"Dry" and "wet" beriberi mimicking critical illness polyneuropathy

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Abstract

Three cases with manifestations of right heart failure, shock, metabolic acidosis, and renal failure in varying combination were admitted with paraparesis. Nerve conduction study suggested predominantly motor and mainly axonal type of neuropathy. Rapid reversal of shock, acidosis, and multi-organ dysfunction with timely infusion of thiamine was followed by the complete neurological recovery.

Key Words

Beri-beri, critical illness polyneuropathy, multiorgan failure, thiamine

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Introduction

Beriberi is the clinical disease associated with thiamine deficiency and sub-acute thiamine deficiency will have symptoms of peripheral edema and mixed motor and sensory neuropathy. A more rapid form of wet beriberi is termed as acute fulminant cardiovascular beriberi or Shoshin beriberi, which manifests in the form of shock, severe metabolic acidosis, and renal failure. In individuals without risk-factors for thiamine deficiency, the diagnosis of this condition is often missed.

Critical illness polyneuropathy (CIP) is an acute neuromuscular disorder of severely ill patients in the setting of systemic inflammatory response syndrome, sepsis, and multiple organ failure. In a patient with tachycardia, tachypnoea, and multiorgan dysfunction, weakness may be attributed to CIP, severe electrolyte disturbance, critical illness myopathy, pre-existing neuropathy or possibly Guillain-Barré syndrome (GBS). In this case series, we describe three patients who had the above manifestations and all had an uneventful recovery on treatment with intravenous thiamine.

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Case Reports

Case 1

A 44-year-old non-alcoholic male was admitted with dyspnea, vomiting, upper abdominal pain, oliguria, and weakness of both the lower limbs of 3 days duration. He was admitted thrice in the previous 1 month for symptoms of heart failure at a peripheral hospital, with partial recovery each time upon conservative treatment. On admission, patient had tachypnea, unrecordable blood pressure, heart rate 136/min, elevated Jugular Venous Pressure (JVP), and bilateral pitting edema of legs. Systemic examination revealed right sided S3, systolic murmur of 3/6 intensity at the lower left sternal edge and 3 cm tender hepatomegaly. Bilateral lower limb power was 3/5 with sluggish deep tendon reflexes.

Electrocardiography (ECG) revealed sinus tachycardia with ST depression in leads V3-V6. Arterial Blood Gas analysis showed severe metabolic acidosis [Table 1]. 2D echocardiography on admission showed dilated Right atrium/Right ventricle (RA/RV) with mild tricuspid regurgitation and pulmonary hypertension [Table 1]. A provisional diagnosis of right heart failure with the multiorgan dysfunction possibly due to sepsis or cardiac beriberi was made and he was managed conservatively with broad spectrum antibiotics, sodium bicarbonate infusion, inotropes, and thiamine infusion. Acidosis, shock, and renal failure recovered promptly over from 24 h to 48 h. However, his paraparesis worsened over the next 2-3 days and Nerve conduction study (NCS) suggested predominantly motor, axonal type of peripheral neuropathy [Table 2]. With continued thiamine supplementation, his power in the lower limbs gradually

Table 1: Investigation reports

Parameter	Case 1	Case 2	Case 3	Normal value
Hemoglobin	13.6	11.3	14.2	13-16 g/dl
TLC (on admission)/mm^3	7600	10,600	8,400	4000-11,000
Platelet count/mm^3	253,000	453,000	35,000	150,000-400,000
Creatinine (max)	4.35	1.34	1.9	<1.4 mg/dl
Urea (max)	130	61	151	<40 mg/dl
SGOT/SGPT	91/51	64/36	47/64	<40 U/L
Albumin (on admission)	4.1	3.6	4.3	3.5-5 mg/dl
CPK/CPK-MB	494/51	1208/29	303/11	
Sodium	131	132	124.6	135-145 mmol/L
Potassium	7.6	4.9	5.4	3.5-5 mmol/L
pH: On admission	6.94	7.28	7.26	7.35-7.45
pH: On second day	7.421	7.42	7.42	7.35-7.45
HCO_3: On admission	3.3	14.3	13.2	
HCO_3: On second day	24.8	23.2	22.6	
2D echocardiography	RA/RV dilatation, Mild TR, PASP-50 mm of Hg	Mild TR, PASP-40 mm of Hg, Normal cardiac chamber dimensions	Dilated RA/RV, Moderate TR, PASP 50 mm of Hg	

