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 This is a well-defined oval to round plaque that fails to meet the criteria for generalised morphea. generalized morphea, linear morphea pansclerotic morphea Pathophysiology • autoimmune component is suggested by enhanced T helper 2 (Th2) dependent interleukin 4 (IL-4) activity, which in turn upregulates transforming growth factor beta (TGF -beta). • TGF-beta stimulates fibroblast production of collagen and other extracellular matrix proteins. Features • Unlike systemic sclerosis, morphea lacks features such as sclerodactyly, Raynaud phenomenon, nailfold capillary changes, telangiectasias, and progressive internal organ involvement. • Morphea can present with extracutaneous manifestations, including fever, lymphadenopathy, arthralgias, fatigue, central nervous system involvement, Investigations • Hypergammaglobulinaemia (↑↑IgM , IgG) • peripheral eosinophilia • ↑↑ESR and CRP • Anti-Cu/Zn superoxide dismutase antibodies have been found in up to 90% Treatment • Superficial circumscribed morphea Tacrolimus 0.1% ointment applied twice daily for 12 weeks may be a useful firstline • Generalized, linear, or deep morphea combination therapy with oral prednisone and methotrexate To minimize the risk of relapse, the recommended treatment duration of MTX is at least 2 years. Systemic corticosteroids can be helpful in the inflammatory phases of morphea, but they are not recommended for long-term monotherapy Mycophenolate mofetil is a second-line Prognosis • generally resolves within 3–5 years, although sometimes a patch may persist for over 25 years.

Polymyalgia rheumatica (PMR) Pathophysiology • overlaps with temporal arteritis 30% of patients also have giant cell arteritis. • histology shows vasculitis with giant cells, characteristically 'skips' certain sections of affected artery whilst damaging others • muscle bed arteries affected most in polymyalgia rheumatica Epidemiology • occurring in patients age 50 years or older. • More common in women

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Rheumatology

Features • typically patient > 60 years old very rarely seen in the under 50s. • usually rapid onset (e.g. < 1 month) • typically presents with pain and stiffness of the shoulder and pelvic girdle muscles. • aching, morning stiffness in proximal limb muscles (not weakness) Pain and muscle

stiffness worst in the mornings • mild polyarthralgia, lethargy, • depression, • low-grade fever, anorexia, night sweats • Weight loss Investigations • ESR > 40 mm/hr the next best investigation a high ESR would prompt immediate treatment with steroids. • Raised C reactive protein (CRP) • Alkaline phosphatase is an acute-phase reactant and is raised in approximately a third of patients with polymyalgia rheumatica. • note CK and EMG normal • reduced CD8+ T cells • Normochromic / normocytic anaemia Differential diagnosis • Giant cell arteritis (GCA) GCA and PMR frequently co-exist, cranial symptoms including headache, jaw claudication, and vision symptoms are typically absent in patients with PMR. PMR typically has less prominent symptoms than GCA. Treatment • prednisolone e.g. 15mg/od - dramatic response Response to a moderate dose of steroids can be useful in confirming the diagnosis of PMR. The maximum dose of prednisolone should not exceed 20 mg once daily. Patients should report 70% improvement in symptoms within three to four weeks, and inflammatory markers should have normalised by this point.

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 Calcium and vitamin D supplementation should be initiated for all patients with PMR who are starting corticosteroid therapy. Bisphosphonates should be added for long term steroid therapy. The usual starting dose is 15 mg prednisolone per day. Patients should expect relief of symptoms within 24-72 hours. One of the best ‘tests’ for Polymyalgia Rheumatica (PMR) is how patients respond to corticosteroid therapy. Tapering Tapering should be guided by clinical response. The dose should be increased if symptoms are not well controlled within one week. The effective starting dose should be maintained for two to four weeks after the patient becomes asymptomatic. Generally, the daily dose can be lowered by 1.0-2.5 mg every two to four weeks to find the minimum dose needed to maintain symptom suppression. Once the patient is reduced to 10 mg per day, the daily dose can be tapered by 1 mg every four weeks. Approximately 50-75% of patients can discontinue corticosteroid therapy after two years of treatment. Methotrexate and azathioprine If symptoms relapsed when the dose of prednisolone has been reduced below the current dose, Continue the current dose of prednisolone and start methotrexate used in patients with corticosteroid intolerance or as corticosteroid-sparing agents. These are generally reserved for patients in whom it has been difficult to reduce the prednisolone after prolonged high dosages (for example, 10 mg or more per day for more than a year). These agents should be added to the prednisolone initially, but with a view to slowly reduce and withdraw prednisolone. As with steroid therapy, azathioprine or methotrexate can be discontinued if there has been sufficient response. Prognosis • Rapid improvement often occurs within 24 to 72 hours with low-dose prednisolone.

