... through a left thoracotomy ...

Myasthenia gravis

David Shepherd
Consultant Neurologist, 12 Stones Drive, Ripponden, West Yorkshire, UK; E-mail: divyhily@stones1944.

Practical Neurology, 2003, 3, 184–185

In the past, at Association of British Neurologists’ dinners, I accepted that I was usually the only neurologist in Highland evening dress, but to that idiosyncrasy I can now add the less pleasurable attribute of a malignant thymoma and myasthenia gravis.

Well how did it all begin? With a cough, a dry, brief, non-irritating cough, which started as far back as 1993 and as we will see disappeared after surgery. Around the same time I began to accumulate crumbs in the vestibule of my mouth, external to the teeth and inside the cheek, easier to extract digitally than with my tongue and worse on the left. Orofacial surgeons do describe some sort of rarefocal facial muscular weakness; perhaps that was it, I thought! I used to dictate three medicolegal reports back-to-back and after filling a 1-hour tape, on occasion from 1996 onwards noted that I found it easier to articulate by leaning back in a chair. This disappeared in the summer of 1997 for the best part of a year. At the end of a long clinic dictating to my secretary whilst consuming a cup of coffee and well-earned biscuit, at times I felt as if I might choke and that my speech was strained. She was the only person to notice anything wrong with my voice prior to the summer of 1998. An ear, nose and throat colleague kindly excluded as the cause polyps, adenoids, etc.

On 24 August 1998 I gave a talk to a Multiple Sclerosis Society and felt that my voice was strained and nasal in quality, but no one, even on questioning, noticed anything amiss. On 14 September whilst on a slip road joining a motorway I got brief fluctuating horizontal diplopia on extreme right lateral gaze. In retrospect it must have been ataxic nystagmus. Other occasional similar episodes followed. In October, whilst in Venice, brief episodes of vertical diplopia occurred and quite vicious positional vertigo (occasional episodes of which I had had going back many years and assumed to be benign). Multiple sclerosis seemed more likely than myasthenia at this juncture.

I gave my last lecture on 18 November to general practitioners. I found it more and more difficult to speak, but the audience noted nothing. I sought relief by asking them questions and realized that my own silence allowed partial restoration of articulation. By now it was time for some investigation and auto-antibody tests were sent in early November. There was modest anxiety, impatience and mounting irritation when 4 weeks later there was no trace of the specimens! No apology was ever forthcoming, no anti-acetylcholine receptor antibody result was found, and yet a negative anti-skeletal muscle result appeared many weeks later. On 4 December further specimens were sent to a different establishment (neurology trainees do get nervous venesecting consultants!). The blackest moment of all was on Friday 11 December when a strongly positive skeletal muscle antibody returned thus portending gloom. I arranged for a thoracic CT on 14 December and from the moment contrast was administered, I anticipated the result. A large calcified mediastinal mass and two calcified lesions in the left pleura. Shortly afterwards the acetylcholine receptor antibody result returned strongly positive.

Self-diagnosis after 5 years was complete and now was the time to choose my management team, or who was going to get the short straw!
The neurologist had to be someone considerably younger than me in the hope of management over a prolonged time course. The surgeon chose himself, being the only pure thoracic surgeon at my Regional Centre and a colleague of the neurologist. My, by now dysarthric, opening words to my general practitioner were ‘My notes are remarkably slim but the file will expand significantly’. He signed me off forthwith. By early January 1999 I had intermittent increasing dysphagia, dysarthria and facial weakness, but strangely no diplopia.

The immediate preoperative period was characterized by increasing family concern, but personal resignation. Pulmonary function tests were surprisingly physically challenging even though the results were then normal. Informed consent involved the understatement, ‘The lesion is a bit near the clockwork’.

