

Acute rheumatology

Shahir Hamdulay

Consultant Rheumatologist & General Physician

Northwick Park hospital

Shahir.hamdulay@nhs.net

14.6.2019

DIAGNOSTIC & TREATMENT DILEMMAS

- GCA & PMR
- Monoarthritis
- Polyarthritis
- Multi-system disease

Case 1

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 γ globulins

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

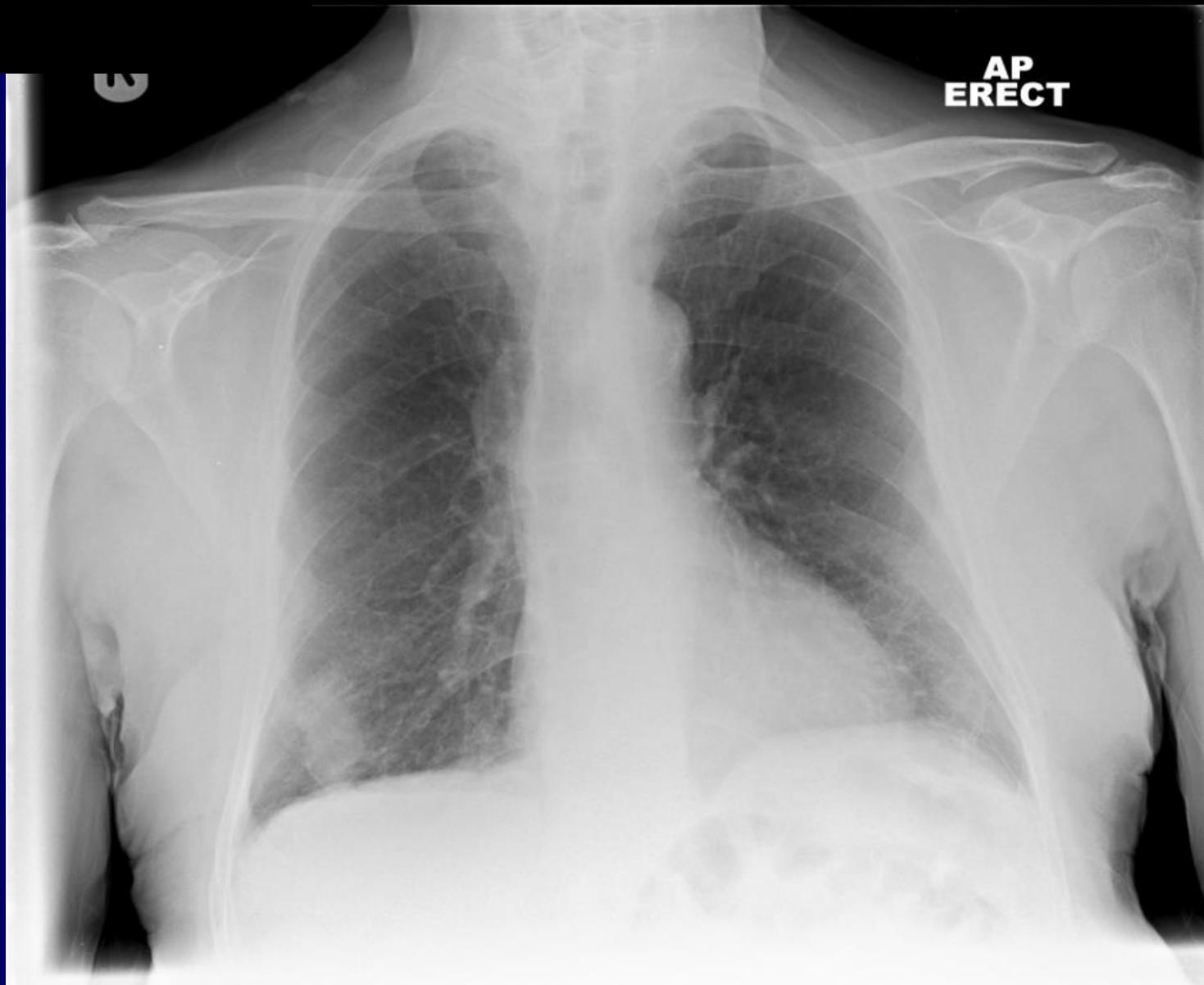


27/10/2010, 10:45:17

C: 2190.5, W: 3743.0

A2

C: 2077.0, W: 4036.0



25/10/2010, 13:18:28

A1

C: 40.0, W: 350.0



Contrast: CONTRAST

Gantry: 0°

FoV: 500 mm

Time: ms

Slice: 3 mm

Pos: -274

HFS



F: B

313 mA

120 kV

Image no: 154

Image 154 of 282

27/10/2010, 11:30:25



C: 991.0, W: 1982.0

RJ5001780775

Seq: SE

Slice: 3 mm

Pos: 6.63453

TR: 4460

TE: 112

AC: 2

R

P

HFS

FoV: 360 mm

Image no: 10

Image 10 of 21

28/10/2010, 10:28:14

F



C: 942.0, W: 1884.0

RJ5001780775



Seq: SE

Slice: 3 mm

Pos: 22.6222

TR: 4460

TE: 112

AC: 2



HFS

FoV: 360 mm

Image no: 14

Image 14 of 21

28/10/2010, 10:28:14

F

C: 972.0, W: 1945.0

RJ5001780775

R

Seq: SE

Slice: 3 mm

Pos: 38.61

TR: 4460

TE: 112

AC: 2

HFS

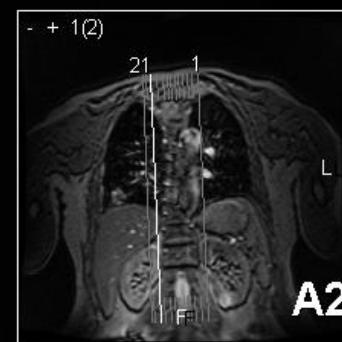
FoV: 360 mm

Image no: 18

Image 18 of 21

28/10/2010, 10:28:14

F



C: 1468.0, W: 2937.0

RJ5001780775



Seq: SE

Slice: 3 mm

Pos: 22.6222

TR: 600

TE: 9.728

AC: 2

P

HFS

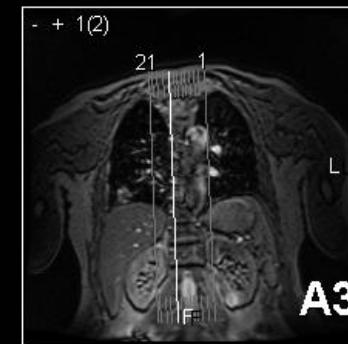
FoV: 360 mm

Image no: 14

Image 14 of 21

28/10/2010, 10:32:22

F



C: 439.0, W: 879.0

R33001700773



Seq: RM

Slice: 3 mm

Pos: 22.6222

TR: 5600

TE: 46.592

AC: 1

P

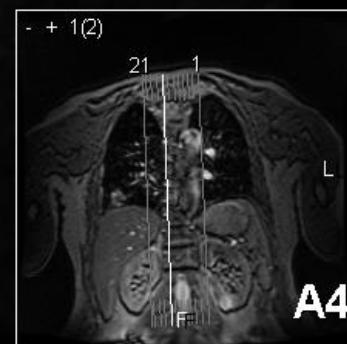
HFS

FoV: 360 mm

Image no: 14

Image 14 of 21

28/10/2010, 10:37:02



- **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> 2weeks
 - 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