Temporal arteritis (Giant cell arteritis (GCA) Overview • also known as giant cell arteritis (GCA). • Temporal arteritis is large vessel vasculitis • overlaps with polymyalgia rheumatica (PMR). • Histology shows changes which characteristically 'skips' certain sections of affected artery whilst damaging others. • It is a clinical emergency. GCA should always be considered in elderly patients with headaches, ocular symptoms (e.g. acute monocular visual loss), systemic symptoms and high ESR. Suspected GCA glucocorticoids immediately, even before diagnostic evaluation by temporal artery biopsy is complete.

Rheumatology

Epidemiology • Sex: ♀ > ♂ • Peak incidence: 70–79 years; rarely seen in patients < 50 years Features • typically, patient > 60 years old The greatest risk factor for (GCA) is aging. almost never occurs before age 50 • usually rapid onset (e.g. < 1 month) • headache (found in 85%) • jaw claudication (65%) is a very specific sign for temporal arteritis. • visual disturbances (50%) secondary to anterior ischemic optic neuropathy 15-20% of patients develop permanent visual loss. • tender, palpable temporal artery • features of PMR: aching, morning stiffness in proximal limb muscles (not weakness) • also, lethargy, depression, low-grade fever, anorexia, night sweats • Large vessel GCA : Subclinical involvement of the aorta and large arteries is frequent, a clinical consequence of which can be aortic aneurysm (in 10 to 20 % of cases). Investigations • Raised inflammatory markers: ESR > 50 mm/hr (note ESR < 30 in 10% of patients). CRP may also be elevated ESR can be within normal range in 5-10% of GCA cases. • Temporal artery biopsy: the definitive diagnostic test skip lesions may be present (certain sections of affected artery whilst damaging others) An adequate length of temporal artery (3 to 5 cm) should be obtained because inflammatory lesions may be present in a segmental fashion. A negative temporal artery biopsy can occur in up to 50% of patients, often because the sampled region was not involved in the pathologic process. Therefore, it is not sensitive enough to rule out temporal arteritis. Treatment should not be delayed while waiting for the biopsy to be performed. • Note: creatine kinase and EMG normal Diagnosis • The American College of Rheumatology 1990 criteria requires 3 of the following for GCA diagnosis:

1. Age >50 y/o
2. New onset localised headache
3. Temporal artery tenderness or decreased pulsation
4. ESR >50mm/hr
5. Temporal artery biopsy positive Treatment • High-dose prednisolone there should be a dramatic response, if not the diagnosis should be reconsidered Current BSR guidelines recommend: Uncomplicated GCA (no jaw or tongue claudication, or visual symptoms) prednisolone 40-60 mg daily Complicated GCA: (with visual involvement and/or jaw/tongue claudication)

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 Evolving visual loss or history of amaurosis fugax: IV methylprednisolone 500 mg-1 g daily for three days, followed by oral corticosteroids Established visual loss: at least 60 mg prednisolone daily • Urgent ophthalmology review. Patients with visual symptoms should be seen the same-day by an ophthalmologist. Visual damage is often irreversible • As GCA requires long-term steroid therapy bone sparing agents (a bisphosphonate and vitamin D) and a gastroprotective drug (e.g omeprazole) should be prescribed. • Also, low dose aspirin should be considered as it has been shown to reduce the rate of visual loss and cerebrovascular accidents in GCA.

Polyarthritis Differential diagnosis • rheumatoid arthritis • SLE • seronegative spondyloarthropathies • Henoch-Schonlein purpura • sarcoidosis • tuberculosis • pseudogout • viral infection: EBV, HIV, hepatitis, mumps, rubella

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Polyarteritis nodosa (PAN) Definition • systemic vasculitis of the medium-sized vessels, with necrotizing inflammation leading to aneurysm formation and tissue ischemia; • most commonly involving skin, peripheral nerves, muscles, joints, gastrointestinal tract, and kidneys . • any organ with the exception of the lung can be affected, Epidemiology • Peak incidence: ~45–65 years • Sex: ♂ > ♀ • more common in middle-aged men Pathophysiology • diffuse vascular inflammation and ischaemia of the affected organs. • PAN is a medium-vessel vasculitis that is a type III hypersensitivity reaction. Association • hepatitis B infection Features • Nonspecific symptoms: (found in 65% to 80% of patients) fever, malaise, arthralgia, weight loss • Neurological involvement: (in 55% of patients) polyneuropathy (mononeuritis multiplex), cerebral ischemia (stroke) • Skin involvement: (in 44%) skin rash, Skin ulcers, nodules livedo reticularis

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• Renal involvement : (in 11%) hypertension, Hypertension is a manifestation of renal ischaemia via activation of the reninangiotensin system. haematuria but red cell casts are absent because glomerular inflammation is not a feature. renal impairment • Coronary artery involvement ; increased risk of myocardial infarction • GI involvement: abdominal pain, nausea, vomiting can present with abdominal pain and melena due to involvement of the mesenteric arteries. • Testicular pain testicular pain from ischaemic orchitis is a characteristic feature uncommon presentation • Usually spares the lungs PAN should be considered in young adults presenting with stroke or myocardial infarction The diagnosis may be confirmed with a biopsy of involved tissue Livedo reticularis Diagnosis • The American College of Rheumatology (ACR) 1990 criteria Three of the following 10 criteria are required:

1. Weight loss ≥4 kg
2. Livedo reticularis
3. Testicular pain or tenderness
4. Myalgias, weakness, or leg tenderness
5. Mononeuropathy or polyneuropathy
6. Diastolic blood pressure >90 mmHg
7. Elevated urea or creatinine
8. Positivity for hepatitis B virus (HBV) infection
9. Arteriographic abnormality

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10.Biopsy of small- or medium-sized artery containing polymorphonuclear leukocytes. Investigations • Hepatitis B surface antigen is positive in 30%, • p-ANCA is positive only in 20%.

