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DIABETIC AMYOTROPHY – REVISITED: A CASE REPORT

Endocrinology

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ABSTRACT

Introduction – Diabetic Amyotrophy is an uncommon type of peripheral neuropathy. It is a disabling but self limiting condition. Good functional recovery is possible with active physiotherapy.

Case summary – We report a 34 year old patient with history suggestive of diabetes with complaints of asymmetric onset sub-acute proximal followed by distal weakness of lower limbs along with wasting with significant history of weight loss without bladder bowel involvement – lower motor neuron type of weakness with lower limb hyporeflexia.

Discussion – This condition usually involves elderly type 2 diabetics. Earlier studies showed that pathogenesis is metabolic derangement but recent insight is that it is an immune mediated process. Till immunosuppressive agents are clearly shown to be effective, good physiotherapy is the only way to limit residual disability.

KEYWORDS

Diabetic Amyotrophy, physiotherapy, immunosuppressive therapy

INTRODUCTION

– Peripheral Neuropathy is the most common and most disabling long term complication of Diabetes, developing in eventually 50% of the patients.[1]The most common form of neuropathy is a symmetrical, predominantly sensory polyneuropathy.[2] Diabetic Amyotrophy is a distinct type of peripheral neuropathy, with its characteristic set of clinical features. The overall prevalence of diabetic Amyotrophy is low, which is 1.1% in patients with type 2 diabetes mellitus and just 0.3% in type 1 diabetes.[3] Early recognition of this entity is crucial as good functional recovery is possible with just active physiotherapy. There has been a paradigm shift in the understanding of the pathogenic mechanisms of this entity, which was first described by Bruns. We report a diabetic patient who presented with symptom complex of diabetic Amyotrophy at the very onset of his illness.

CASE SUMMARY –

A 34 year old male patient working in a home appliance company presented with deep aching pain in his left thigh for last 6 months followed by weakness in left hip and knee joint. Over the course of 1 month, similar complaints appeared in right proximal lower limb, thereby causing difficulty in standing up from squatting position. He started noticing slipping of slippers while walking 3 months later. He also complained of thinning and frailness of bilateral lower limbs associated with fasciculation. History of osmotic symptoms associated with weight loss of around 10 kg was also present for last 6 months.

Patient was of lean built with average nutrition (BMI of 16.31kg/m^2). General physical examination was normal. Sensorimotor neurological examination revealed asymmetric flaccid weakness in lower limbs, proximal more than distal weakness with loss of pain and temperature sensation extending up to ankle. There was considerable wasting of bilateral quadriceps muscle. (Figure 1) The deep tendon reflexes in the lower limbs were diminished and bilateral plantar response was flexor. Neurological examination of the upper limb and other systemic examination were normal.

Based on the history and clinical examination, we kept the differentials of Lumbosacral Plexopathy (either due to diabetes or neoplastic etiology), Chronic Inflammatory Demyelinating Polyneuropathy (CIDP), Cauda Equina syndrome and Progressive Muscular Atrophy. Routine investigations (Figure 2) revealed that the patient had type 2 Diabetes. Nerve Conduction Study (NCS) (Figure 3) of the lower limbs showed length dependent axonal polyneuropathy with axonal degeneration of lower limb plexus. MRI of lumbosacral spine showed no structural abnormality.

A final diagnosis of newly detected Type 2 Diabetes with Diabetic Lumbosacral Plexopathy with concomitant distal sensorimotor neuropathy was made. Patient was initiated on insulin to achieve aggressive and intensive glycemic control. He was also advised active physiotherapy. Patient gained weight and his weakness improved in subsequent follow up visits.

DISCUSSION –

Diabetic Amyotrophy (Greek = muscular atrophy) was first recognized by Bruns[4] in 1890 and the term was coined by Hugh Garland[5] in 1955 when he first reported 12 patients of type 2 diabetes with this condition. Since then, it has been known by various names including Burns-Garland syndrome[6], diabetic polyradiculopathy[7], femoral and femoral-sciatic neuropathy of diabetes[8]and diabetic lumbosacral plexopathy[9].