Differential diagnosis

- Inflammatory diseases
 - RA, seronegative arthropathy 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

Case 3

- 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

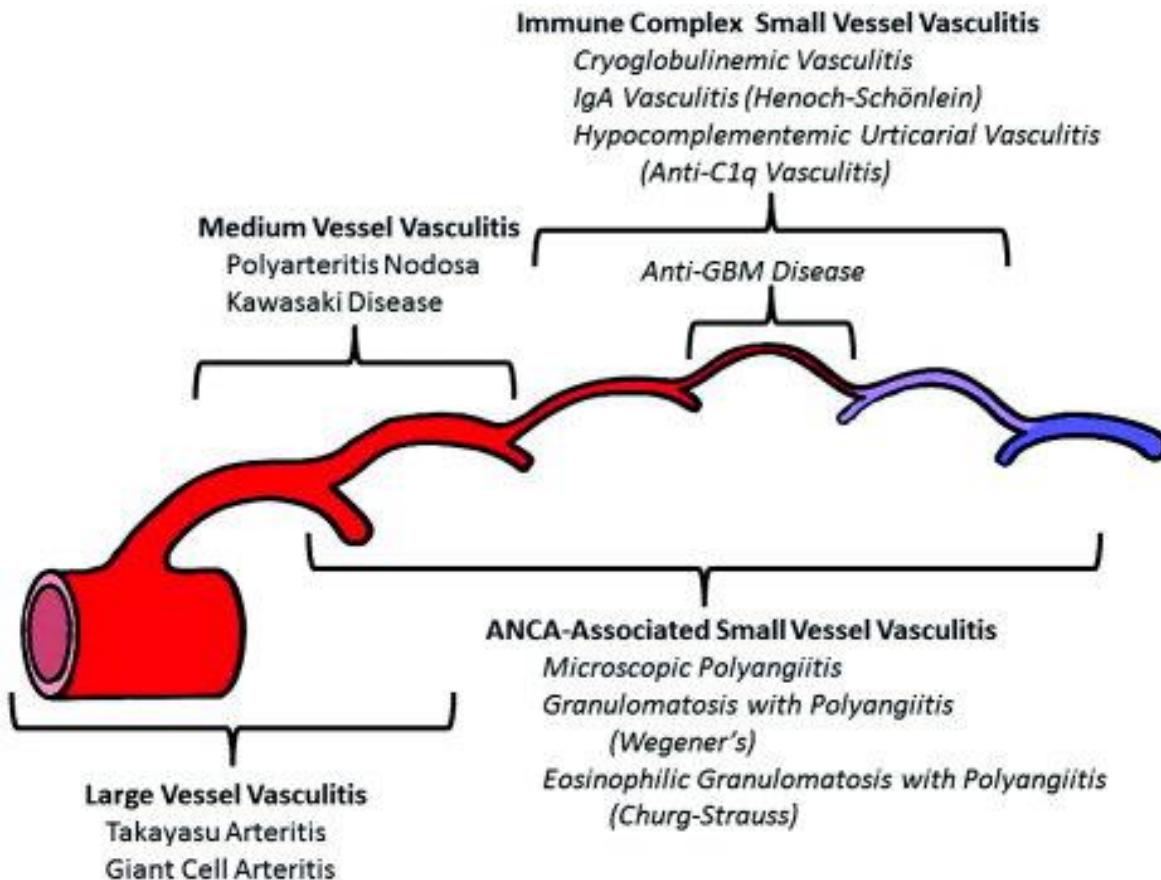


Giant cell arteritis

- 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%)

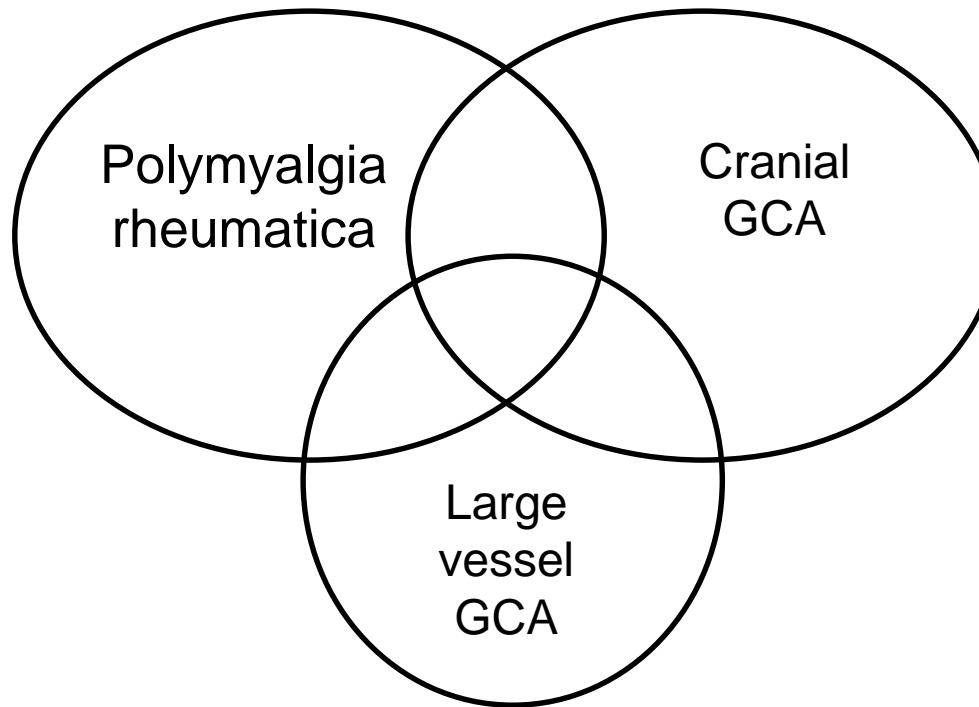
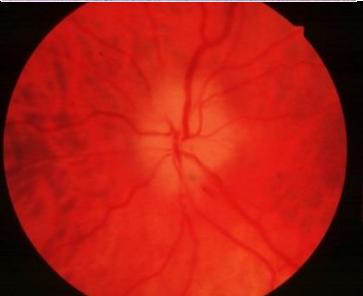
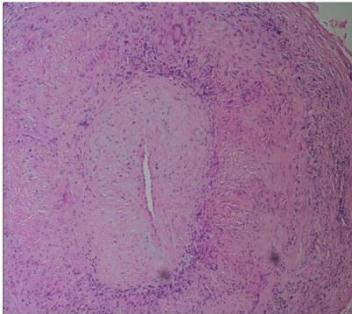
Hunder et al 1990

2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides



Giant cell arteritis

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



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)

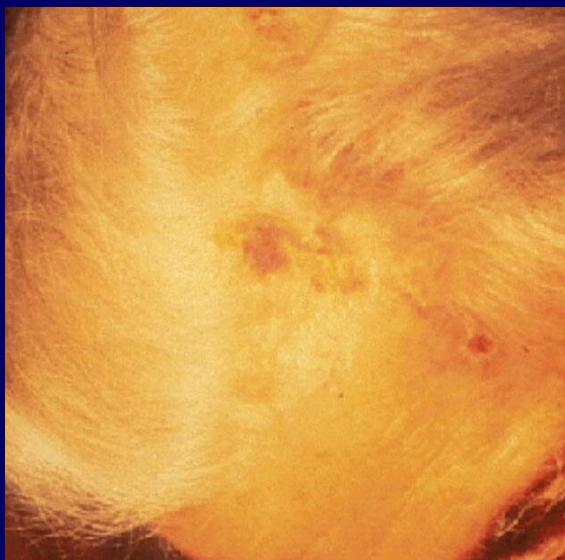
Prior JA et al.

