AMU Clinical Reasoning Session Key Learning Points (15.5.26)

Case 1 Summary

Young lady (in her 20s) presented with 1/52 of paraesthesia (pins & needles) in her feet progressing up her lower limbs which was preceded by a viral flu-like illness a couple of weeks ago. She had no other associated symptoms, nil PMH or regular medication. No recent stressors noted with no previous similar episodes.

On assessment in AMU, she was alert & orientated with nil systemic signs. Asides from paraesthesia in her feet / legs – no other focal neurology inc normal autonomic function. Obs & bloods (inc haematinics, vitamins, TSH) were all NAD. An MRI spine reported demyelinating lesions suggestive of MS for which she was referred to Neurology.

Heuristics (thinking tools) from this case

1. Simple, Specific, Structured (SSS)

   Neurological presentations are classically complex and make you feel like you are stuck in a vast ocean! But this can be simplified with simple starting steps.

   First & foremost, think is this:
   a) *UMN (CNS) vs LMN (PNS) -> motor vs sensory vs mixed (MND)*
   b) *Extrapyramidal -> Parkinsonism signs*
   c) *Cerebellar -> impaired coordination*

   In this case, the patient has a sensory issue (paraesthesia), but it is unclear if it is an Upper Motor (UMN) or Lower Motor (LMN) sign. Let's delve into this next..

2. For & Against approach

   For LMN – paraesthesia without any UMN signs (e.g. hypertonia, hyperreflexia)
   Against LMN – No other LMN signs (e.g. wasting, fasciculations, areflexia)

   For UMN – the key clue is 'bilateral' progressive lower limb paraesthesia which could indicate spinal cord pathology.

<u>Against UMN</u> – No other UMN signs

This case is tricky because all you have is lower limb paraesthesia so on the surface it is difficult to know for sure whether it is UMN or LMN. I think the most important examination is whether there is a sensory level (UMN) or whether it is patchy disturbance (LMN). However, in cases of diagnostic uncertainty (even after using the for & against approach) – ***it is prudent to investigate for both..***

3. <u>Red flag or Red herring</u>

The biggest red flag in this case is ***bilateral & progressive*** *paraesthesia.* ***Bilateral neurological signs*** *should always warrant suspicion* ***of spinal cord pathology. Progressive / ascending*** *is also concerning in this context.*

The lack of any motor weakness although is reassuring – is not a red herring. You do not know whether the patient ***is yet to develop weakness in her disease*** *(esp. with the progressive nature of her paraesthesia).*

We spoke about whether her preceding viral illness is a red flag or red herring. I would argue that it is a red flag. It is a known phenomenon that viral / infective illness can cause deconditioning & activate underlying MS lesions or cause transverse myelitis (inflammation of spinal cord) through an immune-mediated process. Hence, her viral illness can serve as a clue towards spinal pathology.

4. <u>Show your working (like A level maths - management process > diagnosis)</u>

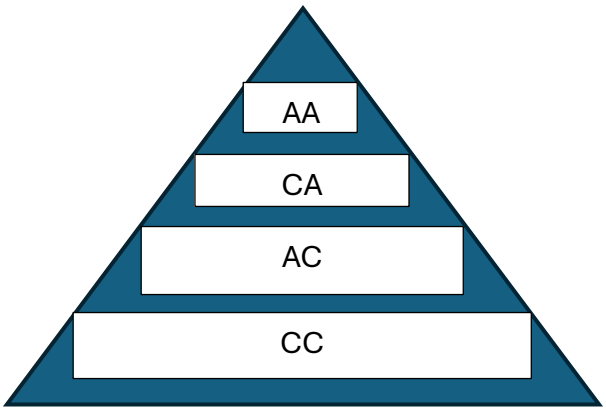
It is difficult to focus on a particular diagnosis in this case (given the lack of associated signs with the paraesthesia). As such, ***the step-by-step management is far more important.*** *Blood tests have effectively ruled out common causes of sensory peripheral neuropathy. As such, the next step is to rule out central causes with an MRI.*

*Ascending paraesthesia after viral illness should prompt consideration of GBS. Importantly, however****, if we focused solely on the diagnosis of GBS rather the management process then we are at risk of not doing / delaying the MRI spine which yielded the diagnosis.***

5. <u>Hallmark features in the clinical context</u>

I think I remember hearing that on taking a hot bath – the patient's paraesthesia worsened – this is the ***Uhthoff's phenomenon which is classical in MS.***

6. Pyramid of likelihood (common conditions are common)



C = Common
A = Atypical

In medicine, the statistical likelihood vastly drops the more atypical the disease & presentation. In this case – MS (200+ per 100,000) is statistically more likely than GBS (1-2 per 100,000). Although the patient's presentation is more common for GBS, the likelihood of MS is higher hence MS (AC = atypical presentation of a common condition) is still more likely than GBS (CA = common presentation of an atypical condition). Additionally, the young age & female gender in this case increases the likelihood of MS.

