Acute Neurology on the AMU

Chair: Professor Philip Smith

Chair: Dr Nick Scriven
Headache

Dr Ben Wakerley
Consultant Neurologist
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Objectives

• Define important **Red Flags**
• Recognize **Dangerous Headaches**
• Selected cases
Red Flags

- Headache during / after pregnancy
- Age of onset > 50y
- Neurological symptoms or signs
- Systemic symptoms or signs
- Secondary factors – e.g. cancer

De Luca and Bartleson, *Neurology Seminars*, 2010
Red Flags

- Head or neck injury
- New onset, type or worsening headache
- **Abrupt or split second onset**
- Triggered: exertion, sex, Valsalva, cough

De Luca and Bartleson, *Neurology Seminars*, 2010
Thunderclap headache

- Acute
- Sudden onset (<60s)
- Pain > 7/10
Think Blood Vessels!
Thunderclap headache

- Subarachnoid haemorrhage
- Intracranial haemorrhage
- Stroke
- Cerebral venous sinus thrombosis
- Posterior reversible encephalopathy
- Reversible cerebral vasoconstrictive syndrome
- Subdural haematoma
- Accelerated hypertension
- Pituitary apoplexy
- Cervical arterial dissection

- Meningitis / encephalitis
- Spontaneous CSF leak
- Third ventricular colloid cyst

- Primary thunderclap headache
- Primary sexual / exertional / cough headache
- Migraine
Circumstances

- Mild trauma
  - Arterial dissection, intracranial hypotension
- Dural puncture
  - Intracranial hypotension
- Fever
  - Infection
- Post partum
  - Sinus thrombosis, reversible cerebral vasoconstrictive syndrome
- Ear, nose and throat symptoms
  - Complicate sinusitis

Ducros and Bousser, BMJ, 2013
Symptoms and Signs

• Neck stiffness
  – SAH, meningitis

• Transient loss of consciousness
  – SAH, 3\textsuperscript{rd} ventricular cyst

• Seizure / focal neurology
  – SAH, cerebral venous thrombosis, stroke

• Papilloedema
  – Intracranial hypertension

Ducros and Bousser, BMJ, 2013
Posture

• Patients avoid lying flat
  – SAH, sinus thrombosis, intracranial hypertension, sinusitis

• Patients avoid standing up
  – Intracranial hypotension

Ducros and Bousser, BMJ, 2013
Headache, think fundoscopy!

Only 14% neurological patients in ER. (Bruce et al. *NEJM*, 2011)
Investigations

- Appropriate imaging
  - CT / CTV
  - MRI / MRV
- CSF
Case 1

- 32-year-old male
- Working out at the gym
- Sudden onset generalized headache
- Like being “kicked in the head”
- Drowsy, stiff neck
Subarachnoid haemorrhage
- 13% delayed diagnosis
- 32% no protocol for acute onset headache
- 11% no neurological examination

- Acute hospitals in England, Wales, Northern Ireland and the offshore islands
- 27 Neurosurgical centres
- 1st July 2011 - 30th Sept 2011
- 687 aneurysmal SAH
How common is thunderclap headache?

- 43 per 100’000 patients / year (11% SAH) Landtblom 2002
- 16 per 100’000 patients / year (25% SAH) Linn 1994

11-25% patients presenting with thunderclap headache have SAH
Why is SAH missed?

1) Failure to appreciate clinical presentation
2) Failure to understand limitations of CT
3) Failure to perform or correctly interpret results of CSF examination

Initially misdiagnosed in 1/3rd patients
The headache in SAH

History

- 70% primarily headache
- Onset – 50% split second, 50% few minutes
- Duration – 1h+
- Pain – worst ever

- Location / characteristics not important
Non-headache symptoms of SAH

- Nausea, vomiting
- Pyrexia
- Photophobia
- Neck stiffness
- Visual disturbances

- Transient loss of consciousness
- Coma
- Epileptic seizure
- Focal neurological signs
- Sudden death
Other things to remember...

- PMHx of polycystic kidney disease, Marfan’s syndrome, etc..
- FHx of aneurysms
Activity at onset – does it matter?

