What's in a name? The Bruns–Garland syndrome

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Today's diabetes world is fast-moving and exciting; knowledge is accumulating at an astonishing rate. To help understand the present, however, it sometimes helps to examine the past.

In this instalment of *Tattersall's Tales*, Robert Tattersall presents the history of Bruns–Garland syndrome, a form of neuropathy, from its first description by Ludwig Bruns in 1890 to that of Hugh Garland in 1960.

It is a common clinical perception that, like buses, certain conditions come in threes. In my experience this has been true of subarachnoid haemorrhage and diabetic amyotrophy. In the literature, amyotrophy also seems to have been the subject of triplets of papers separated by decades. In 1890 the German neurologist Ludwig Bruns (1858–1916) described three patients (aged 58, 59, and 70) with short-duration diabetes; all three developed severe pain in their hips and thighs, followed by a weakness and wasting of leg muscles without objective sensory loss (Bruns, 1890). All recovered fairly quickly on diet alone. In the same year, in the *British Medical Journal*, the London physician Thomas Buzzard (1831–1919) described JK, a 55-year-old carpenter, who:

> *'Began to get pain and tenderness in the front of the right thigh which extended presently down the leg to the foot. The pain was very severe and there was so much weakness that his knee often gave way and let him down. A month after the right the left leg was similarly attacked.'*

This was clearly neuropathic because 18 months later the patient had a perforating ulcer of the foot and gross wasting of the small muscles of the hand (Buzzard, 1890). In early editions of his textbook, Joslin included quite long sections on sciatica, which he regarded as a relatively common complication but which was probably amyotrophy.

In a 1930 article, 'Diabetic neuritis with paralysis', Root and Rogers from the Joslin Clinic described at least one case which sounds like amyotrophy. A Mrs EGA was admitted with severe pain in the left leg, particularly bad at night, which had increased steadily for the past 2 months. She found it difficult to walk upstairs and on examination had atrophy of the left quadriceps which was tender (Root and Rogers, 1930).

For the next 20 years diabetic motor neuropathy disappeared from the journals until revived by the Leeds neurologists Hugh Garland (1903–1967) and Derek Taverner (1914–1998). In their 1953 paper, titled 'Diabetic myelopathy', they described five men aged between 56 and 73 with duration of diabetes for between 6 months and 3 years who complained of pain, maximal in the hip and thigh (Garland and Taverner, 1953). There was wasting of the quadriceps and loss of reflexes in the legs. None of the men had objective sensory disturbance and at some stage three had had extensor plantar reflexes. All recovered so that pathological studies were not possible but, because of the upgoing plantars and the contrast with 'ordinary' diabetic neuropathy, Garland suggested that the lesion was in the spinal cord, possibly secondary to atheroma of the spinal arteries. Taverner always believed that the lesion was peripheral but deferred to his senior colleague. In 1955 Garland published a follow-up of the original five cases and added seven more (Garland, 1955). Patient 1 was originally seen in 1950 with a 2-month history of severe pain and weight loss of 12.7 kg and was totally bedridden. He improved steadily and 4 years later was asymptomatic. After 5 years patient 2 had progressed from being severely disabled to being able to walk normally and 'hop on either foot'. The seven new cases were similar except that two were women. In the discussion Garland suggested that the clinical picture needed to be revised from his 1953 description:

> *'It is still true to say that the most usual features are diffuse pain, weakness, wasting and areflexia, usually asymmetrical and limited to the leg. Less constant features are extensor plantar responses and a raised protein content of the CSF. Electromyographic changes are not always similar; they often suggest a cord lesion, but sometimes the changes appear to be of peripheral origin. It would also seem clear that the lesion is not limited to the lumbar cord and may involve cervical or dorsal segments. Because of the variable findings a "diabetic amyotrophy" rather than myelopathy is perhaps the most suitable designation.'*

Where the aetiology was concerned Garland now discounted arteriosclerosis and blamed glucose 'since the metabolism of the nervous system is largely that of glucose'. He also pointed out that, since only three of the 12 patients had been sent to him with even a provisional diagnosis, most physicians were ignorant of the syndrome. The usual diagnosis, when there was one, was sciatica or some other complication of lumbar osteoarthritis. Sceptics originally questioned the existence of the syndrome and in 1960 Garland, a colourful personality and well-known iconoclast, commented that:

'The reaction of others to my first paper was varied; many thought this was merely a product of my Leeds imagination, but most neurologists soon found one or more examples.'

(Garland, 1960)

On the other side of the Atlantic the concept of diabetic motor neuropathy was revived by Joseph Goodman of Ohio, who, at a meeting of the American Diabetes Association in 1953, reported 17 cases of what he called femoral neuropathy. Sixteen had diabetes, all but one were men and all but two over the age of 40 (Goodman, 1954). He described the following three outstanding manifestations:

- pain in the thigh
- muscular weakness
- absence of the patellar reflex.

The pain could be:

'So severe and intractable that opiates are required and because of failing appetite, there may be loss of weight.'

He emphasised that:

'With control of the diabetes by diet and insulin, the neurological complaints of these patients usually disappeared within three months.'

The disorder has collected a bewildering variety of names. In addition to those already mentioned, it has been called 'diabetic polyradiculopathy', 'diabetic mononeuritis multiplex', 'proximal diabetic neuropathy' and 'diabetic lumbosacral plexus neuropathy'. Since 1963 it has been generally agreed that the wasting and weakness are secondary to a neural lesion (Locke et al, 1963), but there has been little agreement about whether this is due to ischaemia, metabolic derangement, inflammation, necrotising vasculitis or inflammatory demyelination. The present consensus is that the main pathological process is ischaemia from microscopic vasculitis (Dyck et al, 1999). Interestingly an almost identical condition occurs in people without diabetes and may respond dramatically to intravenous methylprednisolone (Dyck et al, 2001). Weight loss in the diabetic condition has frequently been attributed to poor glycaemic control but also occurs, albeit less profoundly, in the non-diabetic variety.

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