Clinical and Electrophysiological Abnormalities amongst the Patients of Diabetic Truncal Polyneuropathy

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Abstract :

Introduction : Diabetic truncal polyneuropathy (DTPN) is a well-recognized form of diabetic neuropathy. Electrophysiological abnormalities have not been described in details in literature. **Objective :** To analyze clinical and electrophysiological abnormalities in patients with DTPN. **Methods :** A retrospective analysis of clinical and Electrodiagnostic studies (EDX) was carried out among nine patients with DTPN who attended our institute in last 12 years over the period of Jan 2003 to Aug 2015. **Results :** All nine patients (6 males, 3 females; aged 48-71 years) had type 2 diabetes mellitus (DM). Three patients were on Insulin and remaining patients were on oral hypoglycemic agents. Eight patients had DM for 2-10 years and one patient was diagnosed at presentation. All patients reported truncal paresthesia in a unilateral thoracic dermatomal distribution. Six patients had abdominal muscle weakness and one had proximal limb muscle weakness. EDXs revealed sensory motor polyneuropathy in all patients. Seven patients revealed abnormality on needle Electromyography (EMG) in limb muscles. Six patients revealed acute/chronic neuropathic changes on needle EMG in paraspinal muscles. Five patients revealed active denervation in abdominal muscles. **Conclusion :** DTPN presents with abdomino-thoracic painful paresthesia over usually two-three thoracic dermatomes with abdominal muscle weakness in two-thirds of patients. All patients had distal sensory motor peripheral neuropathy. Detailed clinical history and examination along with EMG/NCS studies can help in early diagnosis & management.

Key Words : Diabetes Mellitus, Electrodiagnostic studies, Truncal Poluneuropathy .

Introduction :

Type 1 Diabetes Mellitus (DM) is related with deficiency of insulin due to autoimmune destruction of Beta cell in pancreas. Type 2 DM is associated with insulin resistance and hyperglycemic state. Patients with impaired glucose tolerance or impaired fasting glucose can present with neuropathic pain.[1] Peripheral neuropathies are associated with both Type 1 and Type 2 Diabetes.[2] Incidence of neuropathy in Type 2 DM patients is about 45% and Type 1 DM patients is about 54-59%.[2] DM can cause different types of neuropathies.

Types of neuropathy[3]

- Distal symmetric sensory-motor polyneuropathy
- Small fiber neuropathy
- Acute severe distal sensory polyneuropathy
- Autonomic neuropathy
- Diabetic neuropathic cachexia
- Hypoglycemic neuropathy
- Treatment induced neuropathy (Insulin neuritis)
- Diabetic radiculoplexopathy
- Mononeuropathies
- Cranial neuropathies
- Truncal radiculopathy

DTPN is a rare form of neuropathy and the exact incidence is unknown. [4, 5] DTPN is also known as thoracoabdominal neuropathy, trucal mononeuropathy or thoracic radiculopathy.

Pathology :

The exact mechanism of nerve damage in asymmetric DTPN is unknown. However, vascular involvement hypothesis is suspected. Sudden onset, unilateral involvement and spontaneous recovery favors vascular origin of asymmetric DTPN. Transient occlusion of Vasa nervorum(the nutrient artery to the nerve) results in relative ischemia to develop symptoms but not enough to cause permanent damage.[4, 5] Vasa nervorum

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is also a suspected site of lesion in Diabetic patients with Vascular disease.[6] Again, whether or not the nerve damage occurs at radicular level or at the nerve trunk level (after fusion of sensory and motor roots) is unclear. [7] Pain and hypersensitivity being the main symptoms raises suspicion for unmyelinated and myelinated nerve fibers involvement. [8] Acute peripheral nerve injury studies have revealed involvement of minimally myelinated A delta or unmyelinated C fibers resulting in spontaneous activation or reduction of threshold to physiological stimuli. [9]

Again, the exact pathological changes in symmetric DTPN is unclear but believed to be related with dying back phenomenon as patients do not exhibit full recovery.[10]

Diabetic polyneuropathy affecting limbs or mononeuropathy affecting the limbs or cranial nerves have been well described in literature, but very little attention was provided to Diabetic truncal neuropathy. Understanding of spinal nerve anatomy is crucial to learn about DTPN.

