC casts and proteinuria
- Increased IgE levels
- Positive antinuclear cytoplasmic antibodies against myeloperoxidase (p-ANCA) in 15–60% of cases
- Chest X-ray may show infiltrates and pleural effusion.

Treatment

- Most patients respond to high-dose steroids though some cases may require addition of cytotoxic drugs.
- In severe cases, anti-TNF-α agents such as infliximab and etanercept

WEGENER'S GRANULOMATOSIS

- Granulomatous vasculitis of upper and lower respiratory tracts together with glomerulonephritis

Clinical Features

- Paranasal sinus pain and discharge, nasal mucosal ulcerations, saddle-nose deformity and hearing loss
- Pulmonary involvement—cough, haemoptysis and dyspnoea
- Renal involvement-glomerulonephritis, resulting in proteinuria, haematuria

Diagnosis

- The ESR is markedly elevated.
- Serum anti-proteinase 3 (c-ANCA) is positive.
- Kidney or lung biopsy

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Treatment

Treatment is similar to that for microscopic polyangiitis.

GOUT AND HYPERURICAEMIA

Aetiology

Aetiology of gout and hyperuricaemia is given in **Box 9.4**.

Clinical Features

- Asymptomatic hyperuricaemia
- Acute gouty arthritis
- Intercritical period
- Chronic tophaceous gout (tophi and chronic gouty arthritis)
- The MTP joint of the great toe (podagra) in 70% of patients
- Gout can also cause bursitis and tenosynovitis.
- In chronic tophaceous gouty arthritis, crystal deposits appear in cartilage, synovial membranes, tendons and soft tissues. The classic location of a tophus is the helix and antihelix of the ear.

Box 9.4: Aetiology of gout and hyperuricaemia.

Increased purine synthesis de novo:
- Hypoxanthine-guanine-phosphoribosyl transferase (HGPRT) deficiency
- Phosphoribosyl pyrophosphate (PRPP) synthetase overactivity
- Glucose-6 phosphatase deficiency
- Idiopathic

Decreased renal excretion of uric acid:
- Renal failure
- Lead poisoning
- Alcohol
- Drugs: Diuretics, aspirin, pyrazinamide, cyclosporine and levodopa
- Lactic acidosis
- Hyperparathyroidism
- Myxoedema

Increased turnover of purines:
- Myeloproliferative disorders
- Lymphoproliferative disorders
- Cancer chemotherapy
- Haemolysis

- Urate nephropathy
- Obstructive uropathy (nephrolithiasis)

Diagnosis

- Increase in serum uric acid concentration (> 7.0 mg/dL in males and > 6.0 mg/dL in females)
- Aggregated deposits of monosodium urate monohydrate (tophi) in and around the joints
- Synovial fluid examination by compensated polarised microscopy can demonstrate urate crystals. They are seen as slender, needle-shaped, negatively birefringent structures.

Management

Treatment of acute attack:
- NSAIDs are the agents of choice. All NSAIDs are equally effective. Commonly used NSAIDs are indomethacin (50 mg 6 hourly), naproxen and fenoprofen.
- Colchicine is the second choice of drug.
- In patients with polyarticular involvement not responding to NSAIDs or colchicines

Drugs for prophylaxis:
- Allopurinol
- Febuxostat
- Uricosuric agents such as probenecid and sulphinpyrazone

PSEUDO-GOUT, CALCIUM PYROPHOSPHATE DIHYDRATE DEPOSITION DISEASE AND PYROPHOSPHATE ARTHROPATHY

- Crystal deposition occurs in fibrous and articular cartilage.
- Release of calcium pyrophosphate dihydrate (CPPD) crystals into the joint space provokes an acute attack of synovitis—'pseudo-gout'.
- Mostly asymptomatic; identified as an incidental finding on radiographs.
- The knee is the most frequently affected joint.
- Acute attacks of pseudo-gout may be precipitated by trauma, joint surgery, sprain or even a long walk.
- Chronic pyrophosphate arthropathy resembles osteoarthritis involving
  - Knees
  - Wrist

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