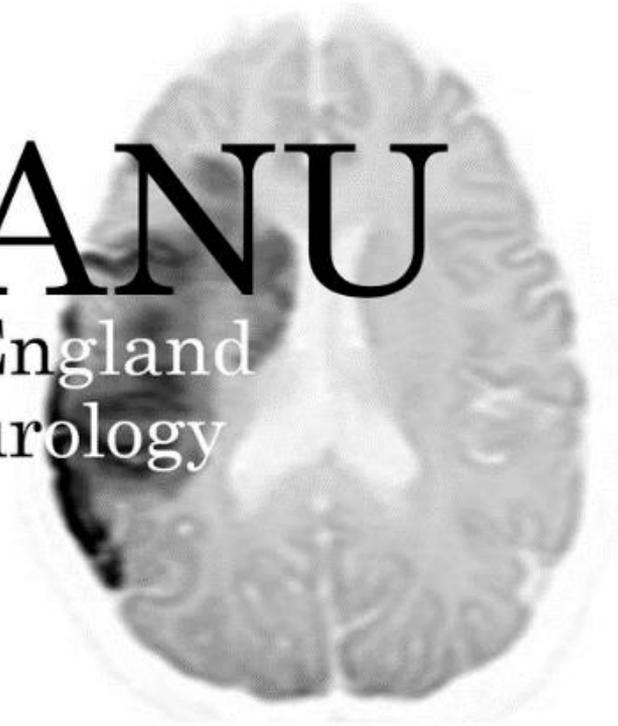


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North of England
Acute Neurology
Update



Eye signs in acute neurology

Case A

- 30 year old male
- 2 day history blurred vision – both eyes
- Mild headaches for 1 week

- O/E
 - VA 6/24 bilaterally
 - Normal fields
 - Normal eye movements

Case A – Differentials?

Acute/subacute syndrome of raised ICP

Primary

- IIH

Secondary

- Cerebral venous sinus thrombosis (CVST)
- *Extracranial* venous obstruction
- Mass lesions
- Hydrocephalus
- Malignant hypertension
- Others: tetracycline (and other drugs) renal failure, SLE, Addison's, hypoparathyroidism...

Case A – Investigations?

- Brain imaging...
with venography

Cerebral venous sinus thrombosis - 1

Key points

- Plain imaging can be normal – beware misdiagnosis of IIH
- MRV notoriously difficult to interpret...
- Look for underlying causes:
 - Rheum: SLE, APL, Behçets
 - Haem: thrombocythemia, protein S deficiency, anti-thrombin III deficiency, activated protein C resistance
 - Infections: mastoiditis
 - Post-partum, post-surgery, on OCP

Cerebral venous sinus thrombosis - 2

- Treatable...don't miss it:
 - Examine the fundi
 - Low threshold for venous imaging
- Treatment:
 - Anticoagulation
 - Look for a cause
 - Document visual fields and VAs – may need therapeutic tap or even lumbar drain
 - Don't worry (too much) about haemorrhage

Case B

- 65 year old male
- Double vision for 1 day
- Painless
- Worse looking to the left
- PMH
 - DM
 - HTN
 - Raised cholesterol

Case B - differentials

Acute painless NVI palsy

First, is it isolated? In the history and examination, specifically look for:

- Orbital disease (e.g. chemosis, proptosis)
- Myasthenia gravis (fatigue/variability, ptosis, eyelid closure weakness)
- Other cranial nerve lesions or brain stem signs (e.g. NVI palsy, Horner's sign)
- Systemic, infectious or inflammatory risk factors (e.g. malignancy, temporal arteritis)
- Severe headache (aneurysm...rare but important)
- Optic disc swelling (false localising sign)

Case B - differentials

- If truly isolated, causes of NVI palsy include:
 - Vasculopathic
 - Most common
 - Non-vasculopathic
 - Be more careful...
 - Traumatic
 - Congenital

Case B - treatment

- Treatment for vasculopathic NVI palsy
 - Watch and wait
 - Can worsen for up to 1 week
 - Can take 12 weeks to improve
- Neuroimaging not required, *unless*:
 - Progressive
 - Non-resolving
 - Any suggestion of non-isolated NVI
- Have a low threshold for checking the ESR/CRP

Case C

- 58 year old man, previously fit and well
- 6 week history of intermittent symptoms
- Double vision whilst driving
- Droopy eyelids in the evening
- Seemed to worsen recently after being given antibiotics for a chest infection

Tangent - pitfalls in myasthenia

- Exposing patient to a trigger:
 - <https://www.myaware.org/drugs-to-avoid>
- Missing a thymoma
- “seronegative” patients
- Steroids
 - High doses can cause “Drachman dip”
- Pyridostigmine:
 - Airway secretions – usually paused around intubated/extubation
 - IV pyridostigmine 30 times more potent...