7. Trajectory focus rather than diagnostic obsession

Diagnostic obsession can sometimes be dangerous as it increases the risk of tunnel vision – i.e. focusing on only 1 or 2 diagnoses (e.g. GBS) rather than the trajectory of the patient's symptoms.

In this case, there are 3 trajectories to consider above diagnostic scratching.

Firstly, investigating the ***progression of her paraesthesia****. Often in medicine****, time & monitoring is as important as the number of tests that we do.*** *If the patient's paraesthesia was settling and she was found to be diabetic or vitamin deficient then we can be more reassured. However, her paraesthesia is progressing upwards and her bloods are unremarkable – this trajectory leads towards further urgent investigation.*

Secondly, monitoring the trajectory of her neurology. The lack of motor involvement is reassuring at that current time but we do not know if it will

progress. Therefore, in this patient, ***daily monitoring through neuro exams is as important as the tests itself!*** *Deterioration from motor weakness in both GBS and MS can lead to serious consequences if not acted on early.*

Thirdly, ***the age and background of the patient.*** *In this case, given her young age and good background – missing a diagnosis or deterioration can be catastrophic, whereas if the patient was 90 years old with frailty & multiple co-morbidities – a pragmatic & conservative approach may be in the patient's best interests.*

Structured Teaching from this case

5 red flags of neurological presentation (GP/ED/AMU setting):

1. Speech, swallow, breathing impairments - in the absence of resp / gastro cause – this should always raise suspicion for potentially rapidly progressive neuro pathology such as MND, MG, GBS and stroke.

2. Fasciculations – look at the patient's tongue, arms, legs and posterior trunk; if present then this could indicate MND esp. with the above impairments.

3. Ptosis – myasthenia gravis (MG) until proven otherwise esp. with fatigability and complex ophthalmoplegia.

4. Ascending weakness with absent ankle reflexes – strongly suggestive of GBS.

5. Spinal cord compression – specific signs are impaired proprioception (best marker), saddle paraesthesia, anal tone and sensory level. Beware that the severity of motor weakness does not always correlate to the cord compression pathology.

If any of the above are present then seek urgent Neurology review and monitor closely inc. FVC. Respiratory muscle weakness from a neurological condition can deteriorate rapidly hence consider ITU escalation if appropriate. If features of cord compression then urgent MRI spine (vet with Radiology) and inform Neurosurgery to be on standby.

MS – Important Facts

You are probably all aware of the relapse-remitting, secondary progressive and primary progressive subtypes. MS can present with various signs dependent on where the lesion is - but optic neuritis, brain stem and spinal cord lesions are most common.

Clinically Isolated Demyelination Syndrome (CIDS) refers to the first presentation of demyelination. Importantly, **not all patients with CIDS develop MS – around 40% do not develop further lesions.**

As such, it's possible that the patient in this case presented with CIDS (esp. given she denies previous episodes). MRI scans are important in this regard – if it shows more than one demyelinating lesion then it is also possible that the patient may have had a previous transient episode (which she did not come to hospital for).

Steroids do not impact on disease progression – they only speed recovery in MS (note evidence is slightly different for variants such as neuromyelitis optica). This explains why some patients have had previous transient episodes which self-resolved prior to needing hospitalization and why neurologists may not treat symptoms which are settling.

A little bit on Transverse Myelitis

Inflammation of the spinal cord. Clinical features depend on whether the motor, sensory or autonomic pathways are affected. Back & radicular pain is also common.

Importantly**, transverse myelitis is not the diagnosis – it is usually secondary to an underlying pathology** which has caused spinal inflammation. This can be due to demyelination (i.e. MS), infection, autoimmune, paraneoplastic, ischaemic or idiopathic causes.

Case 2 Summary

Young lady (in her 20s) with nil PMH or regular medication / ETOH presented to SDEC with a collapse. She described feeling dizzy in the kitchen – leaned onto her boyfriend for a few seconds who lowered her onto the floor. A seizure-like episode occurred for around 2 mins followed by a period of drowsiness (undetermined time). CT head in ED was NAD. She described having a vagal episode 1 year ago. There was a FH of Wolf-Parkinson-White (WPW) syndrome. ECG showed RBBB.

She was managed with Neurology & Cardiology reviews. Neurology advised for EEG & MRI with follow-up. Cardiology advised for reveal device monitoring with echocardiogram being unremarkable.