ANCA is classically negative in PAN. • Angiography: Conventional angiography is the imaging modality of choice, and should be performed if there is a clinical suspicion of PAN. typically demonstrates: microaneurysms and focal narrowing in medium-sized blood vessels. • Biopsy should be performed if angiography is not available or does not conclusively show a medium-vessel vasculitis. Shows: focal and segmental transmural necrotising inflammation with fibrinoid necrosis in medium-sized vessels. pleomorphic cellular infiltrate of lymphocytes, neutrophils, macrophages, and eosinophils. granulomas are absent. Differential diagnosis • PAN are differentiated from the other small- and medium-vessel vasculitides by: absence of anti-neutrophil cytoplasmic antibodies, Glomerulonephritis is not a feature of PAN, but it is common in anti-neutrophil cytoplasmic antibodies (ANCA) vasculitis. Making this distinction early by way of urinalysis for protein, blood, and casts is a simple first-line test that can guide further investigation and treatment. Red cell casts are absent in PAN If there is evidence of glomerular inflammation such as urinary casts, then an alternative diagnosis such as microscopic polyangiitis (MPA) or granulomatosis with polyangiitis (Wegener's) (GPA),must be considered. lung involvement is not seen in PAN, and abnormal respiratory findings should suggest an alternative diagnosis and by confirmation that small vessels (i.e., arterioles, capillaries, venules) are not involved. Treatment • idiopathic PAN corticosteroids and cyclophosphamide • hepatitis B related disease plasmapheresis and antiviral agents. • Azathioprine can be used as maintenance therapy, and typically has fewer side effects than cyclophosphamide. Cyclophosphamide causes premature ovarian failure and infertility in both men and women.

Granulomatosis with polyangiitis (Wegener's granulomatosis) Overview • Granulomatosis with polyangiitis is now the preferred term for Wegener's granulomatosis. • It is an autoimmune condition associated with a necrotizing granulomatous vasculitis, affecting both the upper and lower respiratory tract as well as the kidneys. • the classical triad consists of

1. necrotising granulomatous inflammation of the respiratory tract,
2. glomerulonephritis
3. small-vessel vasculitis. Features • upper respiratory tract: epistaxis, sinusitis, nasal crusting • saddle-shape nose deformity • lower respiratory tract: dyspnoea, haemoptysis migrating alveolar shadowing • rapidly progressive glomerulonephritis ('pauci-immune', 80% of patients) It usually presents with rapidly progressing renal failure (within three months), proteinuria and microscopic haematuria. • also: vasculitis (causing carotid artery tenderness) vasculitic rash, eye involvement (e.g. proptosis), cranial nerve lesions Investigations • c-ANCA (PR3-ANCA (targeting peroxidise-3) positive in > 90%, p-ANCA (MPO-ANCA (targeting myeloperoxidase) positive in 25% cANCA directed against proteinase-3 cANCA is highly specific, but is found in only 50% of patients with disease localised to the respiratory tract and 95% with generalised Wegener's.

 In active Wegener’s disease with renal involvement cANCA is highly sensitive and specific. After disease remission cANCA may remain elevated for years, and is not useful in evaluating patients for relapse. • chest x-ray: wide variety of presentations, including cavitating lesions • tissue biopsy renal biopsy: epithelial crescents in Bowman's capsule Kidneys show vasculitis and glomerulonephritis and occasional (NOT always) granulomata Lung biopsy has a high diagnostic yield show vasculitis and granulomas Biopsy of the upper respiratory tract shows granulomas but not vasculitis. Management • steroids Prednisolone is given in doses of around 1 mg/kg per day initially, after which the dose is reduced rapidly, typically at weekly intervals. In case of renal failure with indications for dialysis, the initial management Methylprednisolone Methylprednisolone should be given immediately, followed by haemodialysis and then cyclophosphamide. • cyclophosphamide (90% response) The combination of prednisolone and cyclophosphamide is now established as the standard therapy and the treatment of choice for induction of remission in Wegener's granulomatosis Cyclophosphamide: Traditionally, oral dose (2 mg/kg per day), but latterly intravenous boluses have proved increasingly popular, given in doses of 0.5-0.75 g/m2 body surface area at intervals of 2 weeks (at least for short periods) to 2 months. If a patient had a vasculitic neuropathy. Current practice is to use cyclophosphamide for induction therapy. • Both rituximab and methotrexate have also been used for induction therapy in ANCAassociated vasculitis, although they would not be first-line treatment. • Azathioprine is used as maintenance treatment following cyclophosphamide • ciclosporin is rarely used in the management of ANCA-associated vasculitis. • Evidence from controlled trials suggests that once remission is achieved azathioprine or methotrexate may be reasonable alternatives to cyclophosphamide. • In refractory Wegener's, both infliximab and rituximab have shown some degree of promise. • plasma exchange • in case of decreased conscious level with acute renal failure (with indication for dialysis) and respiratory function is failing. The first immediate step Endotracheal intubation and positive pressure ventilation, transfer the patient to a critical care setting (especially to protect airway with a GCS 8/15). Prognosis • median survival = 8-9 years