TLC=Total leucocyte count, CPK=Creatinine phosphokinase, CPKMB=Creatinine phosphokinaseMB, SGPT=Serum glutamic pyruvic transaminase, SGOT=Serum glutamic oxaloacetic transaminase, RA/RV=Right atrium/Right ventricle, TR=Tricuspid regurgitation, PASP=Pulmonary artery systolic pressure

Table 2: NCV data of study subjects

	Latency (mS)		Amplitude (mV)		CV (m/s)	
	Right	Left	Right	Left	Right	Left
Case 1						
CMAP						
Median	3.65	3.22	10.0	11.4	48.6	52.1
Ulnar	3.33	3.14	14.2	13.4	51.17	58.26
Peroneal	4.38	4.58	1.2	1.1	46.6	40.18
Tibial	4.48	4.9	9.8	10.5	41.14	38.24
SNAP						
Median	2.63	2.58	14.5 μv	15.2 μv	42.54	44.22
Ulnar	2.28	2.34	13.8 μv	13.4 μv	52.46	50.02
Sural	2.54	2.71	28.4 μv	26.3 μv	42.46	46.24
Case 2						
CMAP						
Median	3.44	2.81	12.9	14.4	57.06	56.89
Ulnar	2.19	2.50	8.5	4.5	58.31	65.07
Peroneal	3.75	3.96	0.7	157.3 μv	54.89	44.41
Tibial	3.54	3.13	8.4	7.7	42.67	44
SNAP						
Median	2.46	2.71	52 μv	44.2 μv	48.78	44.28
Ulnar	1.88	2.38	47.2 μv	23.9 μv	53.19	42.02
Sural	2.50	2.38	28.7 μv	40.2 μv	48	46.22
Case 3						
CMAP						
Median	3.54	3.23	11	7.9	54.27	59.11
Ulnar	3.65	3.65	8	6.3	57.21	60.10
Peroneal	5.52	4.58	0.9	1.7	36.86	36.47
Tibial	5	4.79	3.4	2.73	35.56	37.28
SNAP						
Median	2.67	2.75	16.4 μv	23.1 μv	56.18	54.55
Ulnar	2.63	2.63	17.2 μv	24.1 μv	45.80	45.80
Sural	2.96	3.00	20.3 μv	25.5 μv	40.54	40.00

NCS=Nerve conduction study, CMAP=Compound muscle action potential, SNAP=Sensory nerve action potential, NCV=Nerve conduction velocity, CV=Conduction velocity

improved and he was discharged in 2 weeks with near normal power in the lower limbs.

Case 2

A 25-year-old lady who had delivered a baby 6 weeks back was admitted with generalized anasarca and paraparesis of 10 days duration. She had dyspnea and oliguria since 1 day. Deep tendon reflexes (DTR) suggested global areflexia and bilateral lower limb power was 3/5. Her thyroid function was normal and Anti-nuclear antibody (ANA) was negative. Her investigation results are tabulated in Table 1 and NCS findings in Table 2. A diagnosis of heart failure with polyneuropathy was made and she was treated with IV thiamine infusion and supportive treatment. Edema disappeared over a week and she was discharged after 12 days with near normal power in lower limbs.

Case 3

A 36-year-old alcoholic male was admitted withbilateral edema of legs andbilaterallower limb weakness of 3 weeks duration. He also had oliguria and vomiting of 4 days duration. Bilateral lower limb power was 4/5 and his echocardiography as well as NCV findings are enlisted in Tables 1 and 2. Upon treatment with intravenous thiamine for 1 week followed by oral supplementation, his renal functions and acidosis recovered, edema disappeared. After 11 days he was discharged with complete recovery.