Case D

- 66 year old man
- Headache for 2 days
- Droopy right eyelid since this morning
- Double vision

- PMH
 - DM, HTN, Raised cholesterol
- DH
 - Statin, metformin

Tangent – mono-ocular diplopia

Key question: is the diplopia mono-ocular or binocular?

Mono-ocular diplopia

False image visible **from one eye only**

Common causes include:

- Problem with tear film/cornea/lens/vitreous or retina
- Functional

Usually need to see friendly eye doctor rather than neurology

Binocular diplopia

False image visible when both eyes open.

Produced from eye misalignment of any cause.

Case D - differentials

Acute NIII palsy, pupil sparing

- First, is it isolated? Look for:
 - Orbital disease (chemosis, proptosis, disc swelling etc.)
 - Myasthenia gravis
 - Other CN lesions
 - Brainstem or limb signs
 - Systemic, infections, or inflammatory risk factors...low threshold for checking ESR
 - Severe headache (aneurysms...)

Case D - differentials

- If isolated, is the pupil involved?
 - Yes:
 - Aneurysm until proven otherwise
 - No:
 - Ischaemic – esp in patients with DM, but don't forget GCA
 - Trauma
 - Congenital

Case D - outcome

- CTA normal
- Pain settled in a few days
- Ptosis and ophthalmoplegia improved in around six weeks
- HTN, DM and cholesterol treatment optimised

Case E

- 61 year old man
- 3 weeks of right retro-orbital pain
- 1 week history of double vision, ptosis and reduced visual acuity from right eye
- Some night sweats for a few weeks, increasingly fatigued

- Otherwise fit and well

- No meds

Case E - examination

- Right eye proptosis and chemosis
- Right eye does not move at all
- RAPD in the right eye, VA 6/36
- Reduced sensation right forehead
- Absent corneal reflex

- **Which nerves are involved?**
 - NII NIII, NIV, NV, NVI

Case E – where is the lesion?

Case E - differentials

Subacute orbital apex syndrome

- Malignancy
- Inflammatory disease:
 - Sarcoidosis
 - Systemic lupus erythematosus
 - Eosinophilic granulomatosis with polyangiitis
 - Granulomatosis with polyangiitis
 - Giant cell arteritis
 - Thyroid eye disease
- Trauma
- Vascular
 - Carotid aneurysm/fistula

Case E - imaging

- Mucous blood and necrotic material (arrow)
- Soft tissue infiltration (arrowhead)

Case E - outcome

- Orbital decompression surgery and biopsy
- Steroids administered – VA improved to 6/12
- Histology showed large B-cell lymphoma
- Chemotherapy commenced

Case F

- 54 year old woman
- Sees chiropractor about her neck
- 2 days after neck manipulation, right eyelid drooping noticed
- Husband noticed that right pupil is smaller than the left
- No diplopia or visual loss

Case F - differentials

Horner's syndrome

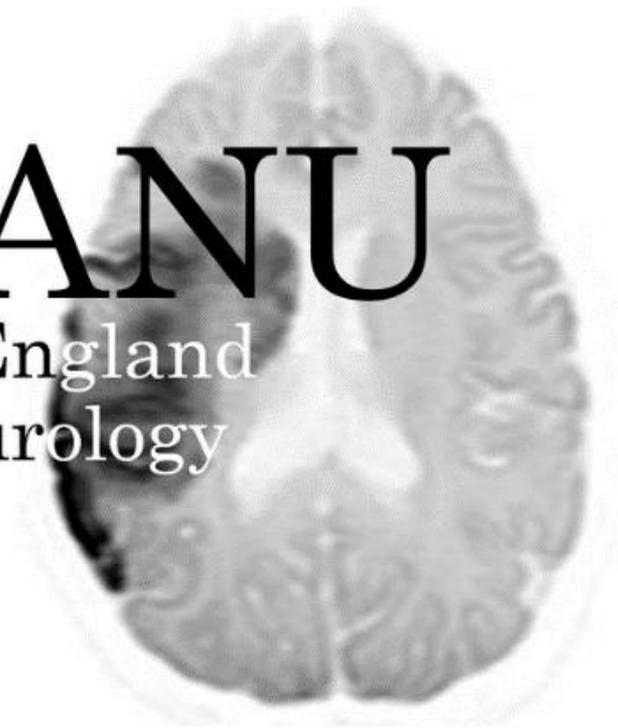
- Lesion anywhere along sympathetic pathway:
 - Lateral brain stem lesions
 - Stroke
 - Tumour
 - Syringobulbia
 - Cervical spinal cord
 - Syringomyelia
 - Lung apex - Pancoast
 - Carotid disease
 - Orbital disease etc.