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

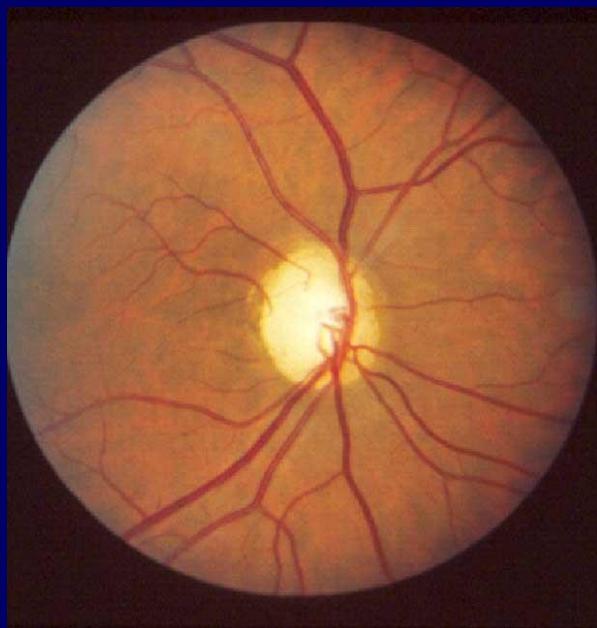
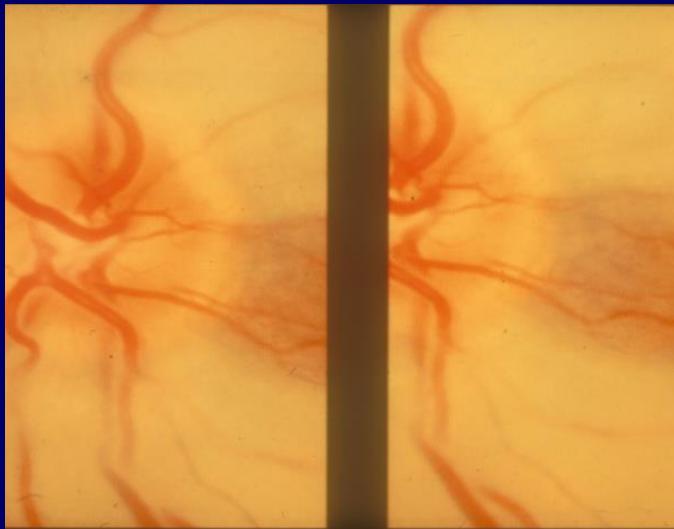
Transient monocular visual loss (amaurosis fugax)

Complete visual loss (irreversible)

