Diabetic Amyotrophy

Synonyms: Bruns-Garland syndrome, asymmetrical proximal diabetic neuropathy, lumbosacral plexopathy and diabetic lumbosacral radiculoplexus neuropathy

Diabetic amyotrophy is a diabetic proximal neuropathy. See also separate Diabetic Neuropathy article. Most (but not all) affected patients have type 2 diabetes.[1]

Aetiology[2, 3]

Diabetic amyotrophy is believed to result from a multifocal immune-mediated microvasculitis, ie an immune abnormality involving vasculitic changes, microvascular insufficiency and ischaemia followed by axonal degeneration and demyelination.[4]

Nerve biopsy shows multifocal nerve fibre loss suggesting ischaemic injury and perivascular infiltrate. It predominantly affects motor nerves of the lumbosacral plexus, particularly the femoral nerve, although autonomic and sensory nerves are also involved.[4, 5]

The condition is a diffuse axonal neuropathy. It falls within a spectrum of different neuropathic syndromes caused by or associated with diabetes. Their aetiology includes metabolic, compressive and inflammatory/immunological mechanisms. They are discussed further in the separate Diabetic Neuropathy article and include:[2, 6]

- Generalised symmetrical polyneuropathies.
- Diabetic autonomic neuropathy, present to some degree in up to 75% of patients with type 2 diabetes.
- Focal neuropathies: mononeuropathies and entrapment syndromes - eg, median nerve neuropathy.
- Diffuse neuropathies: much less common but significant due to their severity and morbidity:
  - Axonal (mainly proximal - eg, diabetic amyotrophy). Most are lumbosacral but thoracic and cervical symptoms can occur.
  - Demyelinating (proximal and distal): chronic inflammatory demyelinating polyneuropathy (CIDP) is an immune-mediated disorder with primary damage to the myelin sheath. It is characterised by symmetrical, progressive weakness and paraesthesiae and tends to affect younger patients. It is closely related to GBS and is considered the chronic counterpart

Epidemiology

Prevalence amongst populations with diabetes mellitus is generally quoted as 0.8%. It is higher in patients with type 2 diabetes (1.1%) than in those with type 1 diabetes (0.3%). In contrast about 50% of patients with diabetes experience peripheral neuropathy. Onset is usually in patients over the age of 50 years, with median age at onset 56 years, although younger patients can be affected. Whilst the vast majority have type 2 diabetes, the condition is unrelated to the severity or duration of diabetes. The condition can be the first presentation of diabetes. A good index of clinical suspicion is needed in cases of proximal neuropathy.[2, 3, 6, 7, 8]

Non-diabetic lumbosacral radiculoplexus neuropathy is a pathologically and physiologically similar condition in patients without diabetes. Age at onset, course, type and distribution of symptoms and impairments, laboratory findings and outcomes are similar. [4]

Presentation[1, 2]

The main features are weakness, wasting and pain, usually in the quadriceps. Symptoms begin on one side but usually spread to the other, although symptoms often remain asymmetric. Onset may be acute or subacute.

Patients complain of pain (often severe), dysesthesiae and paraesthesiae in the proximal lower limbs - usually the front of the thigh, hip or buttock. Pain may be also experienced in the lower back. Weakness and wasting of thigh and leg muscles follow within days to weeks. Sensory impairment may be present but is minimal and occurs in the cutaneous distribution sharing the affected nerve root or peripheral nerve.[6]

Weight loss occurs in more than half of patients. Stepwise progression occurs over months. In about 50% of patients distal symmetrical polyneuropathy co-exists.

On examination there is proximal muscle weakness. Wasting in the quadriceps, hip adductors and iliopsoas muscles is characteristic. There is an absent or reduced knee jerk reflex, although ankle jerks are commonly preserved. Extensor plantar responses may be present and there may be mild sensory loss.

Some patients have symptoms of associated thoracic or cervical radiculopathy.[6]

Differential diagnosis
The differential diagnosis includes other causes of proximal neuropathy including:

- Cauda equina syndrome.
- Multiple sclerosis.
- Guillain-Barré syndrome (GBS).
- Spinal canal stenosis.
- Vitamin B12 deficiency.
- Alcohol dependency.
- Neoplastic lumbosacral plexopathy.
- Chronic inflammatory demyelinating polyneuropathy (CIDP).

The literature describes two cases of diabetic amyotrophy which were initially diagnosed as quadriceps tendon rupture, undergoing surgical repair. The authors say that quadriceps tendonitis should be viewed with caution in patients with diabetes.\(^5\)

It is important to recognise that the condition can be the presenting feature of diabetes.\(^4\)

Investigations\(^2\)

- All patients not known to have diabetes and with a suspected peripheral neuropathy should have a random blood glucose test.
- Baseline haematonic studies to rule out vitamin B12/folate deficiency.
- Lumbar puncture if CIDP is suspected.
- Electromyography/nerve conduction studies show axonal damage in the classic form of proximal motor neuropathy. If it shows demyelination, a diagnosis of CIDP should be considered.
- MRI of lumbosacral spine to rule out structural and neoplastic disorders.

Management

Conservative treatment consists of optimising diabetic control along with very active physiotherapy and analgesia. Co-existing neuropathic pain is managed according to the usual neuropathic pain ladder. Over a period of two decades intravenous immunoglobulins have been found to produce improvement in both clinical and electrophysiological parameters in patients with some diabetic polyradiculopathies. Some reports have suggested they are effective in diabetic amyotrophy.\(^8, 10, 11\)

A consensus statement in 2009 on the rational use of intravenous immunoglobulin (IVIg) for neuromuscular disorders was not supportive, saying that whilst class I evidence exists to support the prescription of IVIg to treat patients with some conditions, including GBS and CIDP, there were no convincing data to substantiate the treatment of diabetic amyotrophy using IVIg.\(^12\)

Cochrane reviews in 2009 and 2012 also concluded that there is presently no evidence from randomised trials to support any recommendation on the use of any immunotherapy treatment in diabetic amyotrophy.\(^13, 14\) They found only one completed controlled trial using intravenous methylprednisolone in diabetic amyotrophy (Dyck 2006). The results have not been fully published and are not available for analyses.

Prognosis

The condition is usually of a self-limiting disorder, lasting up to three years but often causing significant disability in this time. Pain subsides well before the motor symptoms improve, which may take months although mild-to-moderate weakness may persist indefinitely. Some patients also develop associated upper limb or thoracic radiculopathies.\(^3\)

The course is variable and many patients become wheelchair-dependent during the course of the condition. Good functional improvement is common over time although in may be incomplete: weakness, sensory symptoms and absent tendon jerks may persist indefinitely. Some patients experience multiple episodes but a minority of patients progresses to severe and persisting quadriparesis.\(^4, 15\)

Further reading & references

3. Nagary S, Somashekar C, James CM; Diagnosis and management of diabetic amyotrophy. Endocrinol Metab J. June 2010


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