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Microscopic Polyangiitis • Microscopic polyangiitis is similar to wegener’s granulomatosis except in 3 things:

1. it only affects small blood vessels in the lungs or kidneys. No nasopharyngeal damage like wegener's
2. Associated with p-ANCA antibodies. anti-MPO (pANCA, 45%) antibody is strongly positive than anti-PR3 (cANCA, 30%)
3. No granuloma on biopsy

Churg-Strauss syndrome • Churg-Strauss syndrome is an ANCA associated small-medium vessel vasculitis. • also known as Eosinophilic granulomatosis with polyangiitis Features • asthma • paranasal sinusitis • mononeuritis multiplex • blood eosinophilia (e.g. > 10%) • Serum IgE is very commonly elevated and correlates with disease severity. • pANCA positive in 60% • Commonly associated with antimyeloperoxidase antibodies. • Non-fixed pulmonary infiltrates visible on chest radiographs • Rarely, it can cause ischaemic optic neuropathy, which presents with visual loss. Leukotriene receptor antagonists may precipitate the disease Diagnosis • It is diagnosed clinically, although a biopsy should be sought for pathological confirmation. • Skin biopsy reveals small-vessel arteriopathy with granuloma formation and is the diagnostic investigation of choice. Blood vessels with extravascular eosinophils on biopsy. PR3 antibody is associated with Wegener's granulomatosis, MPO antibody is associated with microscopic polyangiitis

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Treatment • High-dose methylprednisolone, with or without cyclophosphamide is the treatment of choice Prognosis • Without treatment, the 5-year survival rate for Churg-Strauss syndrome is around 25%; with appropriate therapy this rises to over 60%.

Idiopathic pulmonary haemosiderosis Definition • recurrent episodes of diffuse alveolar hemorrhage of unknown aetiology Prevalence • rare • tends to occur in younger people Features • pallor, • weakness, lethargy, • dry cough and occasional haemoptysis • no extrapulmonary features. • After recurrent episodes of hemorrhage, pulmonary fibrosis may develop due to iron accumulation. Investigations • no abnormal immunological features, which differentiates it from Goodpasture syndrome and wegener’s • Gas transfer is elevated as blood is already in the alveolar space. • chest radiograph and high resolution computed tomography demonstrate ground glass alveolar opacities that are often bilateral. • final diagnosis requires lung biopsy documentation of large numbers of hemosiderinladen macrophages in the alveoli, without evidence of vasculitis, capillaritis, inflammation, granulomas, or deposition of immunoglobulins in any specific pattern. Treatment • glucocorticoids +/- another immunosuppressive agent (eg, azathioprine, or cyclophosphamide) pulmonary hemorrhage without immunological features Idiopathic pulmonary haemosiderosis pulmonary hemorrhage + immunological features Goodpasture or wegener’s

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Henoch-Schonlein purpura Overview • Henoch-Schonlein purpura (HSP) is an IgA mediated small vessel vasculitis • involving mainly the blood vessels of the skin, GI tract, the kidneys and the joints. • 90% of cases of HSP occur in children aged 2-10 years but can occur in any age group. • In children, (HSP) is the most common cause of vasculitis affecting the kidneys. • typically commoner in males, • may follow an infectious agent. • It can present one to three days following infection of an IgA secreting mucous membrane (commonly following pharyngitis, but can occur following