Heuristics (thinking tools) from this case

1. Hallmark features in clinical context

   This is a classical example of a 'muddy water' case where several pathologies are possible with no clear single cause.

   Let's look at the hallmark features of the possible pathologies:

   - *Cardiac – RBBB, FH of WPW, convulsive syncope can occur*
   - *Seizure - tonic clonic & post-ictal; both unclear from history*
   - *Vagal – preceding dizziness but no clear provoking factor*

   Based on this, we can see that cardiac syncope esp. ARVC is the most concerning pathology to rule out.

2. Disproportionate signs

   Perhaps the most important heuristic in this case ***– RBBB as the disproportionate sign.*** *Often, in medicine, there are one or two disproportionate signs which completely changes the way we perceive and manage a case. If not for the RBBB (and FH); it is likely that the patient may have been managed with an outpatient holter monitor and safety-netting.*

   RBBB alone can be an idiopathic finding or a sign of obesity or smoking. However, in the context of syncope in a young patient – this warrants further investigation which explains the reveal device insertion by Cardiology.

3. Hypothesis A vs Hypothesis B (where hypothesis A is the focus)

   A helpful way to approach cases is by structuring reasoning into a main hypothesis A which is the more likely and the less likely into hypothesis B.

In this case – hypothesis A would be cardiac syncope whilst hypothesis B would be epilepsy. Seizure is less likely given the lack of specific signs and the fact that seizure-like movements (convulsion syncope) are known to occur in cardiac causes due to cerebral hypoperfusion.

Although a vagal episode is most likely on statistical balance – it is prudent to rule out the more serious pathologies before this is labelled.

4. Trajectory focus rather than diagnostic obsession

   The implantation of a reveal device and neurological tests are reflective of trajectory planning rather than diagnostic hunting. The interest does not lie in finding out which specific type of cardiomyopathy or epilepsy the patient has – it is more about risk planning to ensure safety.

   As such, as clinicians working in GP/ED/AMU, a lot of the time we must accept we may not arrive at the diagnosis and instead focus on ensuring safe trajectory planning.

5. Practical approach (not always protocol or pattern-based approach)

   We see countless patients (esp. in SDEC) with syncope due to a vagal / postural cause. We must be careful that our fast protocol / pattern approach does not stop us from seeing a different presentation that warrants further attention using a practical approach (preventing a case of the boy who cried wolf).

Structured Teaching from this case

PASSOUT (mnemonic for causes of collapse)

Postural / vagal – benign & most common. 3 Ps of provoking, preceding, postural.

Arrhythmia – such as type II Mobitz or complete heart block, tri-fascicular block etc.

Seizure – either true epileptiform or non-epileptic attack disorder (NEAD)

Sugar – hypoglycaemic episode

Output – reduced cardiac output which can be due to saddle PE, structural (HOCM, ARVC), valve (critical AS), end stage HF and acute MI.

Unusual – rare causes (as always in medicine)

Transient – neurological due to bleed which can be SDH, SAH, ICH; prognosis worsens in this order.

Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)

ARVC is a cause of reduced cardiac output. It is an inherited cardiomyopathy in which normal RV muscle tissue is replaced by fibrous tissue. It can cause cardiac symptoms including syncope and sudden death in the young. ECGs often show bundle branch block, T-wave inversions in V1-3. A normal echo does not rule out ARVC as it could be in its mild stage with cardiac MRI usually being the definitive test.

Seizure

Seizure is not a diagnosis – it is a sign that the **brain is unhappy** (abnormal electrical discharge). Pathology both inside and outside the brain can lead to seizures.

Think of seizures as thresholds – a seizure occurs once the threshold is reached.

A normal person has a high seizure threshold – i.e. it would take a severe pathology to cause a seizure such as encephalitis, traumatic brain injury, haemorrhage etc.

An elderly / co-morbid person (e.g. dementia, cerebrovascular disease, cerebral palsy) has a low seizure threshold – i.e. it could take a small pathology to cause a seizure such as constipation, dehydration, infection etc.

An otherwise healthy person with a low seizure threshold – primary epilepsy due to congenital or structural reasons in the brain.

Differentiating between GTC seizure vs functional NEAD:

- Look closely at the seizure activity – true seizures have equal rigidity and rhythmic jerking of all 4 limbs.
- Lateral tongue biting with molar incisions is specific whilst tip of the tongue biting and urinary incontinence is not.

- GTC seizures have constant impaired consciousness throughout with no responses to any external stimuli throughout the seizure.
- Post ictal phase should last >15 mins.
- Haemodynamic instability occurs with ABG showing hypoxia & raised lactate esp. in GTC seizures >2 mins.

