ME AND MY NEUROLOGICAL ILLNESS

Chronic inflammatory demyelinating polyradiculopathy responding

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Having retired from general practice 4 years ago I am writing an account of an unusual disease responding to an unusual treatment. Inexplicable bouts of evening fever occurred through 1998 when I was 57-years-old. At first I did not associate these with the onset of bilateral ulnar nerve paraesthesiae in early 1999. My partners had kindly purchased a lap top computer for me so that I could also work as practice manager. I had spent hours learning to use it, sitting in an awkward position, so I assumed I had a compression neuropathy of the ulnar nerves. I continued life as normal, working hard, riding my horse on my half day off, and running a watch repair business in the evenings (I had qualified as a watchmaker 25 years earlier). I ignored the numbness that developed on the lateral borders

of both feet because I had a previous history of lumbar disc disorder.

At the end of 2000 I began to notice unusual fatigue and shortness of breath when ascending stairs to see patients, eventually I found it impossible to do a full day's work without a rest in the afternoon. I took myself off for routine bloods and discovered I had a haemoglobin of 9.2g/dL and a serum creatinine of 250 $\mu\text{mol/L}$. I consulted my own general practitioner and went to see a renal physician, and a neurologist who told me I had a compression neuropathy. Relieved that I did not have motor neuron disease, I failed to query the unusual fact that I apparently had four separate nerves being compressed.

Investigations by the renal physician, which included a vasculitis screen and renal biopsy,

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failed to find the cause of my small contracted kidneys with chronic interstitial change. I retired in May 2001. Erythropoietin improved my anaemia and fatigue, I felt well, took my Border collie dog for a long walk on the marshes every morning and worked full time in my watch repair business. I played at the bridge club on Monday afternoons and attended Rotary lunches on Wednesdays. Life was great; my renal impairment deteriorated only slowly and the repair business was thriving.

One morning in July 2002, walking back from the marshes, my legs failed to move forwards. Fortunately my Border collie was already back on her lead – she went into power-drive walk and towed me home. I managed to hobble at home with a walking stick but was not safe

enough to walk the dog. One evening when taking off my jersey I fell over when my vision was obscured, thereafter I had to undress whilst sitting down. The occasional evening chills and fevers continued.

Shortly after all this, at a routine appointment with the renal physician, loss of position sense and sensory ataxia were confirmed leading to a rapid appointment with another neurologist. His clinical examination showed total absence of any tendon reflexes, symmetrical long sock and short glove sensory loss with gross sensory ataxia and weakness in several muscle groups leading to a diagnosis of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), later confirmed by nerve conduction studies. Blood was analysed to exclude other causes of peripheral neuropathy, including protein electrophoresis to exclude paraproteinaemic demyelinating neuropathy.

Treatment started with prednisolone and azathioprine, which gave an initial, but unsustained benefit. The neuropathic pains then started. They began after lunch every day, it became uncomfortable to sit in my armchair, my buttocks were tender and the pain spread down the back of my thighs. By the time the news on TV started at 6 o'clock I had to stand or kneel to watch it, in the early evening the pain increased in intensity causing me to lie on my face on the sofa, crying out at times with the pain, achieving temporary relief by gently slapping my thigh. Eventually I would go to bed with the strongest analgesic I could find and sleep for perhaps an hour. I would then awake with increasingly severe pain also affecting my upper arms. Analgesics had no effect at this stage, sleep evaded me, at times I would cry out with the pain, at other times weep. By 2 a.m. the pain would subside allowing me to sleep. In the morning I would awake at seven, gloriously free from the pain, so much so that at my morning appointments with the neurologist. I would not even mention it, yet it returned with equal savagery day after day. Finally after some months of pain I was admitted to a neurological ward for a sural nerve biopsy. I was not impressed by the skill of the junior surgeon undertaking the procedure so I was not surprised when the biopsy failed to show any significant abnormality. During my inpatient stay I asked the resident house officer if I could try gabapentin. The result was dramatic, I have remained free from pain ever since.

