A patient that changed my practice

not the Guillain–Barré syndrome

Myles Connor
Division of Neurology, Department of Neurosciences, University of the Witwatersand, Johannesburg, South Africa;
Email: connorm@chiron.wits.ac.za
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Taking over a colleague's patient is not easy. The handover time is fraught with potential pitfalls, particularly if the clinician is inexperienced. As a neurology trainee I was involved in the management, or rather mismanagement, of a patient and learned to be vigilant at the time of handover, particularly when things do not seem to be quite what they should be.

THE STORY

The patient, a 69-year-old woman, was to be transferred to a rehabilitation facility the day I joined the unit. The diagnosis of Guillain–Barré syndrome was based on clinical features and albumino-cytological dissociation in the cerebrospinal fluid. The hospital's neurophysiologist was on leave and the senior trainee had performed the nerve conduction study. He felt the findings were compatible with an acute demyelinating and axonal polyneuropathy. The patient had received intravenous immunoglobulin and, over the week prior to discharge, with the physiotherapist's help she had made a significant improvement. She had responded to treatment and so the diagnosis was sealed. Transport was arranged and everyone was happy with her move to the rehabilitation unit. But, something on the brief examination during the morning round just didn't quite fit. She had a wrist drop out of proportion to the rest of her weakness, and marked asymmetry of her reflexes. I inspected the notes carefully.

She had been admitted to a district general hospital 2 weeks prior to being transferred to our hospital. She had complained of a week-long history of neck pain and weakness, initially of her upper limbs and neck, and later her lower limbs. The weakness had progressed over 2 weeks, was proximal at first, and had then moved distally. All this followed a week after a bout of diarrhoea, during which her general practitioner noted blood and protein in her urine. He had diagnosed a urinary tract infection and treated her with antibiotics.

On admission, she had reported some loss of sensation of her left arm and leg, and her right fingertips. She also complained of occasional, nonspecific diplopia, associated with light-headedness, which occurred when sitting up. There was no bladder or bowel disturbance. She gave no history of immunisations, foreign travel, surgery, exposure to lead or precipitants of the Guillain–Barré syndrome, apart from the bout of diarrhoea. Twelve years previously she had two basal cell carcinomas removed and at the age of 39 had an appendectomy and sterilisation. She was on no medication and smoked 10 cigarettes a day.

ON EXAMINATION AT OUR HOSPITAL

The patient was pale and had a mottled discolouration of both forearms. She had no lymphadenopathy and the rest of the general examination was normal. In particular there was no evidence of autonomic instability and her vital capacity was normal.

On neurological examination, she was alert and co-operative. Her eye movements were normal and she had very mild neck flexion weakness. Tone was normal-to-decreased and she had grade 4 power proximally in her right arm and both legs, deteriorating to grade 3 distally. Her left arm was similarly involved but she had only grade 2 power on flexion and extension of the wrist. All the reflexes were diminished but present, apart from absent left biceps, triceps and supinator reflexes. She had decreased light touch, pain and temperature sensation over the left hand and left foot.

In spite of the marked motor asymmetry, unusual onset, asymmetrical sensory involvement and preserved and asymmetric reflexes, she was thought to have the Guillain–Barré syndrome, though mononeuritis multiplex was included as a differential diagnosis.

The investigations are summarised in Table 1. She was treated with intravenous immunoglobulin and by day four was able to mobilize with assistance. By day 11 she was able to walk unaided using a Zimmer frame.

Despite my concerns regarding the diagnosis, I conceded that she had made a dramatic recovery and that the autoimmune screen, including antineutrophil cytoplasmic antibodies, was normal. I allowed her to be transferred.

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RE-ADMISSION TO HOSPITAL

She was readmitted 15 days later. Her mobility had deteriorated over the previous week and she was no longer able to swallow. On examination she was dehydrated, had angular stomatitis and a swollen right parotid gland. Her chest and vital capacity were normal. Neurologically, she now definitely had neck flexion weakness and the power in her arms was grade 3 on the right, worse distally, and on the left grade 4 proximally and 0 distally. Both legs were weak proximally (grade 3–4) but, on the left, dorsiflexion at the ankle was absent. All reflexes were absent apart from the right knee, left triceps and right biceps, which were decreased but present. The plantar response was flexor. She had a glove and stock-