infection of the gastrointestinal tract, bladder or breast). • An important risk factor in adults chronic alcohol intake. • associated with: Helicobacter pylori, hepatitis B and malignancy. Features HSP is characterised by the tetrad of: • purpura • abdominal pain • arthritis, and • renal involvement (haematuria and proteinuria). Patients with proteinuria have a worse prognosis than patients with haematuria alone. • palpable purpuric rash (with localized oedema) over buttocks and extensor surfaces of arms and legs (due to a cutaneous vasculitis) • abdominal pain (due to gut vasculitis, which may be severe in some cases, leading to bloody diarrhoea) • polyarthritis (common symptom) • features of IgA nephropathy may occur e.g. haematuria, renal failure HSP nephritis becomes clinically manifest in only 20-30%. It usually presents as macroscopic haematuria and proteinuria Of those patients with renal involvement, as many as 10% may develop chronic renal failure and end-stage renal disease. However, fewer than 1% of all patients with HSP suffer this poor prognosis. Diagnosis • Skin biopsy and immunofluorescence demonstrate leukocytoclastic vasculitis with IgA deposition,(meaning lots of white blood cells in the skin around small blood vessels) which is pathognomonic for HSP. Immunofluorescence studies will reveal IgA deposits within blood vessel walls Treatment • analgesia for arthralgia • treatment of nephropathy is generally supportive. All patients with hypertension and proteinuria (greater than 1 g/day) should be started on an angiotensin-converting enzyme (ACE) inhibitor, which may control the BP and proteinuria. Once the BP has been controlled, patient should have a renal biopsy, and if this showed changes of a crescentic glomerulonephritis (GN), then an immunosuppression regime similar to that used in renal vasculitis should be started (probably with high dose steroids in the first instance +/- cyclophosphamide). There is inconsistent evidence for the use of steroids and immunosuppressants Management of HSP in adults often involves the use of immunomodulatory or immune-suppressive regiments (in contrast to children where the majority of cases resolve spontaneously).

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Prognosis • usually excellent, HSP is a self-limiting condition, especially in children without renal involvement • There is often a more complicated course in adults, and 50% of patients who present with renal involvement develop renal insufficiency. • around 1/3rd of patients have a relapse MRCPUK-part-1-September 2019 exam: What is the most likely renal outcome in HenochSchonlein purpura? Full renal recovery

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Kawasaki disease Overview • Kawasaki disease is a type of vasculitis which is predominately seen in children. • Whilst Kawasaki disease is uncommon it is important to recognise as it may cause potentially serious complications, including coronary artery aneurysms Features • high-grade fever which lasts for > 5 days. Fever is characteristically resistant to antipyretics • conjunctival injection • bright red, cracked lips • strawberry tongue • cervical lymphadenopathy • red palms of the hands and the soles of the feet which later peel Diagnosis • Kawasaki disease is a clinical diagnosis

as there is no specific diagnostic test Management • high-dose aspirin Kawasaki disease is one of the few indications for the use of aspirin in children. Due to the risk of Reye's syndrome aspirin is normally contraindicated in children. • intravenous immunoglobulin Combination therapy with intravenous immunoglobulin (IVIG) and aspirin during the acute phase of Kawasaki disease produces a more marked antiinflammatory effect and reduction in coronary artery abnormalities than does aspirin alone. • echocardiogram (rather than angiography) is used as the initial screening test for coronary artery aneurysms Complications • coronary artery aneurysm (25% of cases) • lakayasu1s arter1 t1s

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Takayasu’s arteritis Definition • Chronic inflammatory granulomatous pan-arteritis of the major arteries It typically causes occlusion of the aorta (the ascending arch of the aorta) The subclavian artery is commonly affected, and subclavian steal syndrome may occur The brachial, radial and ulnar arteries can also be involved. Pathology • continuous or patchy granulomatous inflammatory process involving macrophages, lymphocytes, and multinucleated giant cells which causes progressive occlusive disease of the aorta and its branches. Epidemiology • most commonly affects women (the ratio of women to men is 8:1). • typical age onset of 25-30 years. • most common in Asia. Features • questions commonly refer to an absent limb pulse. • systemic features of a vasculitis e.g. malaise, headache • unequal blood pressure in the upper limbs • carotid bruit • vascular symptoms such as claudication. (intermittent claudication) • systemic symptoms of fever, arthralgia and weight loss. • neurological symptoms such as transient ischaemic attacks. • Cardiac features include angina, heart failure, and aortic regurgitation. • Renal manifestations may include mesangial proliferative glomerulonephritis. • aortic regurgitation (around 20%) • ESR and CRP are usually elevated, • levels of pentraxin 3 may be a useful marker of disease activity. Associations • renal artery stenosis Treatment • Corticosteroids with the addition of steroid sparing second agents such as methotrexate or azathioprine are the mainstay of therapy. Prognosis • With good care, 15-year survival rates approach 90%.

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Subclavian steal syndrome (SSS) The proximal part of left subclavian is blocked on left side so no flow in vertebral and to left arm. Blood from right vertebral enters left vertebral and flows back to supply left arm Etiology • Atherosclerosis • Cervical rib • Takayasu's arteritis Features • Presyncope (sensation that one is about to faint) • Syncope (fainting) • Neurologic deficits • Blood pressure differential between the arms • severe memory problems • hands showing circulation problems (hands can have blotchy patches of red and white) (associated with other stigma to vascular disease (e.g. vascular insufficiency ulcers of the foot).

