Dickens, dystonia and dyskinesia

The characters of Charles Dickens' novels have a plethora of pathology. The Pickwickian syndrome is well known. Sir Ruskin's main made some observations on neuropsychiatric disorders.1 Apart from these, it provides interest and amusement to diagnose other disabilities suffered by Dickens' characters.

Little Dorrit has characters with two types of motor disorder:

1. Jeremiah Flintwinch: focal dystonia with torticollis

The following are among the passages which leave little doubt of the diagnosis: "His head was awry; and he had a one-sided, crab-like way with him, as if his foundations had yielded at about the same time as the house, and he ought to have been propped up in a similar manner..." "His neck was so twisted that the knotted ends of his white cravat usually dangled under one ear... altogether he had a weird appearance of having hanged himself at one time or other, and of having gone about ever since, halter and all, exactly as some timely hand had cut him down...

"... Mr Flintwinch... with his neck twisted and one eye shut up, stood smoking..."

The neck was crookedly out of Mr Flintwinch's mouth, as if it circulated through the whole of his very figure and came back by his wry throat, before coming back to mingle with the smoke from the crooked chimneys and the mists from the crooked river...

"To support this diagnosis of torticollis with facial dyskinesia is the frequent observation that Flintwinch... "scraped his horny cheek...", or "... stood leaning against the wall scraping his jaw..." or "... un-pocketed one of his hands to scrape his jaw...". This frequent gesture may represent the "geste antagoniste" which relieves the dystonia by counter-pressure. The original illustrations by "Phiz" portray the torticollis and possibly the "geste antagoniste" (fig.).

2. Pancks, the debt collector: Gilles de la Tourette's syndrome, or multiple tic?

Mr Pancks was a hyperactive, eager, obsessive, chronically nail-biting young man who was always "perspiring and puffing and snorting about in eccentric directions."

There are numerous descriptions of his inappropriate vocalisations: "Mr Pancks here made a singular and startling noise, produced by a strong blowing effort in the region of the nose, unattended by any result but that of acoustic one..." "He blew off that sound again, and it occurred to his companion for the first time that it was his way of laughing... he might not have been quite in earnest, but the short hard rapid manner in which he shot out these principles, as if it were done by mechanical revolution, seemed irreconcilable with barter..." and "Mr Pancks acknowledged this recommendation with such an extraordinarily abrupt, short, and loud utterance of the monosyllable "Oh" that even the unwieldy Patriarch moved his blue eyes in something of a hurry to look at him."

The patient was a 22 year old right handed computer operator. In January 1986, while she was approximately three weeks post-partum from her first pregnancy and following about two weeks of an influenza-like illness, she awoke with pain and stiffness in the right shoulder and rapidly progressive weakness in the proximal right arm. She was referred to another hospital, where an electromyogram, performed in February 1986, showed increased insertional activity with positive sharp waves and fibrillation potentials in the right biceps and some degree of suppression of the left biceps, triceps, and lateral condylar muscles. There was minimal recruitment on voluntary effort in these muscles in the form of a few rapidly firing complex polyphasic potentials of large amplitude and long duration. Short trains of positive sharp waves were also seen in the right biceps. Motor nerve conduction velocity studies, including ulnar F waves from the right abductor digiti quinti muscle, were normal, as were sensory nerve action potentials from the right median nerve. Her physician concluded that she had upper brachial plexopathy and muscle strengthening exercises were prescribed.

She presented to our unit for the first time in April 1986. She reported some improvement in both strength and her feeling of numbness. She was unable to give a possible family history. On examination, she had obvious weakness of the right shoulder girdle, with mild to moderate atrophy of the infraspinatus, supraspinatus, and deltoid muscles. She showed no fasciculations. Strength was rated at 2/5 in the infraspinatus, supraspinatus, and biceps, 3/5 in the deltoid, 4/5 in the brachioradialis, and 5/5 elsewhere.

There was mildly decreased pinprick sensation over the lateral aspect of the upper arm and elbow. The remainder of the examination, including tendon reflexes, was entirely normal, except for the right biceps and brachioradialis reflexes, which were slightly reduced (2+ compared with 3+ elsewhere).

