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A Case of Subacute Combined Degeneration of the Spinal Cord with Bilateral Shoulder Charcot Joint

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Abstract: ***Subacute combined degeneration (SCD) of the spinal cord is a rare neurological condition associated with vitamin B12 deficiency, often presenting with diverse musculoskeletal and systemic manifestations. This report describes a unique case of bilateral Charcot joints in the shoulders of a 56 - year - old female patient with a 15 - year history of left and right shoulder pain, numbness and pain in the left palm for 7 years, weakness of both upper limbs for 2 years, and bilateral lower limb weakness for the past 6 months. The patient had a dietary history suggestive of vitamin B12 deficiency, including strict vegetarianism for the past 20 years, without regular supplementation. Laboratory tests revealed a significantly low serum vitamin B12 level of 120 pg/mL (normal range: 200 - 900 pg/mL), along with findings of macrocytic anemia (mean corpuscular volume [MCV] of 110 fL), consistent with megaloblastic anemia. The peripheral blood smear showed hypersegmented neutrophils, further supporting the diagnosis of vitamin B12 deficiency. The patient initially presented to the orthopedic outpatient department (OPD) due to her shoulder pain, and X - rays revealed complete resorption of the humeral heads and glenoid fossae, along with joint effusions and loose bodies. An MRI of the shoulders and cervical spine was performed simultaneously to evaluate her musculoskeletal and neurological symptoms. The MRI confirmed extensive joint destruction in both shoulders and revealed characteristic hyperintensities in the bilateral posterior columns of the cervical spinal cord, confirming the diagnosis of SCD. This case emphasizes the rare coexistence of SCD and bilateral Charcot joints in the shoulders, highlighting the importance of advanced imaging techniques in diagnosing and managing such complex presentations. The findings underscore the necessity of early recognition and treatment of vitamin B12 deficiency to prevent severe complications like neuropathy, joint degeneration, and anemia, contributing to the understanding of atypical manifestations of this deficiency.***

Keywords: Subacute Combined Degeneration (SCD), Vitamin B12 Deficiency, Bilateral Charcot Shoulder, Neuropathic Joint Disease, Shoulder Arthropathy, Megaloblastic Anemia, Cervical Spine Lesions, MRI Findings in SCD, Chronic Shoulder Pain, Neurological and Musculoskeletal Complications

1. Introduction

Subacute combined degeneration (SCD) of the spinal cord is a rare but serious neurological disorder resulting from vitamin B12 deficiency. It primarily affects the dorsal and lateral columns of the spinal cord, leading to a combination of sensory and motor dysfunctions. If left untreated, SCD can progress to irreversible neurological damage. Vitamin B12 deficiency is often associated with dietary insufficiency, especially in individuals following strict vegetarian diets without appropriate supplementation. Clinically, SCD presents with a wide spectrum of neurological symptoms, including sensory deficits, weakness, and ataxia. In advanced stages, it can affect both the upper and lower limbs, causing significant morbidity. (1)

One of the lesser - known complications of vitamin B12 deficiency is neuropathic arthropathy, or Charcot joints, which typically affects weight - bearing joints such as the knees and ankles. Charcot joints in the upper extremities, particularly in the shoulders, are exceedingly rare. Neuropathic arthropathy is characterized by the progressive destruction of joints due to loss of proprioception and protective pain sensation, leading to severe joint instability, deformity, and dysfunction over time.

This paper presents a rare case of bilateral Charcot joints in the shoulders in a patient with subacute combined degeneration. The patient's condition was complicated by longstanding vitamin B12 deficiency, megaloblastic anemia, and progressive neurological symptoms. Despite the typical presentation of Charcot joints in the lower extremities, this case underscores the importance of considering upper extremity involvement, especially in patients with concurrent neurological and dietary risk factors. Early diagnosis of vitamin B12 deficiency is crucial, as timely treatment can halt or reverse many of its complications. This report adds to the limited body of literature on the association between SCD and Charcot shoulder joints, highlighting the role of advanced imaging techniques in diagnosis and management.

2. Case Presentation:

A 56 - year - old female, with a history of strict vegetarianism for the past 20 years, presented to the orthopedic outpatient department with complaints of progressive bilateral shoulder pain for 15 years, numbness and pain in the left palm for 7 years, weakness in both upper limbs for 2 years, and bilateral lower limb weakness for the past 6 months. The shoulder pain was initially intermittent but gradually worsened over time, leading to significant functional impairment. She also reported difficulty with activities involving her arms and hands, as well as gait instability due to lower limb weakness.