Buerger’s disease Overview • Thromboangiitis obliterans (Buerger’s disease) is a disease of small and medium-sized arteries and veins resulting in inflammation and ulceration, in which the distal

vessels become blocked in the hands and feet. • There is no excessive atheroma and it does not involve the coronary arteries like atherosclerosis. • The disease occurs mainly in cigarette smokers; it has not been documented in nonsmokers. • Although there is florid histological inflammation within vessels, the disease is not a systemic vasculitis, is not accompanied by any elevation in acute phase markers and does not respond to immune suppression. Epidemiology • Prevalence is higher in men and people of Far Eastern origin. • seen in young (usually < 40 years) male smokers. Feature • symptoms of arterial ischaemia resulting in gangrene of the digits. claudication with diminished or absent pulses. The feet or legs may be cyanosed or dusky; the skin is thin and without hair. Ulcerations occur, and necrosis follows • Migratory phlebitis in the superficial vein is present in 40% of cases. Diagnosis • usually clinical. • Arteriogram will show occlusion of distal arteries of the hands and feet. • Histopathology examination of affected arteries reveals fresh inflammatory thrombus within both small and medium-sized arteries and veins, with giant cells surrounding the thrombus. Treatment • Supportive • stop smoking.

Prognosis • can be excellent (i.e. complete resolution of symptoms) with smoking cessation • in some cases, however, amputation is unavoidable

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IBD-associated arthropathy • The history of weight loss, diarrhoea and a mono/oligo-arthropathy suggests a diagnosis of inflammatory bowel disease (IBD). • IBD-associated arthropathy is considered a subtype of seronegative spondyloarthropathy. • A variety of joint involvement has been described, from large joint pauciarticular arthropathy to a rheumatoid pattern polyarthropathy. • Peripheral arthritis is generally non-erosive and the oligoarticular variant particularly may correlate with intestinal disease activity. • Axial arthritis may include inflammatory back pain, sacroilitis, or ankylosing spondylitis and is less likely to correlate with gastrointestinal symptoms. • mechanisms remain unclear. • Treatment of the gastrointestinal disease is not always sufficient for control of arthritis, and biologic agents may be indicated. The description of weight loss, diarrhoea and a mono/oligo-arthropathy suggests a diagnosis of inflammatory bowel disease. (IBD).

Differential diagnoses of arthropathies associated with iron deposition in the joints →brown-stained synovial fluid. • Haemophilia • Haemosiderosis from recurrent haemarthrosis • Haemochromatosis, and • Pigmented villonodular synovitis (PVNS).

SAPHO syndrome SAPHO is an acronym for synovitis, acne, pustulosis, hyperostosis, and osteitis. It is characterised by osteosclerotic bone lesions, sterile osteomyelitis, and a variety of skin lesions. • Synovitis - may be present rarely, and associates with erosions. • Acne - may be severe (conglobate or fulminans) and recur with new bony involvement. • Pustulosis - palmo-plantar pustulosis occurs in approximately 50% of patients, other skin lesions may include psoriasis, hidradenitis suppurativa, acne, and rarely Sweet's syndrome. • Hyperostosis (increase in bone substance) and osteitis (inflammation of the bones) - the bony lesions typically involve the acromioclavicular, and sternoclavicular joints. Other sites include anterior chest wall, sternum,

clavicle, pubic symphysis, spine, and mandible. These lesions are visualised on 99m technetium bone scan or MRI. The cause of the SAPHO syndrome is unknown. Investigation • skin lesions are characterised by neutrophilic pseudoabscesses. • Bone biopsy can reveal sterile osteomyelitis. Diagnosis should be suspected when there is an association of rheumatic pain with a pustular skin disease. treatment • no specific treatment, • some cases remit spontaneously

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• Typical treatment can be used for the arthritic symptoms (i.e. non-steroidal antiinflammatories and disease modifying anti-rheumatic agents). • Isotretinoin and aciretin can be used to treat the skin disease. • In the more severe cases corticosteroids, calcitonin, bisphosphonates and TNF-inhibitors can be used.

Elbow pain The table below details some of the characteristic features of conditions causing elbow pain: Lateral epicondylitis (tennis elbow) Features • pain and tenderness localised to the lateral epicondyle • pain worse on resisted wrist extension with the elbow extended or supination of the forearm with the elbow extended • episodes typically last between 6 months and 2 years. Patients tend to have acute pain for 6-12 weeks • most appropriate to gain short term relief for the patient? Local steroid/anaesthetic injection Medial epicondylitis (golfer's elbow) Features • pain and tenderness localised to the medial epicondyle • pain is aggravated by wrist flexion and pronation • symptoms may be accompanied by numbness / tingling in the 4th and 5th finger due to ulnar nerve involvement Radial tunnel syndrome • Most commonly due to compression of the posterior interosseous branch of the radial nerve. • It is thought to be a result of overuse. Features • symptoms are similar to lateral epicondylitis making it difficult to diagnose • however, the pain tends to be around 4-5 cm distal to the lateral epicondyle • symptoms may be worsened by extending the elbow and pronating the forearm Cubital tunnel syndrome Due to the compression of the ulnar nerve. Features • initially intermittent tingling in the 4th and 5th finger • may be worse when the elbow is resting on a firm surface or flexed for extended periods • later numbness in the 4th and 5th finger with associated weakness Olecranon bursitis Swelling over the posterior aspect of the elbow. There may be associated pain, warmth and erythema. It typically affects middle-aged male patients.