Case F - investigations

- Confirmed right carotid artery dissection
- Rx aspirin

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Quick fire round: *the weird and the wonderful*

- History and a video/picture
- What tests?
- What diagnosis/treatment?

Quick fire 1

- A 30-year-old man
- Progressive dysphagia, dysarthria, and weakness in his arms and legs over 6 months
- Now unable to swallow – presents to ED
- O/E
 - Tongue wasting and ->
 - Widespread wasting and fasciculations, brisk reflexes
 - Normal sensation
- NCS - normal peripheral nerves
- EMG – florid acute and chronic denervation
- Diagnosis: ALS

Quick fire 2

- 50 year old lady
- GP sent in – “generally unwell, not responding to antidepressants”
- Gaining weight
- Tired all the time
- Intolerant of cold

- O/E ->
 - Woltman’s sign

- TFTs – T4 undetectable, TSH 20 (0/4-4)
- Diagnosis: Hypothyroid
 - Thyroxine started ->

Quick fire 3

- 70 year old lady
- 2 year history of gait disturbance
- Now falling – sent in “off legs”
- Memory failing (MMSE 12)
- Family told to look at local nursing homes

- O/E ->

- Imaging ->

- Diagnosis: NPH
- Rx VP shunt ->

Quick fire 4

- 48 year old female
- 2 week history of general malaise
- Now acutely confused, drowsy and started having abnormal movements

- O/E ->

- Na⁺ 130
- LP – protein 0.6, normal cells
- LGI1 antibodies positive in serum and CSF

- Diagnosis: autoimmune encephalitis with faciobrachial dystonic seizures
- Rx steroids +/- more aggressive immunosuppression

Quick fire 5

- 26 year old female
- 2 weeks of change in behaviour – poor memory, confused speech
- Became drowsy and then comatose

- O/E ->
 - Jaw dystonia
 - Opisthotonos
 - Dystonic posturing upper limbs

- Ix
 - USS abdo – ovarian teratoma
 - Serum and CSF positive for NMDA receptor Abs

- Diagnosis: Paraneoplastic NMDA receptor antibody mediated encephalitis

- Rx
 - Immunosuppression
 - Tumour removal

Quick fire 6

Could you have worked it out from witness history alone?

Smart phone videos are **really** useful

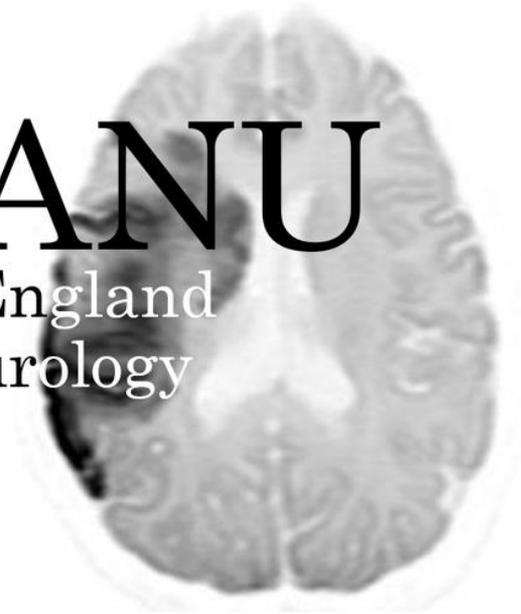
<https://www.nejm.org/doi/full/10.1056/NEJMra1500587>

Quick fire 7

- 4 week history rash on face and hands
- Now struggling to get up the stairs
- O/E ->
- Ix
 - CK 2,500 IU/L
 - Mi2 autoantibodies
 - CT TAP no malignancy
- Diagnosis: dermatomyositis
- Rx: immunosuppression

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Thanks and Questions?

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