anterior ischemic optic neuropathy

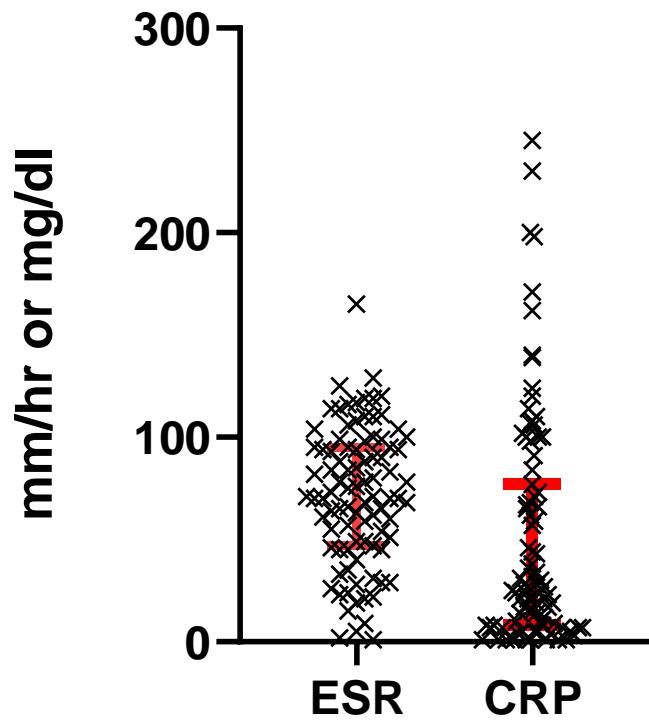
central retinal artery occlusion

Visual field defects

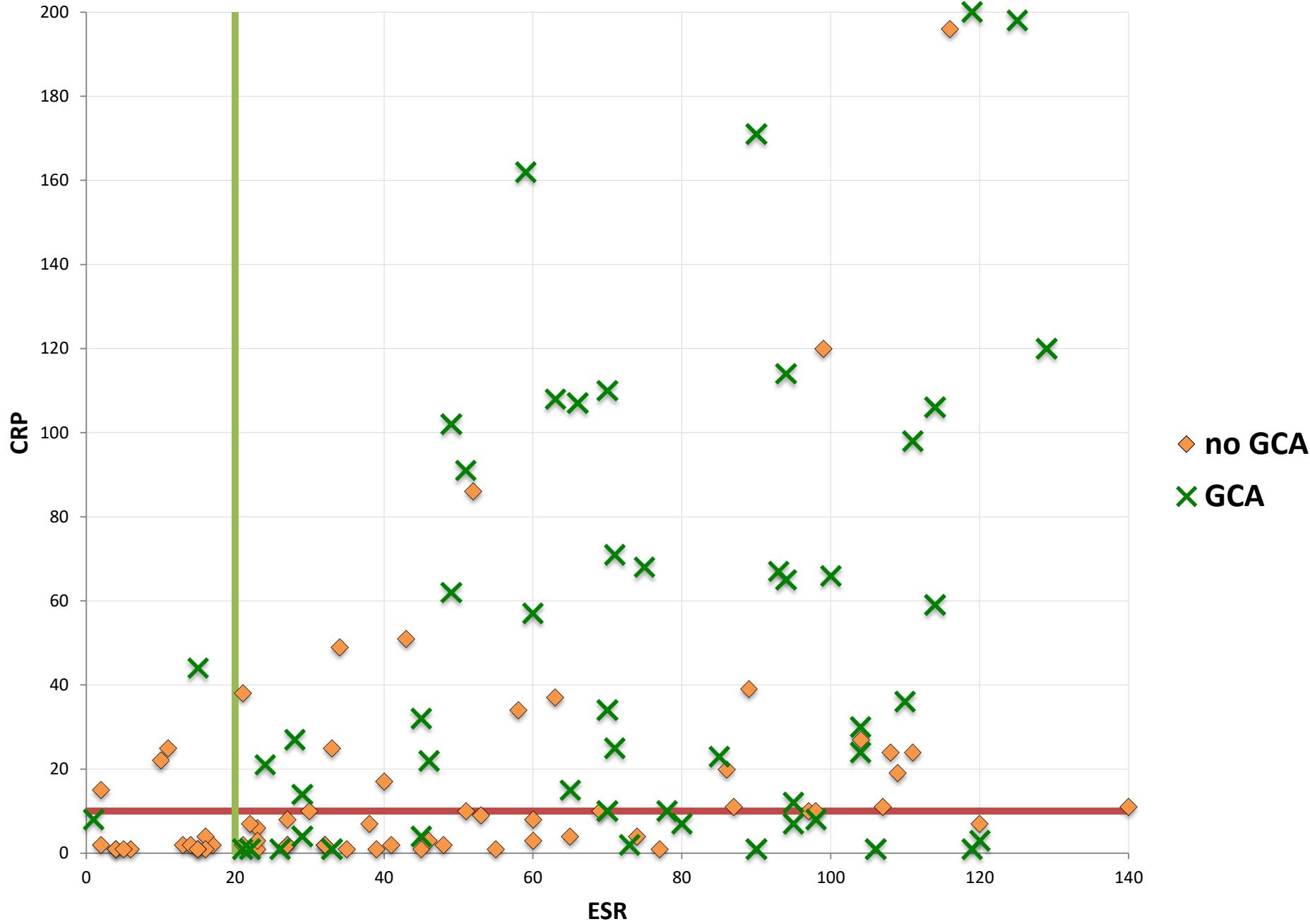


JAN	MONDAY	TUESDAY	WEDNESDAY	THURSDAY	FRIDAY	SATURDAY	SUNDAY
	3	4	5	6	7	8	9
	10	11	12	13	14	15	16
				DD	DD	D - ESTOOLUM	
	17	18	19	20	21	22 (STRAIN)	23
			NOVICAL B/ROBBIE	NOVICAL VEINS ON SIDE BIT OF HEAD	NOVICAL VEINS ON SIDE BIT OF HEAD	VEINS ON HEAD HOSPITAL	VEINS OF HEAD
	24	25	26	27	28	29	30
	U.O. HEAD JAW PAIN	U.O. HEAD DR DAVIS JAW PAIN	U.O. HEAD J/A	U.O. HEAD J/A	U.O. HEAD JAW PAIN	U.O. H J/A	U.O. H J/A
	31	1	2	3	4 RIE U.O/HEAD J/A	5 RIE U.O/HEAD J/A	6
FEB	U.O. HEAD J/A	V.O. HEAD DR DAVIS J/A	U.O. HEAD J/A	U.O/H J/A	DR DAVIS (DAD) OPTICAN TEST SOME AS HOSPITAL/NO HEMIOPAC USE HEAT PAD ON VEINS	J/A	
	7	8	9	10	11	12	13
	14	15	16	17 OPTICAN DEVIATED MY GLASS	18	19	20
	21	22	23	24	25	26	27
	28	1	2	3	4	5	6
ARCH				10	11	12	13