Once Spinal nerve emerges from intervertebral foramina divides in dorsal and ventral rami. Intercostal nerves arise from ventral rami of T2-T6 spinal roots. Intercostal nerves further divide in lateral and medial cutaneous branches and supply the skin of lateral chest and medial chest, respectively. Thoracoabdominal nerves arise from ventral rami of T7-T12 spinal roots. Thoracoabdominal nerves not only supply the skin of lateral and medial abdomen, but also supply the abdominal muscles. The Paraspinal muscles and back of skin is supplied by dorsal rami of spinal nerve. These nerves receive their blood supply from vasa nervorum of intercostal arteries.

DTPN usually presents as pain and dysesthesia in areas of chest or abdomen. Patients may develop weakness of abdominal muscles and abdominal swelling. However, a variant of Diabetic truncal polyneuropathy described in literature has symmetric involvement of multiple intercostal nerves. [10] Patient present as symmetric sensory loss (rather than hypersensitivity) over thoracic and abdominal segments. Sensory loss is seen in tear drop distribution as long length of traversing abdominal body wall nerves. Unlike unilateral DTPN, this subtype does not exhibit recovery. This subtype is usually associated with distal glove and stocking neuropathy and autonomic neuropathy.

Tests helpful to confirm diagnosis of DTPN:

1. EMG/NCS - plays an important role in diagnosis of DTPN.EMG reveals abnormal spontaneous activity in thoracic or abdominal paraspinal or abdominal wall muscles at the corresponding clinical levels in acute phase. Most of these patients also have peripheral neuropathy or amyotrophy on EMG/NCS study. [11]

2. Skin biopsy- of symptomatic region shows loss of intra-epidermal nerve fibers (IENF) compared to asymptomatic region. Follow up skin biopsy after clinical recovery shows return of Intra-epidermal nerve fibers. These findings points to involvement of small nerve fiber involvement and likely location of injury distal to or at Dorsal root ganglion. [8]

3. Thermoregulatory Sweat test- Patients with DTPN reveal areas of Anhidrosis on thorax and abdominal wall corresponding to clinical symptoms and paraspinal muscles denervation.[12] This abnormality raises suspicion for small nerve fiber involvement and coexistent autonomic neuropathy.

Treatment and Prognosis :

Unilateral DTPN is a self-limiting condition and spontaneous recovery is seen in most cases. Prognosis is good and most of the patients recover in 3 months but recovery can take up to 1-2 years. Different pain medications, sodium channel blockers (Dilantin or Carbamazepine), Tricyclic antidepressants, Gabapentin have been used in literature for symptomatic relief with effectiveness. [4] Currently, there is no data to provide preferential use of one medication over other.

DTPN has been described as case reports or case series in literature as it is rare. Mark Ellenberg et al [4] described a large case series of 40 patients with DTPN in 1978. Here, we have analyzed clinical and electrophysiological abnormalities in total 9 patients with DTPN.

Materials and methods:

We reviewed EDX and clinical charts of all patients given diagnosis of DTPN over 12 years during Jan 2003 to Aug 2015, at our institution at NSLIJ health system, NY and retrospective case study was conducted.

Electrophysiological studies were carried out by the two electromyography instruments, using standard methods described by Kimura and Preston and Shapiro. [13,14] Skin temperature was at or above 32°C when appropriate limb temperature was raised and maintained. Motor nerve conduction studies (NCS) were performed on upper and lower extremities with use of surface electrodes. These included stimulation of median and ulnar nerves at the wrist and forearm while recording from the abductor pollicis brevis muscle and abductor digiti minimi muscles of the hand. In the lower extremity, deep peroneal and posterior tibial nerve stimulated at the ankle and knee, while recording from the extensor digitorum brevis muscle and the abductor hallucis muscle were studied. Sensory NCS were performed with use of antidromic techniques[13,14] of the median, ulnar, superficial peroneal and sural nerves. F waves were measured with each motor NCS for which a compound muscle action potential (CMAP) was obtained.

Electromyography was performed using monopolar needle electrodes in limb muscles, paraspinal muscles and abdominal muscles depending on clinical conditon and EDX.

Results :

Total nine patients with DTPN are reported in present study. Six patients were male and three patients were female. Patients were of 48 to 71 years of age. Three patients were on insulin and six patients were treated with oral hypoglycemic agents. Eight out of total nine patients had DM for 2 to 10 years. One patient discovered to be diabetic at the time of presentation. All nine patients reported truncal paresthesias in a unilateral thoracic dermatomal distribution, mostly in two or three segments. Six patients had abdominal muscle weakness and one patient had proximal limb muscle weakness. All patients revealed sensory motor axonal polyneuropathy on NCS. Seven patients revealed abnormality on needle EMG in limb muscles. Six patients revealed acute or chronic neuropathic changes on needle EMG in paraspinal muscles. Five patients revealed denervation in abdominal muscles.