28 January 1999: through a left thoracotomy three, not two, pleural lesions were discovered and resected and a 106 g calcified mediastinal thymoma through which traversed the phrenic nerve, which was sacrificed (hence the origin of my cough for the previous 5 years). Certainly there was postoperative euphoria, as I was myasthenically asymptomatic and pain free, but this was somewhat tempered by the necessity for urinary catheterization (morphine has no effect on this). I had three areas of numbness: the right radial terminal branch supply related to the infra-arterial line, the whole left side of the abdominal wall anteriorly due to stretched or crushed intercostal nerves, and the left T2 distribution in the upper arm presumably due to peri-operative positioning. The last resolved rapidly and the former two over many months, but I have never mentioned any of them to medical staff.

Three days after surgery a bout of atrial fibrillation occurred and though uncomfortable resolved rapidly with beta blockers. I had quite spectacular morphine-induced hallucinations, numerous ants parading out of the suspended ceilings and down the walls when the lights were switched off. I was not convinced by the Chinese nurse’s reassurances, as my secretary’s department had recently been subjected to an invasion of pigeon mites from the ceiling – the product of decades of roosting, dying and decaying ‘cushie doos’ mixed with asbestos fibre in one of our less modern hospitals.

The transition from strong narcotics to paracetamol was abrupt and discomforting. To explain the palpable, audible, but painless, crepitus emanating from the ribs, the surgeon reassured me that, like the main character in ‘Captain Corelli’s Mandolin’, the fragments were bound together, but not with the strings of a musical instrument. The myasthenia was in complete remission for 3 weeks, but then within 2 weeks dysphagia, diplopia and dysarthria to the point of requiring periodic communication only by writing, occurred. Articulation was easier lying flat to counteract profound soft palate weakness. There was increasing facial weakness, proximal limb girdle weakness and specifically marked painless winging of the left scapula (why so focal?). Anticholinesterase medication, although effective for a short duration, induced gastrointestinal side-effects that were only partially contained. Intravenous immunoglobulin proved totally ineffective and oral alternate day prednisolone was slow to show its benefit. At my worst a very positive tension test was psychologically extremely reassuring, but quite astonished my wife. ‘D’ and ‘b’ were the most difficult letters to enunciate and of course my ever attendant nurse was called Debbie!

Moderate, but not sustained, improvement occurred after three sessions of plasmaphoresis. More lasting benefit took a further eight sessions and continued prednisolone. Increasing doses of azathioprine were poorly tolerated and after 8 weeks discontinued because of profound liver function test disturbance which then rapidly, but not completely settled. By August 1999 I was significantly improved, but decided to quit neurology in exchange for family genealogical terms such as umqhile, sasine, testaments and retours.

On steroids, I remained well for a further year but then deteriorated. The relapse began with vertical early morning diplopia for 15 minutes, soon followed by facial weakness and dysarthria. Increased steroid dosage proved largely ineffective after 3 months and it took the addition of a further ten sessions of plasmaphoresis over 2 months to achieve control. I remain in admiration of the haematology nursing staff for their skill, only one failed venepuncture in 21 plasmaphoresis sessions and all lasting for a full plasmapheresis. For me this treatment has been the mainstay of therapeutic benefit, noticeable within 12 hours and sustained for 7–10 days in the acute phase. This was in complete contrast to my experience as a neurologist where virtually no one had benefited. Since March 2001 my condition has been stable on a decreasing dose of alternate day prednisolone, but punctuated by occasional bouts of atrial fibrillation.

What have I learned? Automated sphygmomanometers produce grotesquely fallacious results. Nurses are not aware that in the presence of facial weakness vital capacity readings can be unreliable. Enteric-coated prednisolone is invariably provided irrespective of what is prescribed, and outside the hospital 25 mg prednisolone tablets are unobtainable. I inability to curl the tongue can be the first hint of tongue weakness, well before dysarthria. Alternate-day steroids produce daytime diuresis on the day of steroids and transient hoarseness in the late afternoon, and in addition severe nocturnal calf cramp. Unilateral phrenic nerve paralysis does mean that on bending half one’s abdominal contents migrate into the chest and my forced vital capacity has dropped by a third so I am more short of breath. Finally, every medication has a downside in terms of side-effects that can negate therapeutic benefit, something that I had not fully appreciated from the other side of the couch.