Acute rheumatology

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DIAGNOSTIC & TREATMENT DILEMMAS

• GCA & PMR
• Monoarthritis
• Polyarthritis
• Multi-system disease
A 73 year old woman presents with a 10 week history of pain affecting the cervical spine, both shoulders, lumbar spine and both hips. Early morning stiffness lasts until lunchtime and she feels tired. She has a low grade pyrexia of 37.4°C, bilateral knee effusions and right carpal tunnel syndrome.

Investigations reveal:
Hb 10.1 g/dl MCV 85, Plt’s 480, WBC 14, ESR 81mm/ hour, CRP 27 mg/l
Bilirubin 5, ALP 180, ALT 38, albumin 32
Rheumatoid factor negative, ANA negative
Serum immunoglobulins and protein electrophoresis shows polyclonal increase in γglobulins

How will you manage this patient:
A Prednisolone 20mg OD
B Refer to Rheumatology as OPA
C Prednisolone 60mg OD
D IV Co-amoxiclav
E CT- chest, abdomen & pelvis
Case 2

A man aged 70 years is admitted to MAU with increasing pain and stiffness affecting the back, shoulders and hips. He was diagnosed with polymyalgia rheumatica 4 weeks ago. Currently taking Prednisolone 15mg od. ESR 50, CRP<5. Physical examination is entirely normal.

What will you do next?
A. increase prednisolone to 60mg OD & discharge with Rheum OPA
B. request in-patient rheumatology opinion
C. X-rays lumbar spine
D. Calcium levels
E. CT-chest, abdomen and pelvis
• Final diagnosis:
• Positive for BJP on urine electrophoresis
• Multiple myeloma
Polymyalgia rheumatica

- Inflammatory syndrome with ‘proximal limb pain and stiffness’
- Overlap with giant cell arteritis
- Very rare under age 50
- Common - managed in primary care
- Common indication for long term steroid use
‘Stepwise approach to diagnosis’

• **Diagnosis**
  – Age > 50
  – Bilateral shoulder and/or pelvic girdle pain
  – Morning stiffness > 45 minutes
  – Abrupt onset and duration > 2 weeks
  – Acute phase response (CRP and ESR)
  – Good response to low dose steroid (prednisolone 15-20mg, within 1 week)

• **A diagnosis of exclusion**
  – Infection, cancer, other inflammatory disease - (myeloma)
  – Non inflammatory disease eg. Rotator cuff disease
  – Neurological and endocrine disease

*Edited from BSR guidelines 2009*
Differential diagnosis

- **Inflammatory diseases**
  - RA, seronegative arthropyathy and ankylosing spondylitis, connective tissue diseases, other vasculitis (+/- ANCA)

- **Non-inflammatory**
  - Local shoulder/hip pathology, degenerative spinal disease
  - Fibromyalgia and pain syndromes

- **Neoplasia**
  - Lymphoma, leukemia, Myeloma, solid malignancy

- **Neurological disease- including Parkinsons**

- **Endocrinopathy – including vitamin D deficiency**

- **Chronic infection- TB**

*Michet et al. BMJ 2008*
This 78 year old man has a two week history of pain in the jaw and tongue while eating and a constant headache.

What is the immediate management?

A. temporal artery biopsy
B. prednisolone 30mg once daily
C. prednisolone 60mg once daily
D. temporal artery doppler study
E. cranial arteriography
Giant cell arteritis

- The most common vasculitis
- Large vessel granulomatous vasculitis with a predilection for the extracranial vessels
- May present with visual loss, myocardial infarction and stroke

Classification criteria:
- Age>50 (99%)
- New headache (74%)
- Pain on palpation (64%)
- ESR >50 (85%)
- Histology 85%
- Jaw claudication (37%)
- Eye involvement (32%)
- Large vessel involvement (17%)

Hunder et al 1990
2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides

Arthritis & Rheumatism

Volume 65, Issue 1, pages 1-11, 27 DEC 2012 DOI: 10.1002/art.37715
http://onlinelibrary.wiley.com/doi/10.1002/art.37715/full#fig2
Giant cell arteritis

Large vessel granulomatous vasculitis with a predilection for the extra-cranial vessels

