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UNLOCKING THE MYSTERIES OF SUBACUTE COMBINED DEGENERATION: THE CRITICAL ROLE OF EARLY IMAGING TO UNSHEATH MID-PONS INVOLVEMENT

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ABSTRACT Subacute combined degeneration (SCD) is an uncommon neurological complication due to chronic vitamin B12 deficiency, manifesting as macrocytic anemia and degeneration of the spinal cord's lateral and dorsal columns (1). It typically affects individuals over 40 years old (1), presenting with gastrointestinal, hematological, and neuropsychiatric disorders (2). Symptoms include weakness, distal tingling, numbness, irritability, apathy, drowsiness, and rapid emotional changes (1, 3). This report discusses a 69-year-old woman with SCD extending to the mid pons region. She presented with slurred speech, walking difficulties, memory disturbances, defective cognition, generalized weakness, and mild limb weakness. Past medical history included hypertension, hypothyroidism, urosepsis, anemia, and incomplete B12 replacement therapy. Initial investigations revealed low serum B12 levels (90 picograms per liter), macrocytic anemia, and demyelination in the spinal cord from the ventral pons down to D12. After confirming the diagnosis through MRI, treatment included nasal spray methylcobalamin (Naso B12 - 500 mcg per spray) daily for 18 days, then every other day for 21 days, resulting in improved B12 levels (from 90 to 360 picograms) and neurological symptoms. Additional management included multivitamin infusions, Edaravone, oral folic acid, physiotherapy, and blood transfusions. The patient's neurological deficits improved significantly, and she was discharged with continued B12 supplementation and a protein-rich diet. Early detection and treatment of SCD are crucial for preventing severe neurological complications. This case underscores the importance of thorough diagnostic evaluations, including peripheral smears, hemograms, imaging studies, and electrophysiological tests, for accurate diagnosis and effective treatment.

KEYWORDS : Subacute combined degeneration, vitamin B12 deficiency, macrocytic anemia, spinal cord demyelination, neurological complications.

INTRODUCTION

Subacute combined degeneration (SCD) is a rare yet significant neurological complication resulting from chronic vitamin B12 deficiency. It primarily manifests as a hematological disorder characterized by macrocytic anemia and neurological issues, notably the degeneration of the lateral and dorsal columns of the spinal cord due to demyelination (1). Typically, SCD affects individuals over the age of 40 (1). Vitamin B12 deficiency can give rise to a variety of gastrointestinal, hematological, and neuropsychiatric disorders (2). Initial symptoms of SCD often include a sensation of weakness, which is followed by progressive distal tingling and numbness in both hands and feet (3). These symptoms tend to worsen over time. In addition to these spinal cord symptoms, affected individuals might experience irritability, apathy, drowsiness, suspicion, confusion, and unpredictable emotional changes (1). This case report presents the clinical journey of a 69-year-old woman diagnosed with SCD extending to the mid pons region. It underscores the crucial role of early detection via imaging studies to facilitate effective treatment.

Case Presentation

A 69-year-old woman with a medical history of hypertension, hypothyroidism, urosepsis, anemia, and vitamin B12 deficiency was referred to a neurologist. Her complaints included slurred speech, difficulty walking, memory disturbances, and loss of appetite. Additionally, she experienced defective cognition, generalized weakness, mild limb weakness, and mild slurring of speech. The patient's past medical records indicated that she had received incomplete B12 replacement therapy. For mildly elevated thyroid-stimulating hormone (TSH) levels, she had been prescribed oral thyroxine (50 mcg). Her treatment included a transfusion of one unit of packed red blood cells, intravenous antibiotics, and supportive management for low RBC count. Serum vitamin B12 levels were found to be low, leading to the administration of supplemental B12 orally.

Four weeks later, the patient was admitted to a tertiary care hospital presenting with sub-acute onset weakness in both upper and lower limbs (grade 3/6), loss of joint position sense below the level of C5, and diffuse disorientation. Upon admission, her vital signs were as follows: blood pressure at 180/100 mm Hg, pulse rate at 90 beats per minute, and oxygen saturation at 98%. Extensive investigations included a thyroid ultrasound revealing a hypoechoic nodule with bilateral upper deep cervical lymphadenopathy. Electrophysiological studies indicated sensory-motor axonopathy in the lower limbs. An EEG demonstrated slowed background activity without focal or lateralizing abnormalities. A 2D echocardiogram with color Doppler showed normal valves but a mildly dilated right atrium and ventricle. Blood tests revealed a RBC count of 3 million, macrocytes predominating the peripheral smear, a very high MCV, and decreased serum vitamin B12 levels (90 picograms per liter, with a normal range of 160-950 picograms per liter). Electrolyte assessment showed low serum sodium levels (table 1).