Life that winter was complicated by the sodium losing nephropathy; on two separate occasions I

lost 10 kg due to the associated water loss leading to dehydration, postural hypotension and vomiting. Eventually the excessive salt replacement put me into cardiac failure. The relentless low-grade fever continued every evening for over 8 weeks resulting in full investigation for fever of unknown origin, but no cause was found. Recovery was further delayed by MRSA septicaemia resulting from the presumably unclean insertion of a central line for emergency haemodialysis.

By early 2003 the renal failure was well managed by haemodialysis giving good biochemical control, but the CIDP progressively deteriorated: each week I lost a function that had been present the previous week. In addition to the bilateral ulnar neuropathy, both median and radial nerves were involved leaving just a little strength in abduction of my thumbs while the other muscles in my hands supplied by these three nerves were paralysed. There was complete sensory loss in a short glove distribution. My hands functioned like a pair of flippers, no grip was possible. Axonal damage had led to severe wasting of the small muscles in my hands. I had to use a variety of practical aids for feeding and washing, my wife dressed me. I managed to move about indoors with a walking frame, I could attend Rotary lunches with help, but could no longer grip a card or use watch tools. There were still thirty partially completed repair jobs, and I shall be eternally grateful to the four watch repairers who completed those jobs for me.

Through 2003 prednisolone, azathioprine, intravenous immunoglobulin, plasma exchange, cyclosporin and mycophenolate were all tried; none of them arrested the relentless progression of the disease. The renal failure was well controlled by haemodialysis, the only limb muscles that continued to work were the upper arm muscles enabling me to use a powered wheel chair and to transfer with a sliding board. Social services turned up trumps, they supplied a powered hoist and carers every morning and evening to wash and dress me. A slotted strap round my hand held a spoon allowing me to transfer cut-up food to my mouth. The computer was now my lifeline, using the keyboard with one fatigued index finger became arduous; so I installed a voice-operated program which after perseverance became a very effective tool.

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CIDP, is a sensation of utter isolation and loneliness; none of the seven medical partners in my general practice had heard of it, how do you explain it to friends? A search of the web brought me to two sites <http://www.gbsfi.com> and <http://www.gbs.org.uk> on which there are many expert patients and a wealth of knowledge. There are often medical, or anatomical queries so I log on every morning to do a 'cyber surgery'. The voice operated program allowed me to continue in spite of almost complete quadriplegia. I raised a query about unexplained fever and have now had twelve replies from others in many parts of the world in whom fever of unknown origin occurred during the active phase of CIDP, many of them were also fully investigated for a cause of the fever, though none of their physicians associated it, as I do, with the active auto-immune destructive process.

Realising that I had probably reached the nadir of my disease, I accepted that this was how the remainder of my life would be, one which in view of my family history and the renal failure was likely to be brought to a close by a myocardial infarct. The major strain was on my wife as principal carer for most of the day and night, there is no official support to which a carer can turn. In a way, we were fortunate to have two friends also limited to wheel chairs; their partners were able to give some support and advice. Renal failure turned out to be a blessing; besides no longer having the inconvenience of passing urine from a quadriplegic body, 3 days away every week in the care of renal dialysis nurses relieved my wife of responsibility on those days. As in all chronic diseases, attitude is of prime importance. A cheerful face in adversity reflects on all who come into contact, so much so that people begin to seek contact. I was fortunate in being able to do the 'cyber surgery' each morning, giving me a purpose in life and the facility for helping others.

After discussion with a neurologist more experienced in CIDP, the neurologist I was seeing suggested the use of Rituximab, an antibody against the CD20 antigen found on the surface of B cell lymphocytes. Its use is established in B cell lymphoma and has shown promise in patients with the neuropathy associated with IgM paraproteinaemia. The possible adverse ef-

fects were fairly horrific, but I really had little to lose and no other options. I was started on intravenous infusions of 700 mg of Rituximab, which were repeated every 3 weeks until I had had seven doses. I could hardly believe the transformation. After the first dose I could partly flex my hand, a movement previously impossible as my hand had been paralysed. After 3 months muscle strength recovered in my legs, fatigue diminished and disappeared, the ataxia was less and the pain was reduced.

I installed parallel bars and employed a neurologically trained physiotherapist. Six months after starting Rituximab I could walk with a frame, wash and dress myself and hold normal cutlery. Soon finger-thumb grip returned so that I could play cards. The crowning achievement was to walk my daughter down the aisle at her wedding.

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