Polymyalgia rheumatica

Cranial GCA

Large vessel GCA
Challenges

1. Visual loss and ischemic optic neuropathy in 18-24%
   - Absence of typical symptoms
   - Near normal inflammatory markers

• Significant diagnostic delay for giant cell arteritis
  • Mean of 8 weeks (7.7-17.6 weeks)  
    *Prior JA et al.*

2. Diagnosis
   - tricky for the generalist and inexperienced clinician
   - reliance on inflammatory markers ESR> CRP
   - flawed ethnic demographic data
   - temporal artery biopsy- ‘hassle’ to arrange and potential technical flaws
   - Ultrasound and CT/PET useful but not readily accessible
Thickened temporal arteries
Absent pulsation

Scalp necrosis
Ophthalmic manifestations

Visual disturbance in 6-50% of cases

Transient monocular visual loss (amaurosis fugax)

Complete visual loss (irreversible)
  anterior ischemic optic neuropathy
  central retinal artery occlusion

Visual field defects
<table>
<thead>
<tr>
<th>Jan</th>
<th>Monday</th>
<th>Tuesday</th>
<th>Wednesday</th>
<th>Thursday</th>
<th>Friday</th>
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<td>2</td>
<td>3</td>
<td>4</td>
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<td>6</td>
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</table>

**Notes:**
- **Jan 10:** U.O.-Head; Jaw pain; Dr. Davis; jaw pain.
- **Jan 17:** Normal vision in side bit of head.
- **Jan 24:** U.O.-Head; jaw pain; Dr. Davis; jaw pain.
- **Feb 7:** U.O.-Head; jaw pain; Dr. Davis; jaw pain.
Inflammatory markers in GCA
ESR/CRP Distribution in patients with and without GCA
ESR/CRP in patients with biopsy confirmed GCA
**Investigations**

- **Inflammatory markers** - ESR >50mm/hr, raised CRP

- **FBC** - anaemia, thrombocytosis, WBC usually normal

- **Deranged LFT’s** - Raised ALP, ALT/AST

- **Temporal artery biopsy** - Recommended by guidelines, positive up to 4 weeks after commencing steroids

- **Other investigations**:  
  - Ultrasound, MRA, CT-PET
<table>
<thead>
<tr>
<th>Diagnostic test</th>
<th>TA biopsy</th>
<th>FDG-PET/CT</th>
<th>MRA (multi contrast HR of vessel wall)</th>
<th>color duplex ultrasound</th>
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<tbody>
<tr>
<td>Sensitivity/specificity (percent)</td>
<td>40/100</td>
<td>83/90</td>
<td>89/75</td>
<td>67-100/95 Temporal, Common Carotid and axillary arteries</td>
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<tr>
<td>Safety</td>
<td>Invasive, facial n. injury, infection</td>
<td>radiation exposure</td>
<td>contrast</td>
<td>N/A</td>
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<tr>
<td>Assessing disease activity</td>
<td>contralateral biopsy for recurrence?</td>
<td>in remission may remain positive</td>
<td>unknown</td>
<td>under study</td>
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<td>Cost</td>
<td>$541</td>
<td>$5,185</td>
<td>$7,348</td>
<td>$388</td>
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<tr>
<td>Limitations</td>
<td>does not assess extra-temporal vessels</td>
<td>cannot assess temporal artery</td>
<td>limitations for MRI in general</td>
<td>aorta</td>
</tr>
</tbody>
</table>
The patient with weight loss, fatigue and the unexplained inflammatory response
Rapid access pathways in GCA

• Fast track pathway reduces sight loss in giant cell arteritis: results of a longitudinal observational cohort study.

  Patil P and Dasgupta et al. Clinical & Experimental Rheumatology 2015
Audit 2007-2009

54 suspected cases
21 diagnosed GCA
13 biopsy positive

Only 39% given steroids
24% presented with vision loss

GIANT CELL ARTERITIS PATHWAY

Consider GCA in patients > 50 years old presenting with the following features:
- Abrupt-onset headache (usually unilateral in the temporal area)
- Scalp tenderness with or without abnormal superficial temporal artery
- Jaw, tongue or limb claudication
- Visual symptoms (including diplopia, amaurosis fugax or visual loss)
- Polymyalgia symptoms

Is patient younger than 50 years old?