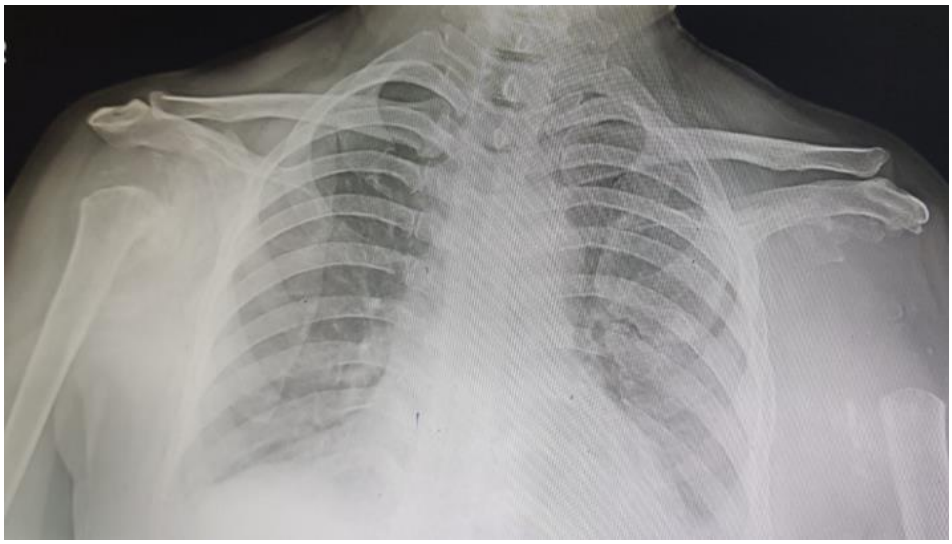
The patient had no significant past medical history or family history of neurological disorders. She denied any trauma or injury to the shoulders. On examination, both shoulder joints exhibited marked instability and deformity, with restricted range of motion. Neurological examination revealed fine touch, vibrations, position sense decreased in the bilateral upper and lower limbs with negative Romberg's sign which is suggestive of posterior column involvement. Crude touch, pain and temperature sensation decreased till the C8 - T1 dermatome levels which is suggestive of spinothalamic tract involvement.

Given the chronicity and severity of her symptoms, the patient was referred for imaging. Bilateral shoulder X - rays revealed complete resorption of the humeral heads and glenoid fossae resulting in an amputated appearance of humeral heads bilaterally, with evidence of joint space distention and the presence of loose bodies. On the left side, there was also a fracture of the scapular body with caudal displacement. These findings suggested advanced neuropathic arthropathy, or Charcot joints, in both shoulders.

To further investigate the neurological symptoms and spinal involvement, MRI of the shoulders and cervical spine was performed. The cervical spine MRI showed hyperintensity in the dorsal and posterior columns on T2 - weighted images, consistent with subacute combined degeneration of the spinal cord due to vitamin B12 deficiency. MRI of both shoulders demonstrated severe joint destruction with complete loss of the humeral heads, joint effusions, and the presence of multiple large bone fragments and fine bony debris.

Laboratory investigations revealed a significantly low serum vitamin B12 level of 120 pg/mL (normal range: 200 - 900 pg/mL), along with macrocytic anemia (mean corpuscular volume [MCV] of 110 fL). Peripheral blood smear examination showed hypersegmented neutrophils, further supporting the diagnosis of megaloblastic anemia secondary to vitamin B12 deficiency.

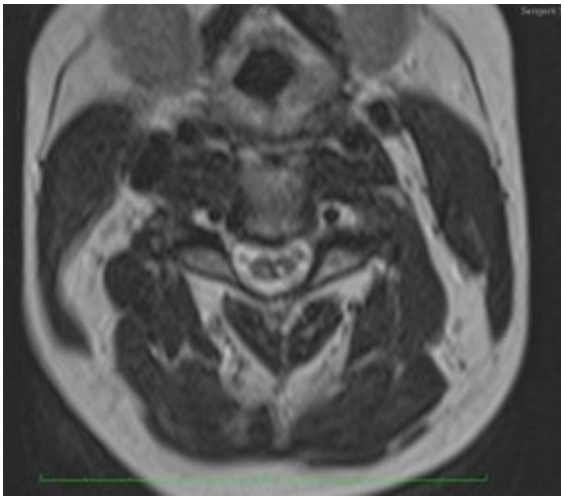
Based on the clinical, imaging, and laboratory findings, the diagnosis of subacute combined degeneration of the spinal cord with bilateral Charcot shoulder joints secondary to vitamin B12 deficiency was confirmed. The patient was initiated on high - dose intramuscular vitamin B12 therapy and referred for orthopedic and neurological rehabilitation to manage her musculoskeletal and neurological impairments.