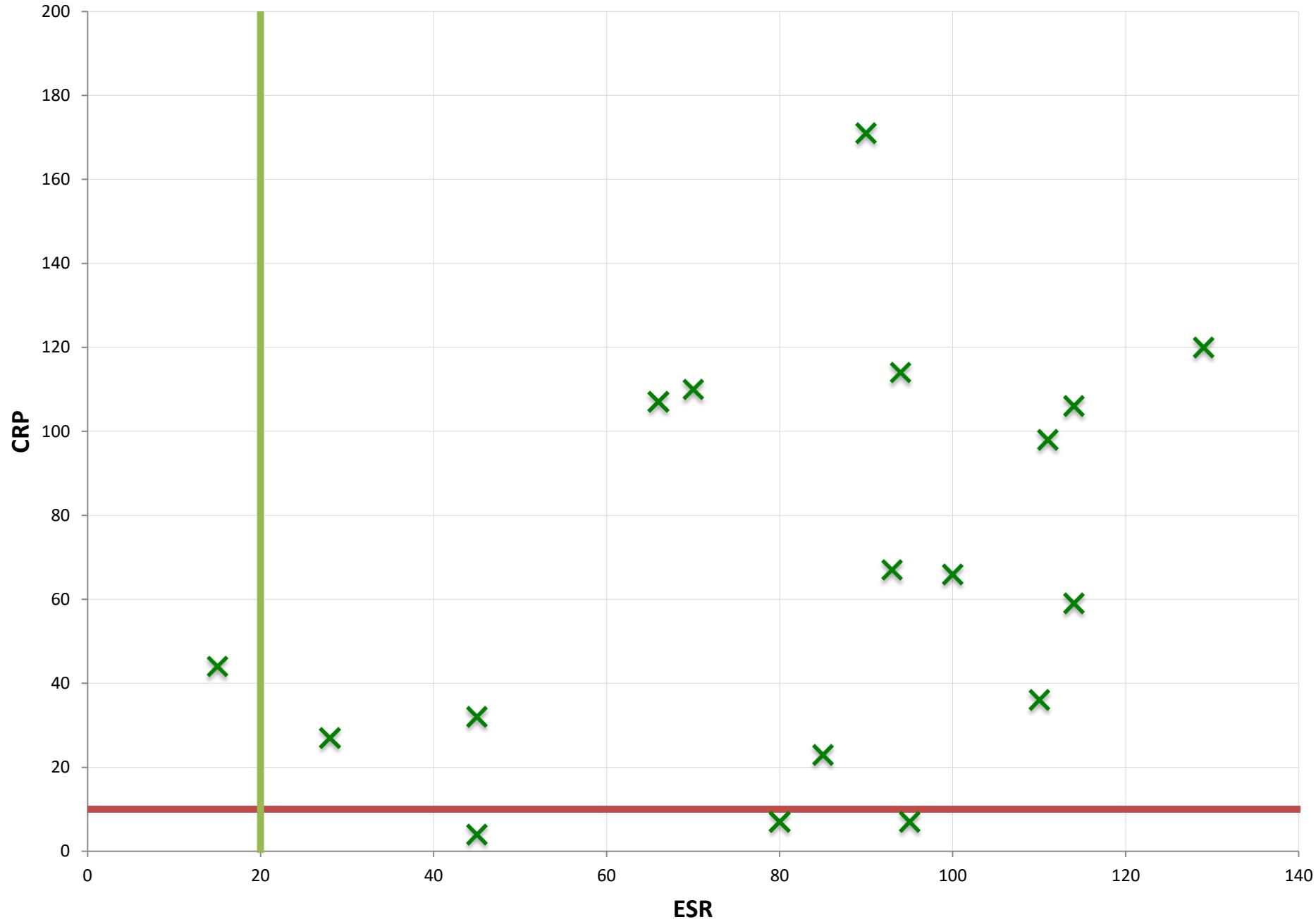
Inflammatory markers in GCA



ESR/CRP Distribution in patients with and without GCA



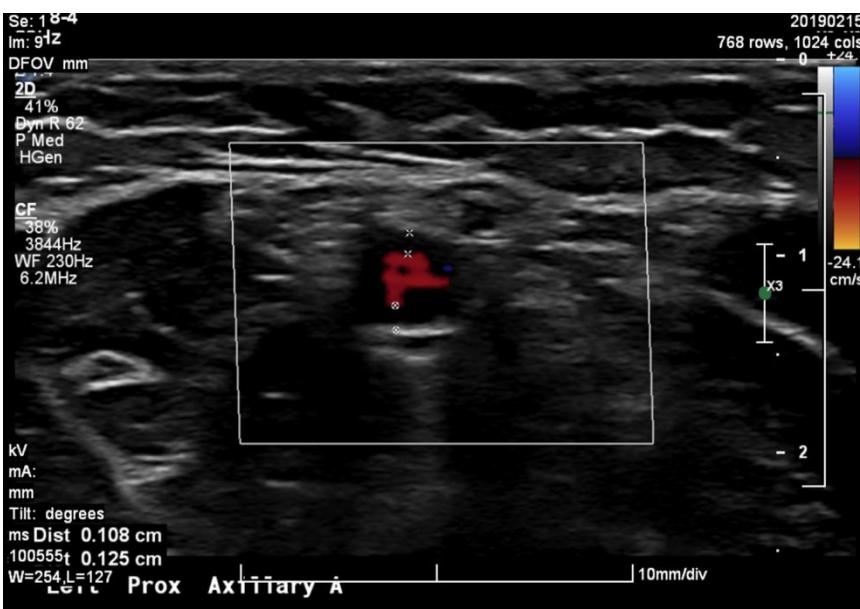
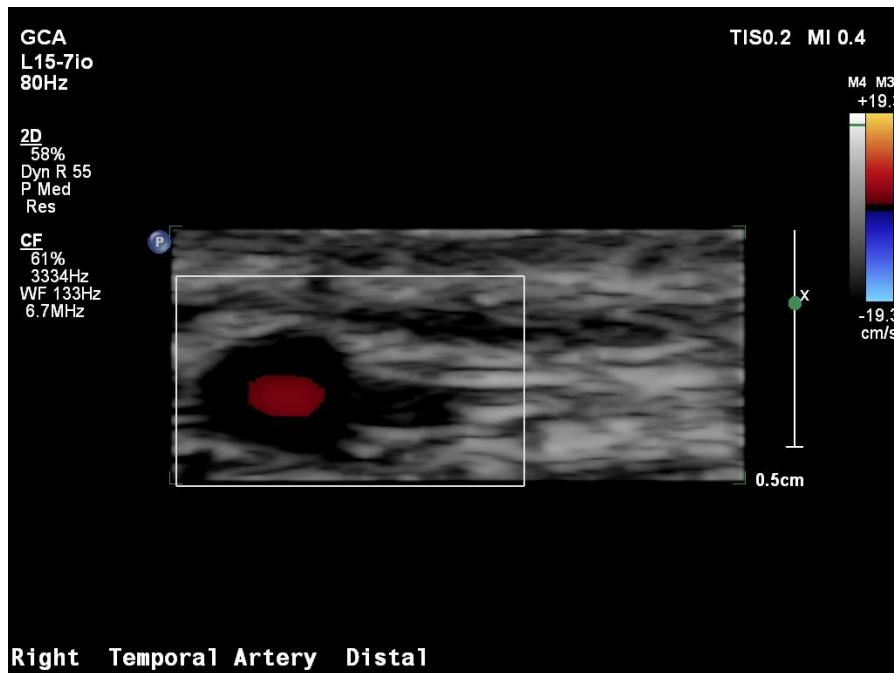
ESR/CRP in patients with biopsy confirmed GCA