YES
DISCUSS WITH RHEUM OR MEDICAL SPR/CONSULTANT
Consider alternative diagnoses:
- Migraine
- Sinusitis
- Facial pain
- TMJ disease
- Dental problems

NO

Is visual disturbance/loss or neurological problems?

YES
ADMIT
Initiate treatment after discussion with responsible Consultant
- IV methylprednisolone (750-1000mg IV)
- Aspirin 75mg + PPI
- Refer Rheumatology & Ophthalmology

NO

SUSPECTED GCA – SUITABLE FOR OUTPATIENT MANAGEMENT

1. Check FBC, U&Es, LFTs, bone profile, clotting, ESR & CRP, G&S
2. Discuss all cases with Rheumatology SpR (Mob 07825 014746 or B lp 387, 388, 393) or Consultant In hours
3. Commence Treatment with steroid + aspirin 75mg and PPI cover
   a. Prednisolone 40mg daily if no visual symptoms or jaw claudication
   b. Prednisolone 60mg daily if jaw claudication but no visual symptoms
   c. Admit if visual symptoms or focal neurology!

1. ALL PATIENTS MUST BE REFERRED TO RHEUMATOLOGY USING ICE
2. REFERRAL PROFORMA FOR BIOPSY MUST BE FILLED & FAXED (PAGE 4)
   A temporal artery biopsy will be arranged within the next 2 weeks, with a rheumatology clinic appointment to follow for results and further treatment.
3. Provide GCA and temporal artery biopsy information leaflet to patient (see pages 2-3 of this document)

Giant cell arteritis pathway: August 2017
Differential diagnosis

- Intracranial infections
  - Sinusitis
  - Intracranial/ dental abscess
- Trigeminal neuralgia
- Neoplasia- intracranial and extracranial
- Cervical spondylosis/ occipital neuralgia
- Polyarteritis nodosa
Case 4

A patient with well-controlled Rheumatoid arthritis on methotrexate 20mg weekly, sulphasalazine 1g bd and prednisolone 5mg OD presents with a painful, swollen, right knee on a Friday evening. He cannot move the knee. There is no history of trauma.

Temp 37.4°C, WCC 10, CRP 53, ESR 80.

What do you do?

A  Analgesia and expedite Rheum OP r/v  
B  Increase prednisolone 20mg and expedite Rheum OP r/v  
C  Increase prednisolone 20mg, give antibiotics and expedite Rheum OP r/v  
D  Aspirate joint and give antibiotics  
E  Aspirate joint, increase prednisolone 20mg, expedite Rheum OP r/v
Septic Arthritis

Prompt diagnosis
Aspirate joint and give antibiotics

- Knee 55%
- **Polyarticular 12%**
- Hip 11%
- Ankle 8%
- Shoulder 8%
- Wrist 7%
- Elbow 6%
- Others 5%

<table>
<thead>
<tr>
<th>Gram Positive Cocci</th>
<th>Adults (%)</th>
<th>Children (%)</th>
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<tr>
<td>S. aureus</td>
<td>35</td>
<td>27</td>
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<tr>
<td>S. pyogenes, S. pneumoniae, S. viridans Group</td>
<td>10</td>
<td>16</td>
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<table>
<thead>
<tr>
<th>Gram Negative Cocci</th>
<th>Adults (%)</th>
<th>Children (%)</th>
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<tbody>
<tr>
<td>N. gonorrhoeae and meningitidis</td>
<td>50</td>
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<td>H. influenzae</td>
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<thead>
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<th>Gram Negative Bacilli</th>
<th>Adults (%)</th>
<th>Children (%)</th>
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</thead>
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<tr>
<td>E. coli, Salmonella and Pseudomonas species</td>
<td>5</td>
<td>9</td>
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<tr>
<td>Mycobacteria and Fungi</td>
<td>&lt;1</td>
<td>&lt;1</td>
</tr>
</tbody>
</table>
Who should aspirate?