• Probably irrelevant (Landtblom 2002)

• 61% of SAH during rest
• 66% of non-SAH during rest

• 1.5% of SAH during sex
• 6.5% of non SAH during sex

• SAH during sleep is rare
Investigations

• **All** patients presenting with thunderclap headache must have an unenhanced CT brain

• 100% < 6h  (Backes et al. *Stroke*, 2012)
• 98% < 12h
• 90% < 24h
• 50% < 5days
If CT negative...then LP

- Ideally delay until 12h
- Experienced operator
- Measure pressure
- 4th bottle for xanthochromia
- Protect sample from light and get to lab quickly

100% sensitivity 12h – 2 weeks
Other causes of Xanthochromia
1) Jaundice (total bilirubin > 10 mg/dL)
2) Increased CSF protein (>150 mg/dL)
3) Rifampicin
4) Excess dietary carotenoids

• <48h usually 1000+ RBCs/dL
What next?

• If SAH → neurosurgeons → CTA

• Aneurysmal (85%)
• Idiopathic perimesencephalic (10%)
• Intracranial arterial dissection, AVM etc... (5%)
3 scenarios

Thunderclap headache = CT ± CSF

1) Abnormal CT
2) Normal CT, abnormal CSF
3) Normal CT, normal CSF
Abnormal CT

- Subarachnoid haemorrhage
- Intracranial haemorrhage
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Normal CT, abnormal CSF

- **Subarachnoid haemorrhage**
  - Intracranial haemorrhage
  - Stroke
- **Cerebral venous sinus thrombosis**
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- Primary thunderclap headache
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- Migraine
Normal CT, normal CSF

- Subarachnoid haemorrhage
- Intracranial haemorrhage
- Stroke
- Cerebral venous sinus thrombosis
- **Posterior reversible encephalopathy**
- **Reversible cerebral vasoconstrictive syndrome**
- Subdural haematoma
- **Accelerated hypertension**
- Pituitary apoplexy
- Cervical arterial dissection

- Meningitis / encephalitis
- **Spontaneous CSF leak**
- Third ventricular colloid cyst

- Primary thunderclap headache
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- Migraine
Normal CT, normal CSF

• Revisit the history
  – Drugs (e.g. cocaine)
  – Neck injury

• Re-examine the patient
  – Arterial tenderness
  – Horner’s syndrome
Additional investigations

Look at the vessels.....

• MRI / MRV / MRA
• CTA (intra/extra cranial)
Case 2

- 22-year-old male
- Snowboarding holiday
- Sudden onset right-sided headache and neck pain
Carotid artery dissection

Features

8% headache only (20% thunderclap)

Cerebral stroke

Horner’s syndrome

Common causes

Spontaneous
Trauma (e.g. deceleration injury)
Connective tissue disorders

Vertebral artery dissection

Features

Lateral medullary stroke

Common causes

Spontaneous
Trauma (e.g. deceleration injury)
Connective tissue disorders
Case 3

- 28-year-old female
- 2 weeks post partum
- Sudden onset generalized headache
- Vomiting
- Problems with vision
Sagittal sinus thrombosis

**Features**

- 10% thunderclap headache
- Papilloedema
- Focal seizures
- Venous infarcts
- Altered consciousness

**Common causes**

- Thrombophilia
- Pregnancy
- Nephrotic syndrome
- Meningitis / sinus infections
- Dehydration
- Underlying malignancy

D-Dimer **NOT** helpful
Pituitary apoplexy

Features

Bitemporal hemianopia

Third nerve palsy

Common causes

Pregnancy
Hyper / hypo-coagulable states
Pituitary radiation
Posterior reversible encephalopathy syndrome (PRES)

Features
- Visual disturbances
- Focal seizures
- Altered consciousness

Common causes
- Pregnancy
- Drugs
- Hyper / hypotension
- Deranged electrolytes
Cerebral reversible vasoconstrictive syndrome

Features
- Recurrent thunderclap headaches
- Seizures
- Focal neurological deficits
- 25% SAH
- Triggers: exercise, coughing, bathing

Common causes
- Post partum (50%)
- Drugs (e.g. Cocaine, cannabis)

CT brain and CSF often normal

Intracranial hypotension

Features
15% thunderclap headache
Postural headache

Common causes
Dural puncture
Migraine

• Common cause of thunderclap headache

• History
  – Previous migraine?