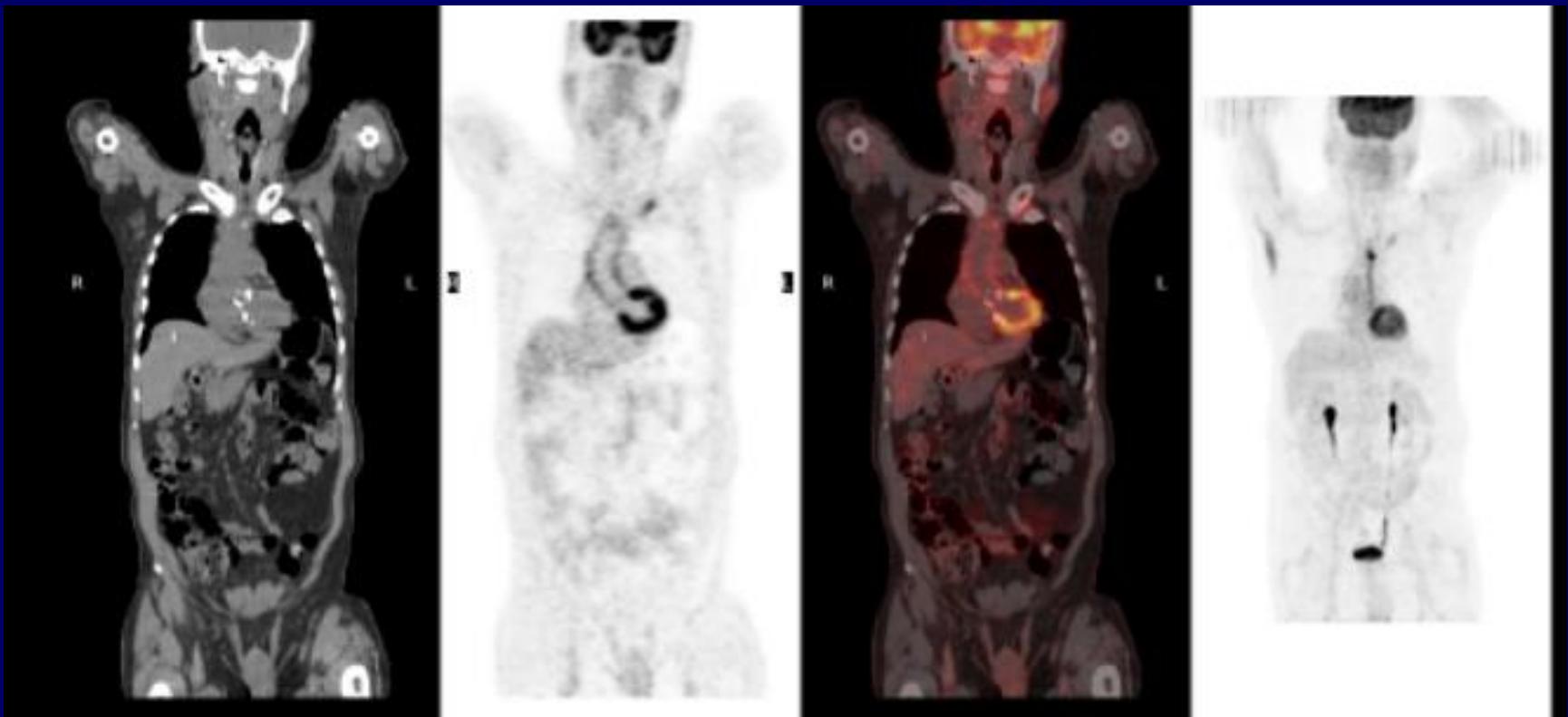
Investigations

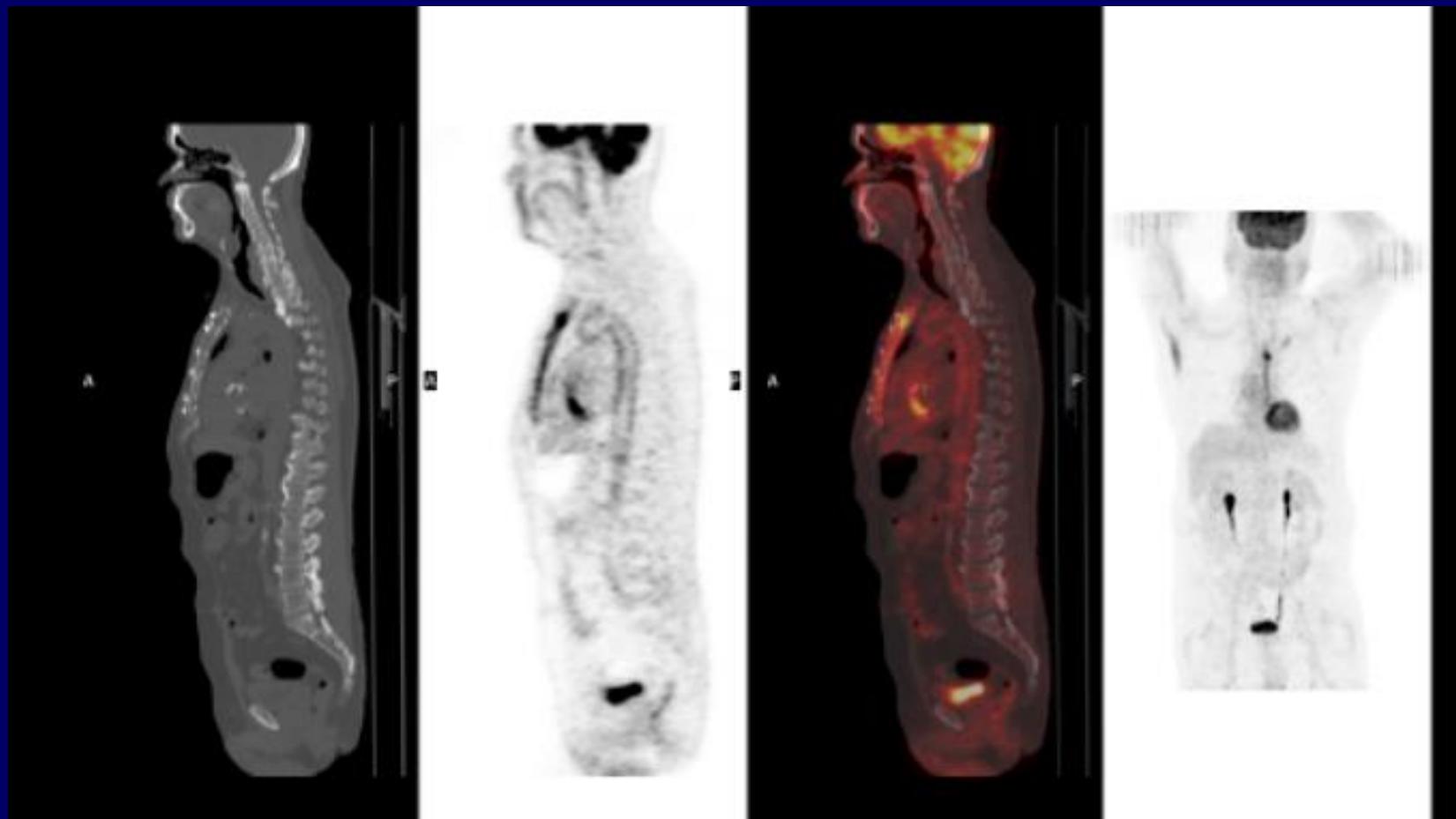
- Inflammatory markers- ESR >50mm/hr, raised CRP
- FBC- anaemia, thrombocytosis, WBC usually normal
- Deranged LFT's- Raised ALP, ALT/AST
- Temporal artery biopsy-
Recommended by guidelines, positive up to 4 weeks after commencing steroids
- Other investigations:
 - Ultrasound, MRA, CT-PET

Diagnostic test	TA biopsy	FDG-PET/CT	MRA (multi contrast HR of vessel wall)	color duplex ultrasound
Sensitivity/specificity (percent)	40/100	83/90	89/75	67-100/95 Temporal, Common Carotid and axillary arteries
Safety	Invasive, facial n. injury, infection	radiation exposure	contrast	N/A
Assessing disease activity	contralateral biopsy for recurrence?	in remission may remain positive	unknown	under study
Cost	\$541	\$5,185	\$7,348	\$388
Limitations	does not assess extra-temporal vessels	cannot assess temporal artery	limitations for MRI in general	aorta



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.

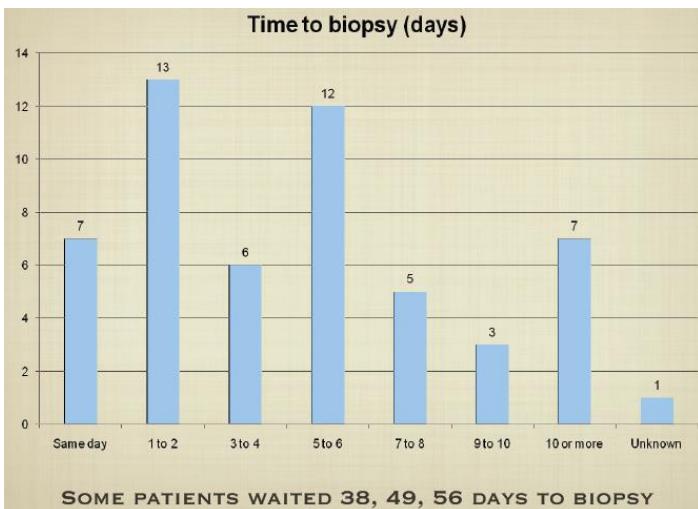
Patil P and Dasgupta et al. Clinical & Experimental Rheumatology 2015

Audit 2007- 2009

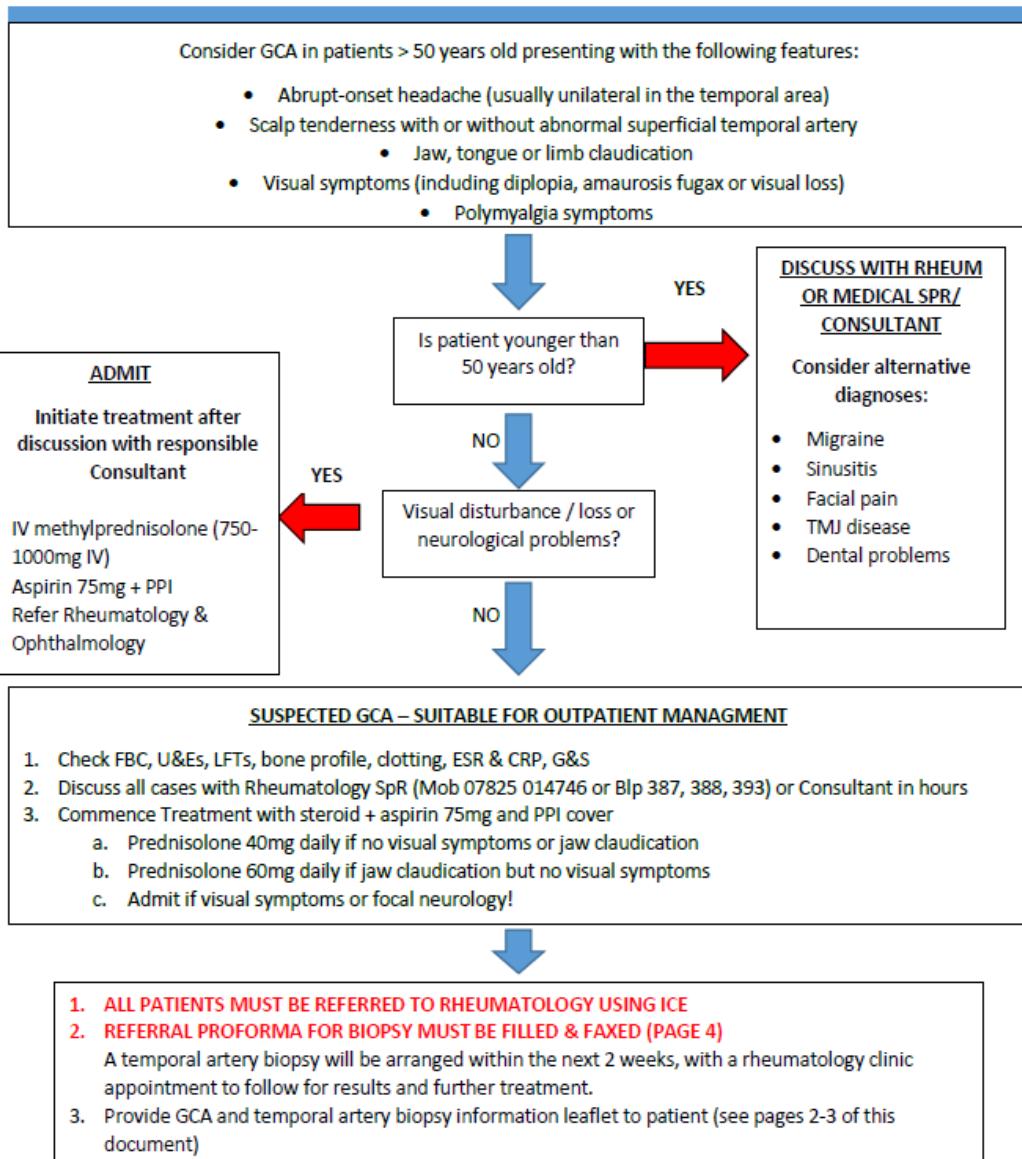
54 suspected cases
21 diagnosed GCA
13 biopsy positive