Discussion

Blankenhorn[1] criteria for beriberi heart disease diagnosis include enlarged heart with normal sinus rhythm, edema and elevated venous pressure, peripheral neuritis, nonspecific ST-T changes in ECG, no other evident cause, history of dietary deficiency, and response to thiamine or autopsy evidence. All the three cases in this series had right heart failure with edema and elevated JVP. Reversible pulmonary hypertension with dilated RA/RV due to an increased pulmonary arterial blood

flow and elevated LV end-diastolic pressure is the predominant presentation in Shoshin beriberi.[2]

Metabolic polyneuropathy shows the pathology of acute axonopathy, which is important for differentiating from the axonal form of GBS. In thiamine deficiency, Adenosine triphosphate (ATP) decreases because of a decrease in pyruvate dehydrogenase activity which is said to be also associated with a decrease in Na^+-K^+-ATPase activity. Our patients may have developed acute axonal polyneuropathy simultaneously due to a rapid thiamine deficiency caused by vomiting, as well as a state of chronic pre-existing thiamine deficiency. In thaimine deficiency neuropathy NCS shows reduced lower limb sensory and motor responses with only mild slowing of conduction velocities. Nerve biopsy shows preferential degeneration of large myelinated axons with absent segmental demyelination. Associated with endoneural edema is present. Unlike other forms of axonal polyneuropathies, recovery is relatively rapid in thiamine deficiency.

CIP is an acute or sub-acute axonal length dependent neuropathy and is viewed as motor predominant or exclusively motor disease.[3] Apart from distal axonal degeneration of motor fibers causing flaccid weakness, decreased or absent deep tendon reflexes; other features of CIP include decreased amplitudes of compound muscle and sensory action potentials with minimal conduction slowing, normal or mildly increased levels of blood Creatinine phosphokinase in a critical care setting. In CIP, nerve biopsy shows degeneration of both motor and sensory nerve fibers without evidence of inflammation, vascular injury or demyelination. Associations and risk factors include immobility, malnutrition, medications including corticosteroids, and aminoglycosides, dialysis, low albumin, and high blood glucose and underlying medical conditions. Recovery is often delayed and incomplete in CIP, and clinical deficits and electrophysiological abnormalities persist in majority even after 5 years.[4]

Other possibilities of polyneuropathy in acutely ill patients include Acute inflammatory demyelinating polyneuropathy (AIDP), uremic polyneuropathy and diphtheritic polyneuropathy. AIDP can be triggered by a critical illness and NCS shows prominent conduction block or slowing. Acute motor axonal neuropathy (AMAN) variant is clinically and electrophysiologically indistinguishable from CIP; however, nerve biopsy in the former shows endoneuiral edema and mononuclear cell infiltration, segmental demyelination and axonal loss in severe cases. Uremic polyneuropathy can occur in acute setting, especially after the initial few hemodialysis. It is predominantly a demyelinating type of sensory neuropathy and NCS shows reduced Sensory nerve action potential (SNAP) amplitudes with prolonged latencies and reduced conduction velocities. Diphtheria can be associated with cardiac failure secondary to cardiomyopathy and can present with acute mixed sensory motor neuropathy of demyelinating type in 15% of cases. Furthermore, critical illness myopathy is clinically difficult to differentiate from CIP, but may have predominant proximal muscle weakness with frequent diaphragmatic involvement. SNAP is normal on NCS, Electromyography (EMG) shows in-excitable muscle and Creatinine phospho-kinase (CK) is elevated to >20 times the normal.

The limitations of the present study included the inability to do the serum thiamine levels and nerve biopsy due to feasibility constraints. Diagnosis of Shoshin beriberi is commonly based on monitoring the therapeutic response to thiamine replacement,[5] in the appropriate clinical setting. The only definitive treatment of beriberi is the rapid intravenous administration of thiamine, which improves the adverse hemodynamic situation within minutes to hours, and is diagnostic of this rare disease.[6] To conclude, these patients had Shoshin beriberi, which is the severe form of thiamine deficiency with right heart failure and Multi-Organ Dysfunction Syndrome; coexisting with polyneuropathy. Both were reversible with thiamine replacement, the former within from 24 h to 48 h and the later over few days to weeks.

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