• Diagnosis of exclusion

• Caution!! Triptans quite effective for thunderclap headache in SAH...
Key points

- Thunderclap headache, think **vessels**
- CT brain ± CSF ± look at vessels
- Headache, think fundoscopy
Further reading

• Davenport R. Sudden headache in the Emergency department. *Pract Neurol*, 2005

• Ducros A, Bousser M. Thunderclap headache. *BMJ*, 2013
Weakness

Dr Geraint Fuller
Gloucester Royal Hospital
Teaching and Learning....

- Case based
- Bedside
- Active involvement

- Some cases
  - To discuss with the person sitting next to you
- Some clinical tips about examination
- You get to vote (sort of)
Weakness

• Acute admissions

• Clinical cases

• What to look for

• Strategies for diagnosis
You see what you are looking for
Patient GH

• A 57 year old man got onto a plane feeling fine.
• During the 1 hour flight his legs became numb and on landing he could not get out of his seat.
• He taken to A and E.
• Cranial nerves and arms normal.
• Reduced tone both legs.
• HF 3 3; HE 3 3; KF 3 3; KE 3 3; FDF 4 4; PF 4 4
• Knee reflex and right ankle reflex absent; left ankle reflex present. Plantars unresponsive.
• Loss of sensation to upper thigh
• Catheterised
Patient GH

• What is the diagnosis?

A) Guillain Barre syndrome
B) Multiple sclerosis
C) Spinal cord compression
D) Stroke
Wrong question!
Neurology and Detective Writing
Kempster and Lees, Practical Neurology Dec 2013
Neurology and the Underground
Stopping at all the stations of the diagnostic process

Symptoms and signs → Synthesis → Syndrome → Diagnosis

Weak legs

Multiple sclerosis
Neurology and the Underground

• Missing stops

Makes you

• Go too far
Stopping at all the stations of the diagnostic process

- Weak legs
- Localisation of lesion(s)
- + time course
- + tests
- Multiple sclerosis
Patient GH

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- Type of weakness?
  - Upper or lower motor?

- Distribution of weakness?
  - Spinal cord?
  - Cauda equina?
  - Peripheral nerve?

- Distribution of sensory loss?

- Autonomic involvement?
  - Spinal cord?
  - Cauda equina?
• MRCP and ‘Cases’ – ‘all’ the information

• Real world
  • Information correct?
  • All information you need?
  • What is missing?
Some difficulties

- Real patients, cases in talks, cases in MRCP and cases in detective stories

Real patients – ask yourself:
- What information is missing?
Patient GH

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Patient GH

• Is anything missing?

Modalities of sensation:
• Vibration sense
• Joint position sense
• Temperature
• Pin prick

Could there be a level...?
Dissociated sensory loss

Sensory level to umbilicus
Patient GH

- Weakness and reflex loss above L1
- Loss of spinothalamic sensation; preserved posterior column sensation
- Loss of bladder function

=> Where is the lesion?
- Anterior spinal cord syndrome at T12 or above

=> What is the lesion?
- Anterior spinal artery stroke
Missing Information

- Uncritical examination of sensory system
- Failure to examine the sensation on the trunk
Sensory Examination Made Easy....

Vibration Sense

Temperature
Sensory Examination Made Easy...

- Vibration Sense
- Temperature
- Joint position sense
- Pin prick
- (light touch.....)

- Distal to proximal
- Abnormal to normal
- Delineate edge of normal
- Look for level if you think there might be one......
• Think about the sensory distribution in all 4 modalities
  – Missing information

• Remember to look for a level on trunk
  – Missing information

• Imagine you are ‘shading in a drawing’
Myotomes and Dermatomes
Patient CD

- 35 year old man
- Previously fit and well
- 3 weeks ago right eye droopy couple of days
- 1 week of difficulty eating; food getting stuck behind lips; swallowing normal.
- Some difficulty with lifting his hands above his head.
- Well
- Bilateral facial weakness
- SA 4 4; EE 4 4; EF 4+ 4+; WE 5 5; FE 5 5
- Legs 5 5
- Reflexes symmetrical +
- Plantars flexor
- Sensation normal
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What level of the nervous system is involved?