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Shoulder problems The table below summarises the key features of common shoulder problems: Condition Notes Adhesive capsulitis (frozen shoulder) Common in middle-age and diabetics Characterised by painful, stiff movement Limited movement in all directions, with loss of external rotation and abduction in about 50% of patients Supraspinatus tendonitis (Subacromial impingement, painful arc) Rotator cuff injury Painful arc of abduction between 60 and 120 degrees Tenderness over anterior acromion

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Prepatellar bursitis The most useful in initial diagnosis of prepatellar bursitis Crepitation of the knee

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Polymyositis Polymyositis is the commonest cause of inflammatory muscle disease in people under 50years-old (inclusion body myositis is the commonest in those over 50-years-old). Definition • Inflammatory disorder causing symmetrical, proximal, painless muscle weakness Pathophysiology • thought to be a T-cell mediated cytotoxic process directed against muscle fibres Epidemiology • Typically affects middle-aged • Female: male 3:1 Associated conditions • Connective tissue disorders • Interstitial lung disease → evaluate with chest x-ray and pulmonary function tests. • Malignancy , commonly Adenocarcinomas , stronger for dermatomyositis, than for polymyositis. The most appropriate next investigation CT chest, abdomen and pelvis

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Features • Proximal muscle weakness +/- tenderness • Raynaud's • Mechanics hands found in a subtype of polymyositis called anti-synthetase syndrome or Jo-1 syndrome →fissuring and cracking on the distal digital pads of several fingers. • Respiratory muscle weakness • Interstitial lung disease: e.g. fibrosing alveolitis or organising pneumonia • Dysphagia, dysphonia Investigations • Elevated creatine kinase (the initial investigation) • Electromyography (EMG): abnormal in almost all patients (90%). Triad of:

1. Short, small polyphasia motor units
2. Fibrillation and sharp waves
3. Bizarre, repetitive discharges
   • Muscle biopsy the definitive investigation to establish the diagnosis Histopathology endomysial mononuclear inflammatory infiltrate with CD8 T cells (MHC class I) and muscle fiber necrosis. • Anti-Jo-1 antibodies seen in pattern of disease associated with lung involvement, Raynaud's and fever • Antinuclear antibody - Positive in one third Treatment • Prednisolone is the mainstay of treatment, at an initial dose of 1 mg/kg/d. • In patients who fail to show improvement, disease-modifying steroid-sparing agents may be added. • A high-protein diet and supervised exercise may further improve symptoms. Prognosis • Most patients have a favourable response to corticosteroid therapy, and 5-year survival rates approach 80%.

Dermatomyositis Proximal weakness with normal reflexes and sensation and absence of fasciculations: without skin lesion →polymyositis with skin lesion → dermatomyositis Definition • Dermatomyositis is a variant of an inflammatory myositis causing symmetrical, proximal muscle weakness and characteristic skin lesions, for example a purple Heliotrope rash on the cheeks and eyelids or Gottron's papules: roughened red papules over extensor surfaces of fingers

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Pathophysiology • Autoantibodies binding to the vasculature, muscle atrophy, and lymphocytic inflammation • caused by CD4 T cells that cause perimysial inflammation and atrophy. Features • Features of polymyositis (proximal muscle weakness, Raynaud's, respiratory muscle weakness, interstitial lung disease, dysphagia, dysphonia) • Pathognomonic skin features Heliotrope rash in the periorbital region a violaceous or erythematous rash in a symmetrical distribution involving periorbital skin. its presence is highly suggestive of dermatomyositis . Gottron's papules: roughened red papules over extensor surfaces of fingers • Other skin lesions Photodistributed erythema, poikiloderma, nailfold changes Mechanic's hands: (rough, cracked skin) Fingers telangiectasia: Nail fold capillary dilatation. Shawl sign: macular rash over back and shoulder V-neck sig : Violaceous erythema or poikiloderma involving the anterior chest • ↑↑↑risk of malignancy Associated features • Malignancies (dermatomycotic increases the risk of malignancy more than polymyositis). typically lung cancer, found in 20-25% • Interstitial lung disease (ILD) occurs in at least 10% Image shows: Gottron’s papules Image shows: Heliotrope rash Investigations • Elevated creatine kinase → the most helpful initial test • EMG • Muscle biopsy high levels of the complement component C5b-9 around the capillary vessels. Perimysial inflammation with lymphocytic infiltrate • ANA positive in 60%

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• Anti-Mi-2 antibodies are highly specific for dermatomyositis, but are only seen in around 25% of patients • Screen for malignancy Management • Prednisolone • Glucocorticoid-sparing agents: azathioprine or methotrexate. Prognosis • Relatively good, with most patients reaching remission after 2–3 years, except of course where there is an associated underlying malignancy.