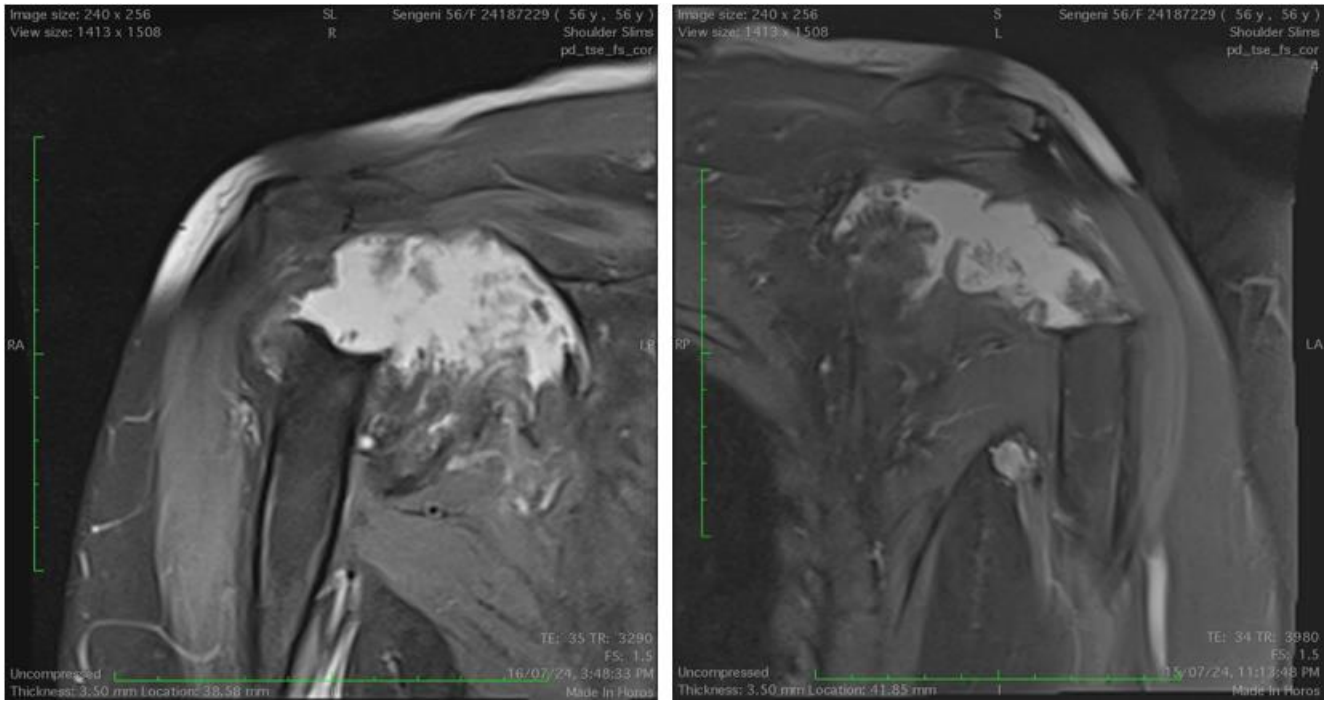
X-ray Findings – Bilateral Shoulders:

- **Right Shoulder:**
  There is complete resorption of the humeral head and glenoid fossa, along with the coracoid process. The joint space is distended, and loose bodies are visible within the joint.
- **Left Shoulder:**
  Complete resorption of the humeral head and proximal portion of the humeral shaft is noted, with blunted margins. A fracture is evident in the body of the scapula. Additionally, there is resorption of the glenoid fossa and coracoid process. The scapula appears to be displaced caudally.