Table 1 - Basic Investigations

Test	Level		
Serum B12	150 picograms		
Haemoglobin	8 gms/dl		
ESR	78 mm		
SGPT	70 IU		
SGOT	37 IU		
Blood sugar	165 mg (random)		
Blood urea	23 mg		
Serum creatinine	1.2 mg		
Serum Vitamin D level	30 ng/ml		
Serum proteins	Albumin 3.5 grams	Globulin 2.7 grams	
Urine	alb	sugar nil	
Urinary protein for 24 hours	24 mg		
ECG	Normal axis	mild LVH	and normal T waves
X-ray	Chest normal		
Peripheral smear	Significant macrocytic	hypochromic profile with normal platelets	
	Low RBC count		
	MCHC 54 pg		

MRI of the brain and spinal cord illustrated acute demyelination from the ventral pons(figure 1) down to the D12 level(figure 2,3), with marked demyelination in the posterior and lateral columns of the spinal cord, thereby confirming the diagnosis of SCD.

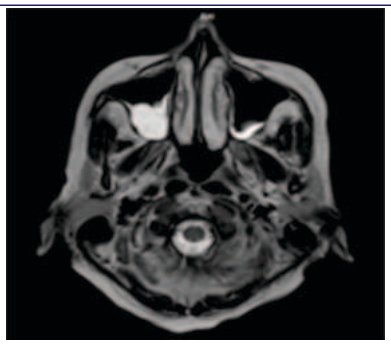

Figure 1 Ventral Pontine Acute Demyelination

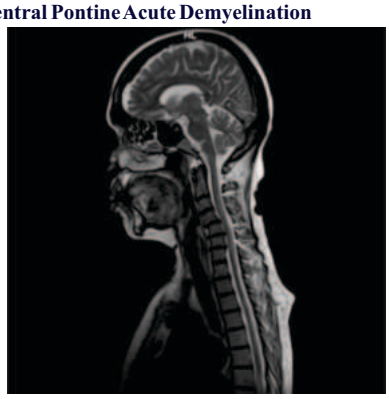

Figure 2 Acute Demyelination At C Spine Level

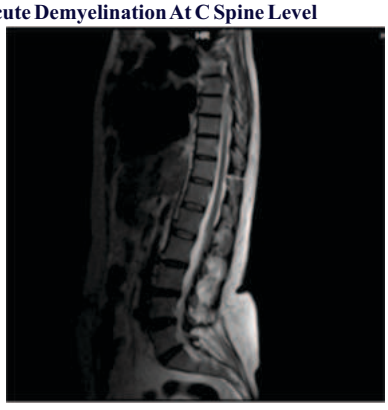

Figure 3 Acute Demyelination At Level Of D Spine

Treatment Given

Upon confirming the diagnosis of SCD through MRI, the patient was treated with nasal spray methylcobalamin (Naso B12 - 500 mcg per spray) daily for 18 days. Following clinical improvement, the dosage was adjusted to every other day for an additional 21 days, with serum B12 levels being monitored throughout this period. The patient's B12 levels increased from 90 to 360 picograms within three weeks, and her neurological symptoms showed significant improvement. Additional supportive management included multivitamin infusions, Edaravone (75 mg IV twice daily), and oral folic acid (10 mg daily). The patient also received physiotherapy and blood transfusions to elevate her hemoglobin level to 12 grams.

Outcome

Following the treatment regimen, the patient's neurological deficits improved to Grade IV/6 within 14 to 30 days, accompanied by an increase in serum B12 levels. The patient was discharged with recommendations to continue parenteral B1, B2, and B6, oral folic acid, multivitamin tablets, and a protein-rich diet. On follow-up, the patient's muscle power and mental state had normalized, indicating a positive response to the treatment.

CONCLUSION

SCD is a significant cause of myelopathy that has the potential for complete recovery if diagnosed and treated promptly (4). The diagnosis relies on clinical presentation and serum vitamin B12 levels, with MRI being crucial for identifying spinal cord demyelination (5). This case emphasizes the need to consider SCD in patients presenting with neurological symptoms and a history of vitamin B12 deficiency. Comprehensive diagnostic evaluations, including peripheral smears, hemograms, imaging studies, and electrophysiological tests, are essential for accurate diagnosis and effective management. Early identification and treatment of macrocytic anemia can prevent severe neurological complications, highlighting the importance of thorough diagnostic evaluations in patients with suspected spinal cord involvement.

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