Discussion :

Diabetic truncal poly neuropathy mostly present as unilateral unpleasant hypersensitivity or pain rather than loss of sensation in one to two roots. Most common roots involved are from T5- T10. In present study, the most common presentation of DTPN was sensory paresthesia & most commonly in T6-7 root distribution. Motor nerves involvement is rare compared to sensory nerves though can present as weakness of abdominal wall muscles. [4, 5] Data from current study revealed, about 66% of patients have

Table 1: Clinical Presentation of Patients

Case No.	Age (years)	Gender	Diabetes Type	Duration of Diabetes	Paresthesias	Weakness
1	59	M	NIDDM	At presentation	T6-7	Abdominal weakness Proximal weakness
2	62	M	NIDDM	2 years	T7-9	Abdominal weakness
3	68	F	NIDDM	4.5 years	T6-7	Abdominal weakness
4	49	M	NIDDM	6 years	T6-7	Abdominal weakness
5	50	F	NIDDM	4 years	T8-9, ?T10	Abdominal weakness
6	71	F	IDDM	10 years	T9-10	No weakness
7	64	M	IDDM	6.5 years	T9-10	No weakness
8	48	M	IDDM	3.5 years	T5-6	No weakness
9	51	M	NIDDM	4 years	T6-7	Abdominal weakness

Table 2: Electrophysiological Abnormalities amongst patients

Case No.	Sensory NCS Abnormality	Motor NCS Abnormality	Limb EMG Active denervation	Para spinal Muscles EMG Active denervation	Abdominal Muscles EMG Active denervation
1	Present	DN+ FN	Present	Present	Present
2	Present	DN	Present	Present	Absent
3	Present	DN	Absent	Present	Present
4	Present	DN	Present	Present	Present
5	Present	DN	Present	Absent	Absent
6	Present	DN	Absent	Present	Absent
7	Present	DN	Present	Absent	Present
8	Present	DN	Present	Absent	Absent
9	Present	DN	Present	Present	Present

abdominal wall muscles weakness. Recurrent episodes of diabetic truncal neuropathy were also described.[5,15] Focal paralysis of abdominal muscles can present as abdominal hernia.[7,15-18] Weight loss was often accompanying symptoms.[5] In one of the largest case series of 40 cases described in literature by Mark Ellenberg4 , DTPN was more commonly found in older individuals and has equal gender distribution. In Present study, DTPN was observed more commonly after age of 50 and predominantly in men. In most of the literatures, the majority of the patients were having long-standing history of Diabetes; unilateral sensory root involvement was more common compared to bilateral or motor root involvement; nocturnal exacerbation of pain was frequently noticed by patients; other types of Diabetic neuropathy such as diabetic peripheral neuropathy, diabetic amyotrophy and autonomic neuropathy were often seen in patients with DTPN. Present study revealed eight patients were known cases of DM & only one patient discovered to have DM after diagnosis of DTPN; all patients have peripheral neuropathy and one patient has Diabetic amyotrophy.

Conclusion :

Unilateral DTPN is a rare but is a very important presentation seen in patients with DM. It is important to differentiate from symmetric DTPN, which is usually seen in advanced cases of diabetic peripheral neuropathy. Patients may present with thoraco-abdominal paresthesia or abdominal muscle weakness. Diabetic peripheral neuropathy, autonomic neuropathy or amyotrophy is also commonly seen in patients with DTPN. Clinicians should have high index of suspicion for diagnosis of DTPN as this condition is self-limiting and spontaneous recovery is noticed in most of the patients compared to peripheral diabetic neuropathy. Lack of awareness may results in unnecessary invasive or noninvasive gastrointestinal/abdominal/cardiac work up, delay in diagnosis and wastage of resources. Clinical diagnosis is obtained by history and examination & EMG/NCS studies can also help in diagnosis.

Abbreviations :

DTPN- Diabetic truncal Polyneuropathy

DM-diabetes mellitus

EDX- Electodiagnostic studies

Abd weakness- Abdominal muscles weakness

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