Factors when considering anti-epileptic drugs (AEDs):

- Trigger for the seizure (e.g. brain pathology)
- Clarify if true seizure (exclude functional or rigors from sepsis)
- Severity of seizure (focal vs GTC vs status)
- Number of seizures (single vs multiple)
- Risk balance (seizure risk vs AED side effects)

For example – AEDs would not be given in an alcohol withdrawal seizure or a functional seizure. If an elderly frail patient with cerebrovascular disease had a single focal seizure due to LRTI – you could consider that AED could potentially bring more harmful SEs.

Indications for starting AEDs:

- More than one seizure
- One seizure if structural brain pathology
- One seizure if severe / status
- One seizure if the risk of further was deemed unacceptable (e.g. occupation)

For the patient in this case, it could be reasonable to delay starting AEDs given she has had 1 seizure without severe consequences and a structurally normal CT. The teratogenic risks and side effects in a young female patient may outweigh benefits at this stage, hence neurology may decide in first fit clinic follow-up.

Case 3 Summary (short discussion)

45-year-old lady was referred by her GP to AMU with 2/12 of feeling generally unwell which had markedly worsened over 2/52. She admitted to vomiting but denied any weight loss. Blood tests showed significantly deranged LFTs (INR, albumin, ALP, ALT, bilirubin) with AKI 3 and IDA. She had a significant alcohol history but had stopped drinking 10 days ago. 3/12 prior to this admission, she had been reviewed in SDEC for IDA which was presumed secondary to menorrhagia hence discharged after IV iron.

On assessment, she appeared icteric but was not in withdrawal with no clinical signs of acute liver decompensation. She had tender hepatomegaly but no signs of ascites. Alongside supportive treatment, an USS abdomen was requested (given her AKI).

Later, CT CAP reported metastatic cervical cancer with inguinal lymph node spread and significant liver mets.

Heuristics (thinking tools) from this case

1. Show your working (management process > diagnosis)

   The diagnosis of metastatic cervical cancer was an unexpected finding. The clinical presentation would appear more in keeping with acute liver failure from alcohol use (with abrupt withdrawal effects) or malignancy from a liver / GI source. This highlights the importance of focusing on management steps over diagnosis.

2. Practical approach (not always protocol or pattern-based approach)

   What later transpired was the fact that the patient had inter-menstrual bleed as the cause of her IDA in SDEC 3/12 prior to admission – in retrospect this does explain her diagnosis of metastatic cervical cancer. Although we do not know the circumstances of the patient's SDEC assessment – this case demonstrates the ***importance of a practical approach over protocol / pattern approach*** *(it would be easy to label IDA as menorrhagia given the large numbers of patients that pass through SDEC with this).*

   Focusing on the history & examination is therefore absolutely crucial *– this would increase the chances of finding out from the patient that her inter-menstrual bleeding was the cause of her IDA.*

3. Trajectory focus rather than diagnostic obsession

   Given the unexpected diagnosis in the context of her clinical presentation – it is far ***more important to focus on her trajectory of potential decline from her acute liver and renal failure than diagnostic hunting.*** *Prioritising management of her organ failure over scanning would not have changed her overall outcome (arguably would have improved her status in the acute setting).*

As such, the decision to do an USS over a CT given her renal failure (risk of contrast nephropathy) was the right decision at that time, even if it was the CT that eventually yielded the diagnosis.

Structured Teaching from this case

Liver Disease (Rule of 3)

3 stages of liver disease:

- Fatty liver
- Steatosis
- Cirrhosis – irreversible stage

3 aetiologies of liver disease:

- Alcohol related liver disease
- Metabolic dysfunction related liver disease
- Specific aetiology related liver disease

Alcohol and metabolic dysfunction related liver disease – reflects the increasing combined incidence of the two.

Specific aetiology related liver disease – this is the liver panel to check for viral hepatitis, autoimmune hepatitis, hereditary haemochromatosis, Wilson's disease, hepatocellular carcinoma, alpha-antitrypsin deficiency, primary sclerosing cholangitis (PSC).

3 clinical signs of liver decompensation (AAA):

- Asterixis (liver flap)
- Altered consciousness (hepatic encephalopathy)
- Ascites (shifting dullness)

Decompensation only occurs in liver cirrhosis. It can affect any or all of the blood tests (albumin, INR, bilirubin, platelets, ALP, ALT) but most common is raised INR or bilirubin.

3 complications of liver decompensation:

- Variceal UGI bleed (portal HTN)
- SBP (infected ascites)
- Hepato-renal syndrome (end-stage)

3 treatments of liver decompensation:

- Treat the underlying cause
- Clear the bowels (lactulose)
- Supportive therapy for organs