- Knee:
  - Normal joint
  - Prosthetic joint
    - YOU!!!
    - Orthopaedics

- Hip:
  - Orthopaedics / Radiology

- Shoulder, elbow, wrist, ankle:
  - Rheumatology / Orthopaedics / Radiology
  - Gram stain, MCS
  - Crystals
  - AFB
Predisposing factors

10% of patients with an acutely painful joint have a septic arthritis

- Age >80 years
- Diabetes mellitus
- Rheumatoid arthritis
- Presence of prosthetic joint
- Recent joint surgery
- Skin infection
- Intravenous drug abuse, alcoholism
- Prior intra-articular corticosteroid injection
- Indwelling catheters
- Immunocompromised eg. HIV

- More than one predisposing factor amplifies/ augments risk.
Points on septic arthritis

- Haematogenous v’s local spread
- Presenting feature of endocarditis
- 80% will have fever, joint pain and swelling
- Polyarticular presentation 15%
- Blood cultures positive in 50%

- Treatment
  - Antibiotics 2 weeks intravenous, 2 weeks oral
  - Surgical washout/ aspiration- no evidence base

- Outcomes
  - Mortality 10-15% (50% if polyarticular, staph aureus)
  - Functional outcomes
Hot Swollen Joint Pathway

Ensure Adequate Analgesia

Non-Traumatic, acute, hot, swollen joint with restricted movements?

Consider septic arthritis

Prosthetic Joint?

No

Yes

REFER ORTHO IMMEDIATELY

Exit

History should include (circle):
- On anticoagulants? - check INR
- Diabetes? - check BM
- Previous Joint Pathology?
- Any Hx of trauma?
- Hx TB/ TB contacts?
- Sickle Cell Disease?
- Genitourinary symptoms?
- Systemic source of infection, chest, urine

Aspirate Joint
(if prosthetic joint or anti-coagulated do not aspirate joint- seek senior review)

Can ask ortho on-call for help with aspiration

Take blood cultures
- FBC, U&E’s, LFT’s, ESR, CRP, Clotting screen, Urate

Start Antibiotics:
- Benzylpenicillin 1.2g IV QDS and Fluclucocillin 1g IV QDS
- Or if Gonococcal disease suspected: Fluclucocillin 1g IV QDS and Ceftriaxone 2g IV OD
- If penicillin allergic: Refer to Guideline/Discuss with ID/Micro

X-ray relevant joint

Refer to Infectious diseases for further care- Bleep 506 in-hours and 007 overnight

Any evidence of Sepsis/ SIRS?

Start Sepsis Bundle

All steps completed

Time:  

Signed:
A 58 year old man presents to A&E at 3am with sudden onset right wrist pain with swelling.
He is unable to use his wrist.
Hypertensive and taking bendrofluazide
Apyrexial, heart rate 100 per minute
Examination- very tender, hot, red, swollen wrist
Wbc- 11.3, neutrophil 8.2 CRP 105, Cr 105 (no known CKD)
Uric acid 380 (normal 200-450)

• What will you do next
  A aspirate the wrist
  B commence intravenous flucloxacillin
  C commence diclofenac and iv flucloxacillin
  D commence colchicine and flucloxacillin
  E orthopaedic review for aspiration
A 58 yo male presents to A&E with a 3/7 hx of severe pain in both ankles and right 1\textsuperscript{st} MTP after gastroenteritis (D&V). He has a background of CKD (baseline creatinine 110) and gout. He is on allopurinol 100mg. On examination he has pain and swelling of both ankles and right 1\textsuperscript{st} MTP.

BP is 110/90, P70, T37.4
Creatinine 150, CRP 20, urate normal

What is your management?

A Stop allopurinol, give antibiotics and fluids
B Stop allopurinol, give antibiotics and NSAIDs
C Continue allopurinol, give naproxen and fluids
D Continue allopurinol, give colchicine and fluids
E Continue allopurinol, give colchicine, antibiotics and fluids.
Gout

- Mono > polyarticular. 1\textsuperscript{st} MTP > ankle, knee
- Risk Factors:
  - Trauma, diuretics, Etoh, infection, surgery, dehydration
- Investigations:
  - Urate normal in 40% acute gout, erosions