There has been more insight into understanding of the pathogenic mechanisms of this condition. Earlier thought was that metabolic derangement and vasculopathy played a major role in pathogenesis[10]. Recent studies indicate that it is primarily an immune-mediated process showing axonal degeneration and focal demyelination in the affected area[11]. Necrotizing vasculitis is observed in cutaneous nerve biopsies of patients with diabetic amyotrophy[11].

This entity commonly affects middle-aged and elderly men with type 2 diabetes, although it may occur in young type 1 diabetic patients also[12]. Age of the patients ranged between 35 to 75 years with a mean age of 65 years in a case series of 33 patients reported by Dyck et al[13]. Patients usually have Type 2 diabetes, but in some cases, diabetic Amyotrophy may be the initial presentation of the disease. In our case also diabetes came to light only after the initial presentation of neuropathy.

The onset of neuropathy is usually with acute or subacute deep aching, lancinating or burning asymmetrical lower extremity pain usually involving the anterolateral thigh region. Weakness follows pain which evolves over weeks to months, commonly affecting the proximal thigh (Quadriceps and Iliopsoas) muscles and occasionally with extensor plantar response. Pain and weakness usually involves the contra lateral extremity and distal leg segments later in the course of the illness. Wasting of the Quadriceps muscle (Vastus Medialis) with fasciculation is a characteristic feature. The illness is commonly preceded by unintentional weight loss of more than 10 lbs which is usually reversible[14].

The diagnosis usually requires a high index of clinical suspicion. The nerve conduction studies (NCS) reveal an axonal process in the form of reduced CMAP (Compound Muscle Action Potential) and absent or reduced sensory responses with proportionate slowing of the conduction velocities. The femoral nerve is most commonly affected. Electromyography of the affected muscle may show changes of acute denervation[14].

Mean time to recovery of weakness is about three months (range 1-12 months) and recovery is generally complete by 18 months. Pain is

usually the first symptom to remit. The majority of patients have residual symptoms such as exercise-related pain, stiffness, recurrent pain, residual weakness on walking and difficulty in climbing stairs. Full recovery of weakness occurs in only 10%–20% of patients[15].

Since the pathogenic mechanism is now understood to be patchy microvasculitis and secondary nerve ischemia, various immunosuppressive treatments in the form of prednisone, prednisone in combination with other immunosuppressant (like azathioprine or cyclophosphamide), and intravenous immunoglobulin (IVIG) have been used in treatment. But studies with immunosuppressive agents have shown variable outcome[16, 17]. Randomized controlled clinical trials are needed to better evaluate their efficacy. Patients may require tricyclic antidepressants, anti epileptics and sometimes narcotic analgesics for disabling pain. Regardless of the pharmacological intervention, physical therapy to improve mobility and strength is of paramount importance.

CONCLUSION –

Diabetic amyotrophy is an uncommon disabling but self limiting condition. This complicates type 2 diabetes mellitus but occasionally may be presenting feature. Blood glucose should be measured in any case of peripheral neuropathy. Early institution of active physiotherapy may help to limit the residual disability. Further controlled studies are needed to analyze the role of early treatment with immunosuppressant medications.