Inclusion body myositis (IBM) Definition • a syndrome of diffuse, progressive, asymmetric, proximal, and distal weakness that is generally refractory to immunosuppressive treatment. • The aetiology of IBM is largely unknown. Epidemiology • IBM occurs more frequently in men than women. • More common in older Caucasian males. • the most common acquired myopathy in patients older than 50 years Features • Muscle weakness can affect both proximal and distal muscles unlike polymyositis and dermatomyositis, asymmetry is common. characteristically early affects quadriceps and finger/wrist flexors are usually more severely The onset of muscle weakness in IBM is generally gradual (over months or years). • Dysphagia is common, occurring in 40-66% of patients • Difficulties with breathing →the most common cause of death is respiratory system disorders. Diagnosis • creatine kinase (CK) levels: no striking elevation (less than 10 times normal) • anti-cN1A autoantibodies • Muscle biopsy shows intranuclear or cytoplasmic tubofilaments on electron microscopy. The specific finding is the presence of sarcoplasmic “rimmed” vacuoles Treatment • Optimal treatment for IBM is not known • In contrast to dermatomyositis and polymyositis, IBM is relatively resistant to standard immunomodulatory therapies.

Polymyositis Dermatomyositis IBM Onset Subacute Subacute Slow Commonest < 50 years commonest > 50 years age Commonest < 50 years Affected muscles Proximal Proximal Proximal

and distal symmetry symmetrical symmetrical Asymmetrical Common incidence Female Female Male Skin lesion NO Characteristic rash NO CK Highly elevated (up to 50 fold) Highly elevated (up to 50 fold) antibodies anti-Jo-1 are more common anti-Mi-2 are highly specific anti-cN1A autoantibodies Muscle biopsy endomysial mononuclear inflammatory infiltrate and muscle fiber necrosis. perivascular and interfascicular inflammatory infiltrates with adjoining groups of muscle fiber degeneration/regeneration T cell CD8 T cell CD4 T cell good Good Poor Response to steroids Painless weakness and wasting with selective involvement of long finger flexors and quadriceps is characteristic of inclusion body myositis. Inclusion body myositis occurs in older people, has an insidious onset, and does not associate with striking elevations in CK. Notes & Notes for MRCP
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Mild elevated (up to 10 fold) or normal intranuclear or cytoplasmic tubofilaments

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Fibromyalgia (FM) Definition • Fibromyalgia is a syndrome characterised by widespread pain throughout the body with tender points at specific anatomical sites. Epidemiology • Prevalence: occur in 1 - 2% of the general population • Gender: women are 10 times more likely to be affected • Age: typically presents between 30-50 years old Features • general symptoms: lethargy • musculoskeletal symptoms: chronic pain: at multiple site, sometimes 'pain all over' allodynia: (pain in response to non-painful stimuli) Morning fatigue morning stiffness tissue swelling, • neurological and psychiatric symptoms: sleep disturbance, headaches, dizziness are common patients often look unwell and may appear depressed and anxious. • GIT symptoms: 50% of patients with fibromyalgia complain of diarrhoea and constipation, often associated with abdominal bloating. Diagnosis • The diagnosis of FM should be considered in any patient with ˃three months of widespread, multisite pain without apparent causative found. • is clinical pain in all four quadrants of the body, as well as tenderness in 11 of 18 anatomically defined trigger areas. • The normal ESR in patients with FM contrasts with the high ESR in elderly patients with polymyalgia rheumatica. • Other causes of fatigue should be excluded e.g. hypothyroidism, anaemia and other rheumatological diseases Management • explanation • aerobic exercise: has the strongest evidence base • cognitive behavioural therapy • medication: pregabalin, duloxetine, amitriptyline Key facts: • How to diagnose? A female, presented with a feature of pain and tenderness over multiple area + normal ESR and CRP. • What is the best management? aerobic exercise

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Dupuytren’s contracture Definition • progressive painless contracture of the palmar facial bands, causing flexion deformities of the fingers. • autosomal-dominant condition with variable penetrance. Prevalence • has a male: female predominance of 10:1. • prevalence rates approaching 25% in elderly Scandinavians. • most commonly observed in persons of Northern European descent and affects 4-6% of Caucasians worldwide. Pathophysiology • fibroblast proliferation, and collagen deposition leading to contractures of the palmar fascia. • Interleukin 1

(IL-1) is the most abundant cytokine • Normal palmar fascia is primarily composed of type I collagen; Dupuytren disease is associated with an increase in type III collagen. Risk factor • Alcoholism (10%), • diabetes mellitus (8%). • previous myocardial infarction, • hand trauma, • HIV infection, • cigarette smoking. Features • bilateral in 45%; • in unilateral cases, the right side is more often affected. • The ring finger is most commonly involved, followed by the fifth digit and then the middle finger. The index finger and the thumb are typically spared. • Penile fibromatosis (Peyronie's disease) is seen in about 7-10% of patients. Rheumatoid arthritis seems to protect against the development of Dupuytren disease. Management • Surgery followed by physiotherapy to improve finger function is the recommended course of action. • Collagenase therapy may be an alternative to surgery in some cases.