Management
- Don’t stop allopurinol if already on it & don’t start allopurinol during attack
- NSAIDS + PPI
- If NSAIDS contraindicated colchicine 500mcg tds (bd renal dose)
- If NSAIDS / colchicine both contraindicated – steroids
  - 20-30mg prednisolone od or 120mg IM depomedrone

Allopurinol
- >2 attacks, tophi, erosions, urate stones/nephropathy
- Colchicine/NSAID cover for 3/12, whilst ↑ allopurinol to target urate <0.3
A 78 year old man presents with a swollen left wrist with limited movements. It is warm and very tender on palpation. WBC 11, CRP 250

Investigations?
Diagnosis?
Crystal arthropathy- Treatment

• Acute
  – Analgesia
  – NSAIDs- Naproxen, diclofenac, celecoxib/ etoricoxib
  – Colchicine
  – Prednisolone, steroid injections

• Long term - Gout
  – Allopurinol or Febuxostat, aim urate <300
  – Do not start in acute phase
  – Dietary measures
  – Withdraw precipitating drugs if possible
Case 7

- A 42 year old lady is admitted with painful, swollen joints affecting the hands, knees and elbows. Symptoms started 2 weeks ago. She has 45 minutes of stiffness each morning. Examination reveals swollen and tender MCP joints affecting the right hand, a swollen left elbow and right knee.
- Temperature 37.8
- Blood tests show: CRP 80, ESR 50, WBC 14, Hb 10.4, Plts 450

What will you do next:

A Septic screen including blood cultures
B Commence prednisolone 40mg OD and discharge with Rheum OPA
C Intramuscular depomedrone 120mg and discharge with Rheum OPA
D ASOT, hepatitis and HIV serology
E prednisolone 20mg OD and discharge with Rheum OPA
Polyarthritis- Differential Diagnosis

- Inflammatory
  - Rheumatoid, reactive arthritis, psoriatic and SpA, Sarcoid

- Infective/Post infective:
  - Viral - hepatitis B, C, HIV, dengue/chikungunya
  - Bacterial - septicaemia, endocarditis, post-strep
  - Other - Lyme, syphilis, TB (Poncets)

- Neoplasia, haematological malignancy/ Paraneoplastic
An approach to Polyarthritis (>5 joints)

History & Examination
Acute v’s Chronic (6 weeks)
Morning stiffness > 1 hour
Joint line tenderness, swollen and tender joints
Skin rash

Investigations
Routine including CRP and ESR
Chest x ray and consider x-rays hand and feet
Infection screen - Blood, urine cultures, ASOT, Hep B, C, HIV
Auto-antibodies - ANA, RhF, anti-CCP
Case 8

A 24 year old student is admitted with a 1 week history of fatigue, fevers and multiple joint pains. She has a rash on her trunk which ‘comes and goes’ throughout the day and has an intermittent sore throat

Swollen, tender knees and wrists

Temperature 39.

WBC 17, neutrophilia, CRP 300, ESR 90, ALT 300, Bil 15, ALP 230

- **Diagnostic investigation:**
  - A Septic screen including blood cultures
  - B RhF, ANA and anti-CCP
  - C Ferritin
  - D ASOT, hepatitis and HIV serology
  - E Liver ultrasound
Adult onset Still’s Disease

A rare systemic inflammatory disease characterized by the classic triad of persistent high spiking fevers, joint pains, and a distinctive salmon-coloured rash

**Yamaguchi criteria**

<table>
<thead>
<tr>
<th>Major criteria</th>
<th>Minor criteria</th>
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<tbody>
<tr>
<td>Fever of at least 39 °C for at least one week</td>
<td>Sore throat</td>
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<tr>
<td>Arthralgias or arthritis for at least two weeks</td>
<td>Lymphadenopathy</td>
</tr>
<tr>
<td>Nonpruritic salmon-colored rash (usually over trunk or extremities while febrile)</td>
<td>Hepatomegaly or splenomegaly</td>
</tr>
<tr>
<td>Leukocytosis (10,000/microL or greater), with granulocyte predominance</td>
<td>Abnormal liver function tests</td>
</tr>
<tr>
<td></td>
<td>Negative tests for antinuclear antibody and rheumatoid factor</td>
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Case 9

A 37 year old woman with SLE presents to A&E with a 2 day history of fever (38°C), sweats and a cough. She is on prednisolone 5mg OD, azathioprine 100mg OD and hydroxychloroquine 200mg BD.