A sagittal T2-weighted image demonstrates abnormally increased signal intensity along the dorsal aspect of the cervical spinal cord (indicated by arrows).



An axial T2-weighted image reveals an area of hyperintensity within the posterior columns of the cervical spinal cord.



T2 fat-suppressed (FS) coronal sections of the right and left shoulders demonstrate complete destruction of the humeral head and neck, resulting in an amputated appearance, along with destruction of the glenoid. Bilateral joint space effusions are present with multiple large bone fragments and fine bony debris.

3. Discussion

This case report highlights the rare coexistence of subacute combined degeneration (SCD) of the spinal cord and bilateral Charcot joints in the shoulders of a 56 - year - old female patient. SCD, resulting from vitamin B12 deficiency, primarily affects the dorsal and lateral columns of the spinal cord. This patient presented with a complex array of symptoms, including bilateral shoulder pain for over 15 years, numbness and pain in the left palm for 7 years, and weakness of both upper and lower limbs that progressed over the past 6 months.

The imaging findings revealed extensive destruction in both shoulder joints, with X - rays demonstrating complete resorption of the humeral heads and glenoid fossae, as well as joint effusions and loose bodies. MRI findings corroborated these changes, showcasing complete destruction of the humeral heads and necks and highlighting hyperintensity in the dorsal columns of the cervical spine. This underscores the critical role of advanced imaging techniques in diagnosing both SCD and Charcot joints, particularly in atypical presentations that may not follow the expected patterns commonly observed in weight - bearing joints.

Vitamin B12 deficiency can arise from various causes, including dietary insufficiency, malabsorption syndromes such as pernicious anemia, or complications following gastrointestinal surgery. (2) In this patient, despite a longstanding history of symptoms, the diagnosis was confirmed through comprehensive imaging and laboratory evaluation. While many patients with SCD present primarily with neurological symptoms like paresthesia, gait instability, and weakness, this case emphasizes the need to consider musculoskeletal complications, particularly in patients with a prolonged history of untreated deficiency.

Neuropathic arthropathy, or Charcot joints, is a significant complication of severe neuropathy, often resulting from a loss of proprioception and protective pain sensation, which leads to repeated microtrauma and joint destruction over time. The simultaneous involvement of both shoulder joints in this case is exceptionally rare and reflects the severe and prolonged nature of the underlying neuropathy due to vitamin B12 deficiency.

The MRI findings showing hyperintensities in the posterior columns of the cervical spine were classic for SCD, complementing the laboratory results that confirmed the presence of megaloblastic anemia. These hematological findings further underscore the importance of early recognition and timely intervention, as prompt vitamin B12 supplementation can halt or potentially reverse neurological and hematological complications. Unfortunately, by the time of diagnosis in this patient, joint destruction was likely irreversible, underscoring the importance of early intervention to prevent such debilitating outcomes. [(3)]

In India, where dietary insufficiency of vitamin B12 is prevalent among strict vegetarians and where malabsorption syndromes may go unrecognized, it is crucial for healthcare professionals to remain vigilant in identifying vitamin B12 deficiency, particularly in patients presenting with nonspecific complaints of chronic pain and weakness. This case contributes to the limited literature on Charcot joints in the upper extremities and highlights the importance of advanced imaging in diagnosing complex conditions like SCD. Educational initiatives aimed at increasing awareness of dietary sources of vitamin B12 and the need for supplementation in at - risk populations could be beneficial in mitigating deficiency - related complications [(4)]

Regular monitoring and management of patients with identified vitamin B12 deficiency, especially those presenting with significant neurological and musculoskeletal complications, are essential for improving clinical outcomes and preventing irreversible damage.

4. Conclusion

This case report documents a rare case of bilateral Charcot joints in the shoulders associated with subacute combined degeneration of the spinal cord, secondary to long - standing vitamin B12 deficiency. The case highlights the importance of early diagnosis and treatment of vitamin B12 deficiency to prevent severe neurological and musculoskeletal complications. In particular, clinicians should consider the possibility of Charcot joints in the upper extremities in patients with advanced neuropathy. Early recognition, supported by advanced imaging techniques and prompt vitamin B12 supplementation, can prevent or mitigate many of the irreversible complications seen in such cases. This report contributes to the growing understanding of the atypical presentations of vitamin B12 deficiency and the role of imaging in guiding diagnosis and management.

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