Table 1 Investigation results

Investigation	First admission	Second admission	Normal values
Haemoglobin	9.7 g/dL (normochromic, normocytic)	10.1 g/dL	12–16 g/dL
White cell count	18 □ 10^9/L	27 □ 10^9/L	3.9–9.8 □ 10^9/L
Neutrophil count	14 □ 10^9/L	23 □ 10^9/L	2–7.5 □ 10^9/L
Lymphocyte count	3 □ 10^9/L	–	1–4 □ 10^9/L
Erythrocyte sedimentation rate (ESR)	–	68 mm in 1 h	1–15 mm in 1 h
International normalized ratio (INR)	–	2.35	–
Sodium	135 mmo/l	138 mmol/L	135–147 mmol/L
Potassium	3.4 mmol/L	2.8 mmol/L	3.3–5.3 mmol/L
Urea	2.4 mmol/L	6.4 mmol/L	2.6–7 mmol/L
Creatinine	40 µmol/L	74 µmol/L	40–140 µmol/L
C-Reactive Protein (CRP)	20 mg/dL	–	0–10 mg/dL
Total protein	–	51 g/L	60–85 g/L
Albumin	16 g/L	23 g/L	35–52 g/L
Total bilirubin	7 µmol/L	12 µmol/L	0–21 µmol/L
Alkaline phosphatase	121 U/L	122 U/L	40–120 U/L
Alanine transaminase	38 U/L	27 U/L	5–4 U/L
Gamma glutamyl transferase	–	13 U/L	0–60 U/L
Antinuclear factor (ANF)	Negative		
Antineutrophil cytoplasmic antibodies (ANCA)	Negative		
Serology	Negative for: Ebstein-Barr virus, cytomegalovirus, varicella zoster virus, herpes simplex I & II, *mycoplasma pneumoniae*, *campylobacter jejuni*, influenza A & B, *chlamydia pneumoniae*, *coxiella burnetti*		
Electrocardiograph	Normal	Normal	
Chest X-ray	Normal	Normal	
MRI cervical spine	Normal	–	

ing sensory loss bilaterally. The investigation results are summarised in the Table 1.

She was treated supportively, given antibiotics for presumed sepsis, vitamin K to correct the INR and oxygen, and the electrolyte abnormalities were corrected. She continued to deteriorate over the hours following admission, developed a metabolic acidosis, and was transferred to the intensive therapy unit (ITU). A nerve biopsy was performed and corticosteroids administered. The following day, she developed an acute abdomen and was taken to the operating theatre. The surgeons found an extensive segment of infarcted bowel. Back in the ITU, she suffered a cardiorespiratory arrest. Resuscitation was unsuccessful and she died.

The postmortem confirmed the nerve and bowel histology of polyarteritis nodosa. (Figure 1)

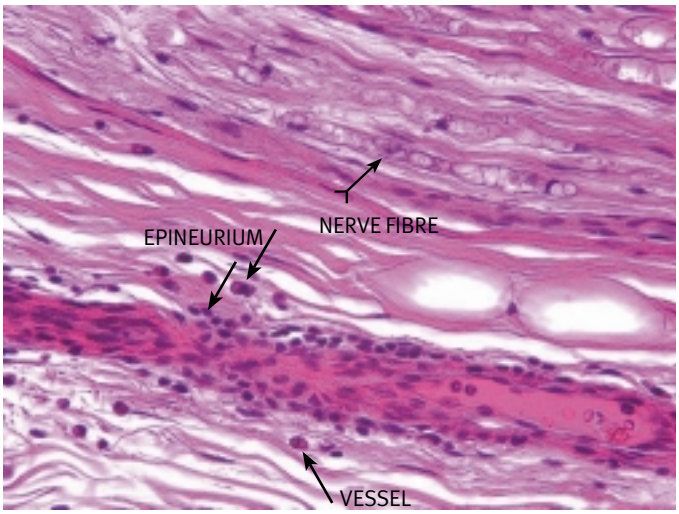

Figure 1 Nerve specimen showing vasculitis (H and E stain). There are inflamed vessels in the epineurium associated with plasma cells (arrows) and lymphocytes (short arrow). The nerve fibres are severely damaged and infiltrated by macrophages (arrow with tail).

whilst Guillain–Barré syndrome may present atypically and have an unusual onset, retained reflexes and some sensory signs, the clinician always needs to look for other causes of an acute polyneuropathy

CONCLUSIONS

I learned a number of valuable lessons from the events surrounding this patient's admissions and outcome. Firstly, whilst Guillain–Barré syndrome may present atypically and have an unusual onset, retained reflexes and some sensory signs, the clinician always needs to look for other causes of an acute polyneuropathy. These must include the causes of mononeuritis multiplex, particularly if the clinical signs are asymmetrical. Negative antinuclear factor and antineutrophil cytoplasmic antibody results do not exclude a vasculitis such as polyarteritis nodosa and, if clinically suspected, a nerve biopsy should be performed. The anaemia and low serum albumin at the first admission were perhaps a clue to something other than an 'ordinary' Guillain–Barré syndrome. An ESR was not available during the first admission, if it had been performed and was as raised as it was in the second admission, alarm bells might have rung and alternative diagnoses pursued. Finally, beware of the situation where the patient is about to be discharged at the time you assume responsibility for their management, particularly when you are uneasy with the stated diagnosis. Lastly, ensure that the diagnosis fits the patient – don't force the patient to fit the diagnosis.

PRACTICE POINTS

- Beware of the 'atypical' Guillain–Barré syndrome.
- Negative autoantibodies do not exclude polyarteritis nodosa.
- Be particularly vigilant when you take over a patient's management, even more so if the diagnosis doesn't quite fit the patient