- Pulse 130, BP 100/70, sats 95% RA. Investigations reveal: WCC 5.4 (neutrophils 2.5, lymphocytes 0.5), platelets 100. ESR 80 mm, CRP 36 mg/l,

*What is your management?*

A  Stop prednisolone and give antibiotics
B  Stop prednisolone, stop azathioprine, give antibiotics
C  Continue prednisolone, stop azathioprine and give antibiotics
D  Increase prednisolone, stop azathioprine and give antibiotics
E  Increase prednisolone, stop azathioprine, stop hydroxychloroquine and give antibiotics
SLE – don’t forget

- Nephritis
  - urine bld, protein
- Haematological
  - Haemolysis, ITP
- Antiphospholipid syndrome
  - PE, DVT, CVA, miscarriage
- Cardiac
  - MI, peri/myo carditis
- Neurological
  - Seizures, psychosis, CVA
SLE – disease activity markers

- Clinical – symptoms, signs
- Laboratory
  - Hb, NØ, LØ, platelets (all may decrease)
  - ESR (CRP)
  - Complement (decrease C4>C3)
  - dsDNA

↑ CRP in SLE
Infection…Infection…
Infection!
(severe arthritis & serositis)
Case 10

• A 26 year old lady is admitted to AAU at 1am with a 1 week history of shortness of breath and haemoptysis

• Over the last four weeks she has become increasingly tired and noticed blood in her urine

• She has painful wrists and several swollen joints

• 2 years ago she developed mouth ulcers, joint pains, pleuritic chest pains.

• She was seen in rheumatology 5 years ago for Raynauds disease

• Blood pressure 100/70, HR 80/minute, Oxygen saturations 98% on 2l
- Urine dipstick- 3+ blood, 3+ protein
- Hb 10.5g/dl WBC 7.4 Plt 254
- ESR 90 CRP<5
- Cr 70 eGFR 95
Case 11

What is your immediate management:

A broad spectrum antibiotics  
B iv methylprednisolone- 1g stat  
C wait until next morning, pending specialist review  
D CT chest  
E commence clarithromycin
What is the most likely diagnosis:
A Goodpastures
B SLE
C Wegener’s/ GPA
D Microscopic polyangiitis
E Legionella pneumonia
• ESR- 90mm/Hr, CRP- <5mg/ml.
• ANA > 1:640, Ro+, La+, Sm+, dsDNA>180
• C3 0.42 g/l, C4 0.04 g/l

• Urine protein: Cr ratio = 180mg/mmol
• SLE
  – Pulmonary alveolitis with haemorrhage
  – Class IV lupus nephritis

• Treatment
  – Pulse methylprednisolone (1g X 3)
  – Iv cyclophosphamide followed by MMF
  – Plasma exchange
Points on vasculitis

• Multi-system diseases defined by size of vessel involved
• CRP usually raised
• A negative ANCA test does not exclude a small vessel vasculitis (SVV)
• Raised PR3/ MPO titre must be taken seriously
• Always check urine dipstick and PCR in suspected SVV
Multi-system disease- differential

- Infection
  - Viral- Herpes/CMV, Hep B, C, HIV
  - Bacterial- sepsis, endocarditis
  - Fungal, atypical, TB
- Malignancy
  - Solid v’s haematological (lymphoma, myeloma)
- Auto-immune/ auto-inflammatory
  - Connective tissue diseases
  - Vasculitis
  - Rheumatoid and SpA
  - Auto-inflammatory
- Other
  - Atrial myxoma
Approach to the patient with multi-system disease

- History and full examination is key
- Urinalysis/ urine dipstick

Acute investigations
- FBC, U&E’s, LFT’s
- ESR (high) v’s CRP (normal in CTD, raised in vasculitis)
- C3 and C4, immunoglobulins
- CXR

Further investigations
- ANA, ENA, dsDNA, RhF, ANCA, anti-GBM, myeloma screen
- Blood cultures and septic screen, HIV, Hep B and C, ASOT
- Imaging – CT/ CT-PET
A 58 year old single man is admitted with painful legs and a rash