Figure 1 demonstrating marked wasting of quadriceps (particularly Vastus Medialis)

INVESTIGATION	VALUE	INVESTIGATION	VALUE
Hemoglobin	13.2 gm%	Arterial blood gas analysis	pH- 7.404, HCO3 – 22.7, PCO2 – 44.7
Total leucocyte count	5010	S. CPK	46 (<60) - WNL
Platelet count	198,000	Total cholesterol/Tg/HDL/LDL	195/159/40/96
MCV	81.9	HIV	Negative
Blood urea	17 mg%	S. Vit B12	450 pg/ml (WNL)
Serum creatinine	1.0 mg% (eGFR-92ml/min)	S. Amylase/lipase	17/8 (WNL)
Serum bilirubin (total/direct)	0.6/0.1 mg%	Stool for fecal fat	Negative
SGOT/SGPT/ALP	28/38/118	ECG	Normal sinus rhythm
Serum protein/albumin	7.4/4.3 gm %	Chest X Ray	Normal
Serum calcium/phosphorus	9.8/3.9 mg%	Ultrasound Abdomen	Mild hepatomegaly
T3/T4/TSH	0.6/5.6/2.4 (WNL)	Fundus examination	Normal with no features of diabetic retinopathy
HbA1c	16.8%	Anti GAD Antibody	4.4 mIU/l (<20 – negative)
Blood sugar (random)	488 mg%	Fasting S. C-peptide	1.2 nmol/l (normal)
Urine - routine & microscopy	Proteins – trace, ketones – present , glucose – 4+	MRI LS Spine	Normal study

Figure 2 showing the investigations of the case

Sensory NCS

Nerve / Sites	Rec. Site	Lat. ms	P Lat. ms	Amp. µV	Dur. ms	Area µVms	Segments	Dist. cm	Vel. m/s
L MEDIAN - Dig II Ortho									
Dig II	Wrist	4.15	4.85	16.0	1.65	9.1	Dig II - Wrist	12	28.9
L ULNAR - Dig V Ortho									
Dig V	Wrist	NR	NR	NR	NR	NR	Dig V - Wrist	14	NR
L SURAL - Lat Mall Antidr									
Calf	Lat Mall	NR	NR	NR	NR	NR	Calf - Lat Mall	14	NR
R SURAL - Lat Mall Antidr									
Calf	Lat Mall	NR	NR	NR	NR	NR	Calf - Lat Mall	14	NR

Motor NCS

Nerve / Sites	Rec. Site	Lat. ms	Amp.2-4 mV	Dur. ms	Area mVms	Segments	Dist. cm	Vel. m/s
L MEDIAN - APB								
Wrist	APB	4.75	6.4	6.10	15.9	Wrist - APB	8	
Elbow	APB	9.85	6.0	7.75	16.3	Elbow - Wrist	23	45.1
L ULNAR - ADM								
Wrist	ADM	4.35	8.3	8.70	33.2	Wrist - ADM	8	
B.Elbow	ADM	11.95	5.6	10.30	24.9	B.Elbow - Wrist	24	31.6
L COMM PERONEAL - EDB								
Ankle	EDB	NR	NR	NR	NR	Ankle - EDB	8	
Fib Head	EDB	NR	NR	NR	NR	Fib Head - Ankle		
R COMM PERONEAL - EDB								
Ankle	EDB	NR	NR	NR	NR	Ankle - EDB	8	
Fib Head	EDB	NR	NR	NR	NR	Fib Head - Ankle		
L TIBIAL (KNEE) - AH								
Ankle	AH	NR	NR	NR	NR	Ankle - AH	8	
Knee	AH	NR	NR	NR	NR	Knee - Ankle		
R TIBIAL (KNEE) - AH								
Ankle	AH	NR	NR	NR	NR	Ankle - AH	8	
Knee	AH	NR	NR	NR	NR	Knee - Ankle		

Motor NCS

Nerve / Sites	Rec. Site	Lat. ms	Amp.2-4 mV	Dur. ms	Area mVms	Segments
R FEMORAL - Vastus						
B.Ing.Lig	Vastus	8.60	3.1	20.45	34.0	B.Ing.Lig - Vastus
L FEMORAL - Vastus						
B.Ing.Lig	Vastus	11.45	4.5	15.80	36.1	B.Ing.Lig - Vastus

Figure 3 showing NCS report which shows axonal length dependent sensorimotor polyneuropathy with axonal degeneration of lower limb plexus.

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