Only 39% given steroids

24% presented with
vision loss



GIANT CELL ARTERITIS PATHWAY



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

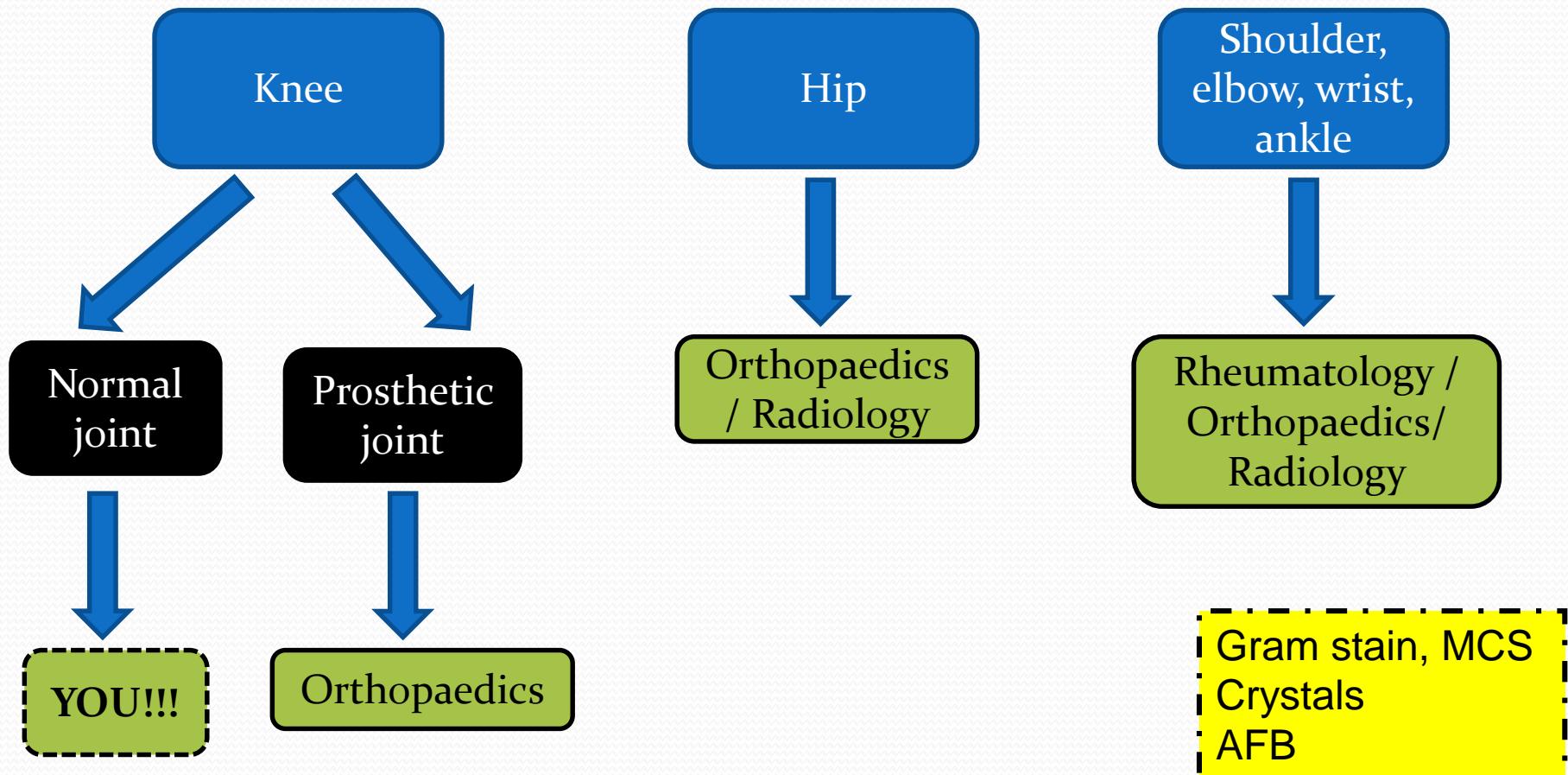
Prompt diagnosis

Aspirate joint and give antibiotics

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

	Adults (%)	Children (%)
Gram Positive Cocci		
<i>S. aureus</i>	35	27
<i>S. pyogenes, S. pneumoniae,</i>	10	16
<i>S. viridans Group</i>		
Gram Negative Cocci		
<i>N. gonorrhoeae and meningitidis</i>	50	8
<i>H. influenzae</i>	<1	40
Gram Negative Bacilli		
<i>E. coli, Salmonella</i>	5	9
and <i>Pseudomonas species</i>		
Mycobacteria and Fungi	<1	<1

Who should aspirate?