A) Brain
B) Brainstem
C) Spinal cord
D) Anterior horn cell
E) Nerve
F) Neuromuscular junction
G) Muscle
Weakness – types

- Upper motor neurone
- Lower motor neurone
- Neuromuscular junction
- Muscle
- Non-organic

Fig. 1 Levels of the nervous system.
- Weakness

- Distribution

- Hemisphere lesion
  - Brain stem lesion
  - Cervical spine lesion

- Hemisphere lesion
  - Brain stem lesion
  - Cervical spine lesion
  - Thoracic spine lesion

- Hemispheric lesion

- Cervical spine lesion

- Cervical spine lesion
  - Thoracic spine lesion

- Generalized neuropathy (may involve face and neck)

- Cauda equina syndrome

- Neuromuscular junction or myopathy (or non-organic) (may involve face/neck)
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Bilateral facial weakness with mild proximal arm weakness.
History of transient ptosis.

Nerve/nerve root

But:
Unexplained ptosis
Reflexes normal
Sensation normal

Neuromuscular

No fatiguability
Bilateral facial weakness without ptosis
Patient CD

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Revisit the history
Evidence of variability?

Re-examine the patient
Evidence of variability?
Evidence of fatigability?

Re-assess the patient
Vital capacity – lying and sitting
Patient CD

- CSF – acellular; protein normal
- Trial of pyridostigmine
- NCS – normal
- Repetitive stimulation – Normal
- Anti-AChR negative
- Anti- MUSK positive
Myaesthenia Gravis

• Anti-acetyl choline receptor antibodies in 85% of generalised MG

• Anti-muscle specific kinase (anti-MuSK) in 30-70% of seronegative cases
  – More bulbar and facial involvement
  – More prone to respiratory involvement
  – Pyridostigmine not effective in some
Mr CS

• 38 year old man
• 1 week before developed pain down back of right leg with numbness of whole foot
• 3 days ago developed numbness in left foot and some weakness in both legs
• Woke with numbness in both hands, left more than right, mainly in thumbs and index fingers.
Mr CS

- Afebrile
- Cranial nerves normal
- R finger extension 4/5; both abductor pollicis brevis 4/5
- R knee flexion 4+/5; R foot dorsiflexion 4-/5; Left 4+/5
- Reflexes
- PI

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Mr CS

- Discuss
- Where?
- What?
- Why?
Mr CS

Question:
Where is the lesion?
A: brain
B: brainstem
C: spinal cord
D: nerve roots
E: peripheral nerves
F: neuromuscular junction
G: muscle
H: non-organic
Mr CS
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Mr CS: Roots or nerves?

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O/E

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- Reflexes – lost right supinator, left ankle jerk
The Myotome Dance
Mr CS: Roots or nerves?

- 38 year old man
- Asthma for 2 years
- 1 week before developed pain down back of right leg with numbness of whole foot
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**Where?**
- Peripheral nerve

**What**
- Multifocal peripheral neuropathy
- Stepwise & progressive

= Vasculitic neuropathy
How to recognise and treat peripheral nervous system vasculitis

E A Marsh1 2 L M Davies3 J G Llewelyn1 2

Pract Neurol doi:10.1136/practneurol-2012-000464

• FBC – eosinophilia
• Inflammatory markers (PV/ESR/CRP)
Patient NH

- 76 year old man
- Diabetic for 5 years on metformin
- 3 month history of numb clumsy hands
- 1 month of difficulty walking.
- Cranial nerves normal
- Arms full power
- HF 4+ 4+
- Plantars unresponsive
- VS lost to ASIS and to wrist
- JPS reduced distal interphalangeal joint hands
- PP and temp reduced to mid forearm and to mid-calf