The patient steadily improved until the summer of 1986, when she reached a steady state, with a sensation of residual weakness without observable neurological deficit on the right side.

She did well until 1988, when she became pregnant for the second time, and delivered a second healthy child on 8 December 1988. Almost immediately after this uncomplicated delivery, the patient began to complain of weakness and soreness in the upper left arm, which gradually spread over a period of four weeks to include almost the entire left arm. Her physician prescribed methylprednisolone, and she tapered this off after a period of three weeks. Our group re-evaluated her on 20 January 1989, during the steroid taper. She had begun to improve, but continued to complain of left arm weakness. Her right arm showed normal bulk, tone, and strength. On the left side she had 4/5 strength for elbow flexion and supination. Sensory examination showed a mild reduction in pinprick along the outer aspect of the left arm, which was also noted on the left. Reflexes were symmetric, and her examination was otherwise normal.

The patient had a repeat electrophysiologic examination on 23 January 1989. Increased insertional activity with positive sharp waves and fibrillation potentials and decreased recruitment pattern were confined to the left biceps. Motor conduction velocity studies of the left median and ulnar nerves were normal. Sensory studies of the left median, left ulnar, and right and left radial nerves, were all normal. Stimulation at Erb's point elicited a
potential of 5 mv over the right and 2 mv over the left biceps with an onset latency of 4.1 ms and 4.2 ms respectively. Antidromic sensory stimulation of the lateral cutaneous nerve of the forearm evoked a potential of 5 uV at a peak latency of 2.8 ms on the left; the corresponding values for the right were 18 uV and 2.5 ms.

The patient was referred to a physiotherapist for restrengthening exercises. By 10 February, when last seen, she had further improved.

The modern history of brachial plexopathy described by Spillane at least 18 years before the brachial radiculitis among diabetics, 26 of whom developed the characteristic pain and neurological deficits while convalescing from other illnesses. The term "neuralgic amyotrophy" dates from 1948, when Parsonage and Turner described 136 British army personnel with this condition.

In this study, "66 of the 136 were in hospital with other conditions when the shoulder-girdle symptoms began, and others had recently recovered from illnesses. In 98 of the cases there was evidence of some precipitating factor." Later the same authors offered follow-up data on 82 patients, and noted that several nerves, most notably the anterior interosseous, were involved individually. The prognosis was good, most attacks resolving completely in two years, but "in a few cases attacks with a recurrence may occur many years after the first attack".

More recently, the spectrum of clinical presentation of this entity and its electrophysiological patterns of involvement have broadened. A familial form, associated with the pathological changes of tomaculous neuropathy, may be separate from the common sporadic form in which this pathological picture does not occur. The familial form does not have a recurrent pattern. With the sporadic form, precipitating conditions have included serum sickness, polio vaccination, tetanus and typhoid vaccinations, malaria, typhus, diabetes mellitus, rheumatic fever, tuberculous, trauma, hepatitis, and systemic lupus erythematosus.

The electrophysiological picture may show any of several described mononeuropathies or mononeuropathy multiplex or isolated areas of the brachial plexus proper. Isolated musculocutaneous nerve involvement, as seen in our patient's second episode, has only rarely been recorded.

Other inflammatory disorders of the nervous system have been associated with the puerperium. McCombe et al studied 61 patients with chronic inflammatory demyelinating polyradiculoneuropathy, nine of whom became pregnant. In all nine, either the onset of the neuropathy or a relapse of pre-existing neuropathy coincided with the pregnancy. They hypothesised that cross-reactivity to second and maternal neural antigens or an immune response due to pregnancy itself might explain this association.

To our knowledge, our patient is the first described in which recurrent alternating unilateral brachial plexopathy coincided with successive pregnancies or puerperal periods. The two quite different clinical and electrophysiological patterns which our patient's two attacks manifested conform to the diversity seen by others already cited in this entity. We have no explanation for the alternating, unilateral pattern of involvement in our patient.

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