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

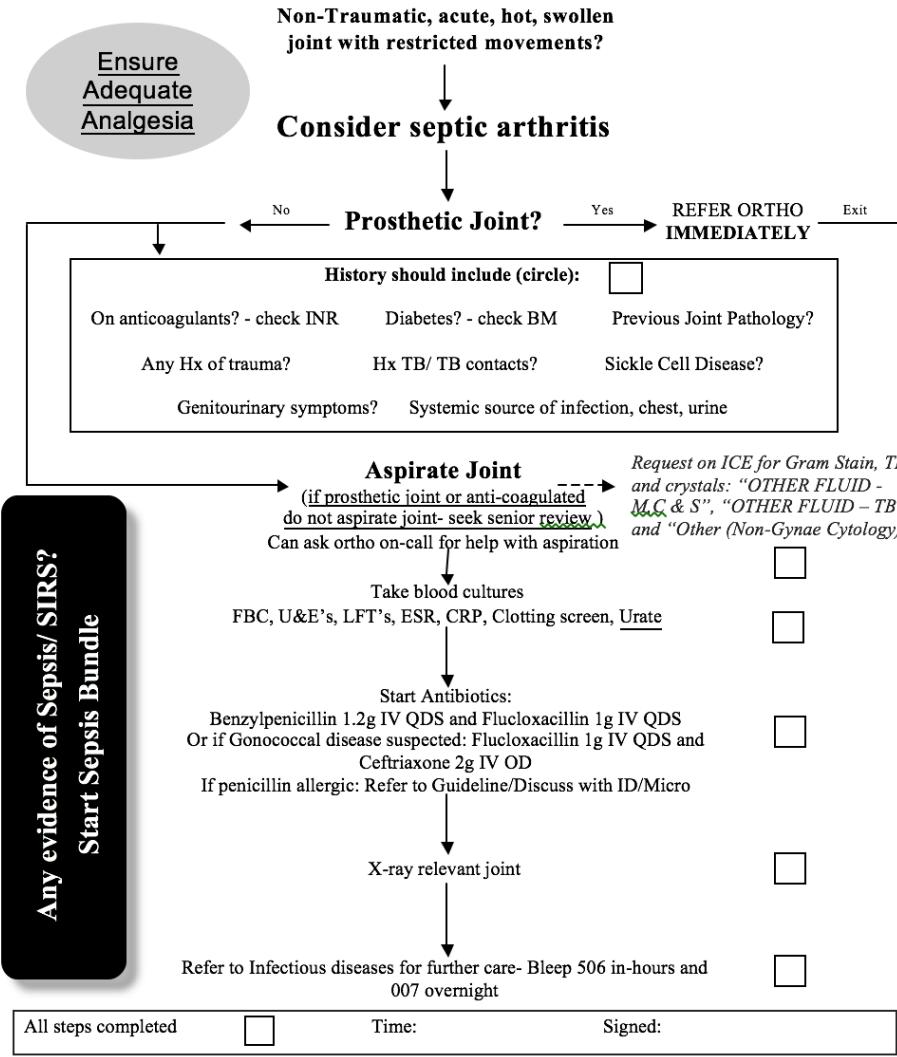
Name:
DOB:
Hospital #

Doctor:
Nurse:

Date:

Time:

Hot Swollen Joint Pathway



Case 5

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

Case 6

A 58 yo male presents to A&E with a 3/7 hx of severe pain in both ankles and right 1st 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 1st 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. 1st 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?

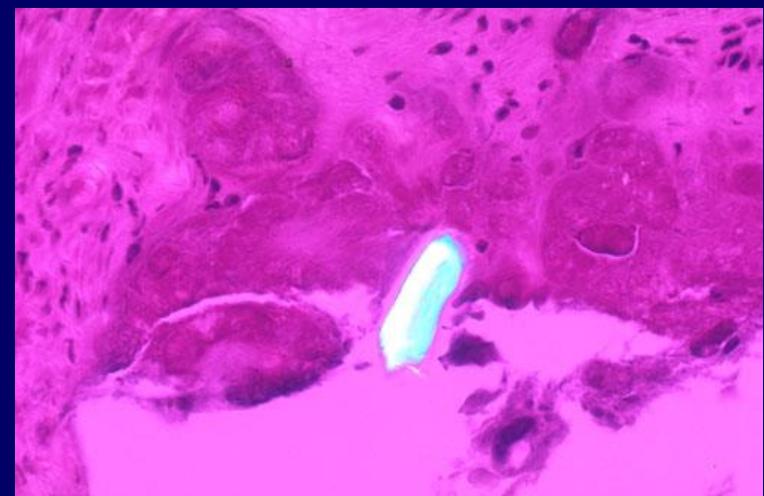
HY, JOHN

C. 4054.0, W. 3882.0

952

R





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

FERRITIN SIGNIFICANTLY RAISED---MAS

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

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
 - \downarrow Hb, NØ, LØ, platelets (all may decrease)
 - \uparrow ESR (CRP \rightarrow)
 - \downarrow Complement (decrease $C_4 > C_3$)
 - \uparrow 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

R

PA



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

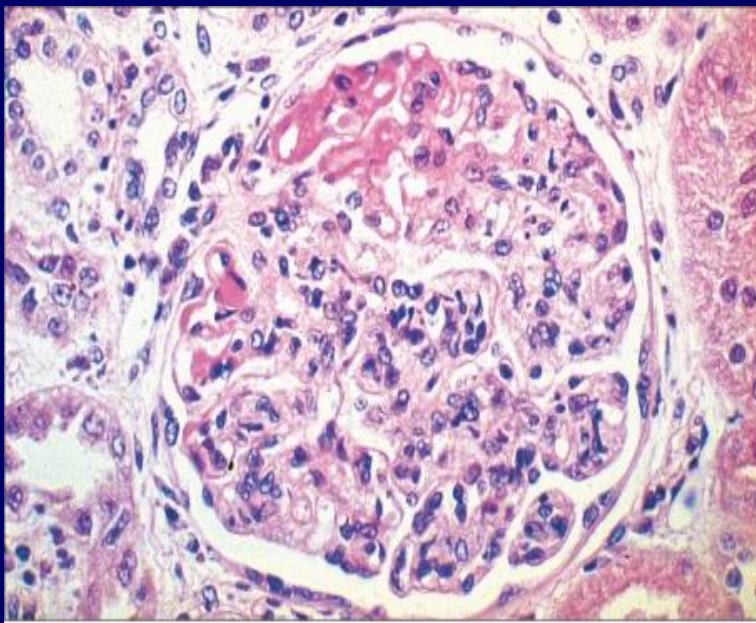
Case 12

What is the most likely diagnosis:

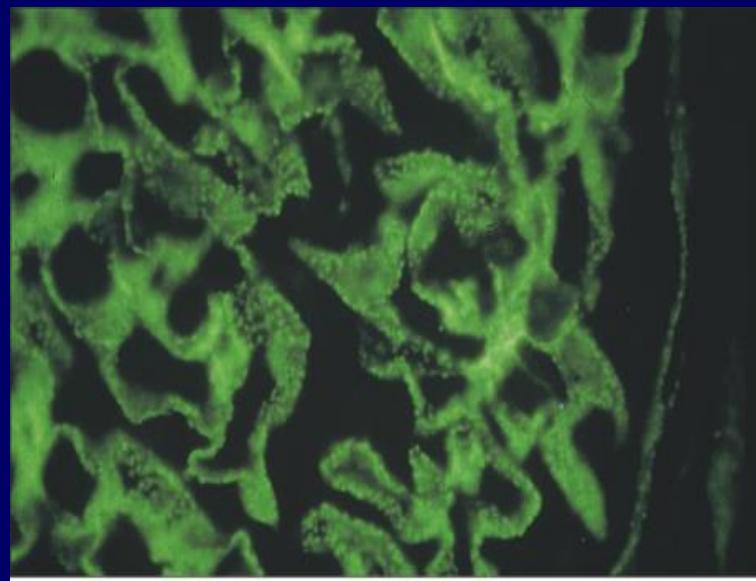
- A Goodpastures
- B SLE
- C Wegener's/ GPA
- D Microscopic polyangiitis
- E Legionella pneumonia

- 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



© www.rheumtext.com - Hochberg et al (eds)



© www.rheumtext.com - Hochberg et al (eds)

- 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, atypicals, 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