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“Numb clumsy hands”
Bias in Neurological Reasoning
Based on AJP 2009;192:561-4

• Attribution bias
  – Has diabetes – problem relates to diabetes
• Availability bias
  – This is the stroke team
  – The patient is on the stroke ward
  – They have had a stroke
• Confirmation bias
  – Well that proves it....
• Framing bias
  – My legs are heavy because they are swollen – why are they swollen?
  – No - why are they weak?
• Hindsight and regret bias
  – Retrospectoscope...I don’t want to miss that again....
• Satisfaction of search bias
Patient IJ

- 58 year old man
- 3 week ago developed numbness going down his right arm. Persisted.
- 10 days ago numbness on back of left thigh to foot.
- 1 day of right facial weakness.
Patient IJ

- Right lower motor neurone facial weakness
- Mild weakness of right shoulder abduction (4+).
- Bilateral weakness of hip flexion (4)
- Loss of both ankle reflexes
- Plantars flexor
- VS lost to knees; JPS normal.
- Soft alteration to pinprick and temperature on lateral right arm and back of left thigh.
Patient IJ

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Right C5 radiculopathy
Left leg radicular symptoms
Recent right facial weakness
Mild bilateral lower motor neurone leg signs

Basal meningitis
Patient IJ

• Chronic meningitis
  – Inflammatory
  – Infective
  – Malignant

• Multiple cranial nerve (especially 7th and 8th)

• Multiple nerve roots (especially cauda equina)

• Need to look at CSF!
Patient AB

• 73 year old
• Diabetes for 10 years
• Breast cancer 6 years ago; lumpectomy; DXT; Chemo
• 4 days of progressive weakness; legs => arms.
• Numbness in both hands and legs.
• Band feeling around waist.
• Normal bowels and bladder
• Now walks with difficulty
Patient AB

• Cranial nerves normal
• Tone normal
• SA 5 5; EF 4+ 4+; EE 4 4; WE 4 4; F Abd 4 4
• HF 4- 4 -; HE 4 4; KF 4 4; KE 4 4; FDF 4+ 4+
• Areflexic
• VS lost to ASIS and fingers
• JPS lost to ankle
• PP and temperature lost to mid calf
Patient AB

- 73 year old
- Diabetes for 10 years
- Breast cancer 6 years ago; lumpectomy; DXT; Chemo
- 4 days of progressive weakness; legs => arms.
- Numbness in both hands and legs.
- Band feeling around waist.
- Normal bowels and bladder
- Now walks with difficulty
- Cranial nerves normal
- Tone normal
- SA 5 5; EF 4+ 4+; EE 4 4; WE 4 4; F Abd 4 4
- HF 4- 4 -; HE 4 4; KF 4 4; KE 4 4; FDF 4+ 4+
- Areflexic
- VS lost to ASIS and fingers
- JPS lost to ankle
- PP and temperature lost to mid calf
Thank you
Patient DN

• 28 year old woman
• 1 week of tingling in both hands
• 3 days of altered feeling in both feet and difficulty going upstairs.
• 1 day of thoracic back pain and increasing difficulty walking.
• Noted tingling on the tip of her tongue
Patient DN

• Cranial nerves normal
• Walking with one.
• SA 4 4; EE 4+ 4+; EF 4+ 4+; FE 4 4
• HF 4- 4-; HE 4 4; KF 4 4; KE 4+ 4+; FDR 4 4
• Areflexic
• VS – normal
• JPS – normal
• PP and T – soft alteration to mid calf.
• 28 year old woman
• 1 week of tingling in both hands
• 3 days of altered feeling in both feet and difficulty going upstairs.
• 1 day of thoracic back pain and increasing difficulty walking.
• Noted tingling on the tip of her tongue

• Cranial nerves normal
• Walking with one.
• SA 4 4; EE 4+ 4+; EF 4+ 4+; FE 4 4
• HF 4- 4-; HE 4 4; KF 4 4; KE 4+ 4+; FDR 4 4
• Areflexic
• VS – normal
• JPS – normal
• PP and T – soft alteration to mid calf.