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Case Report

Case Report of Numb Chin Syndrome (NCS) in Sickle Cell Disease (SCD)

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Abstract

Numb chin syndrome is a rare presentation due to injury to mental nerve characterized by numbness, hypoesthesia, and paresthesia. This syndrome is mostly presented with dental interventions, traumatic injuries, or malignant pathologies. We report a unique case of a 20-year-old male diagnosed with sickle cell disease presented with loss of sensation over right chin area secondary to the vaso-occlusive crisis. This case depicts a complex relationship between neurological complications and hemoglobinopathies of SCD.

Keywords: Sickle cell disease, Vaso-occlusive crisis, numb chin syndrome, hypoesthesia, paresthesia.

1. INTRODUCTION

Numb chin syndrome is a sensory neuropathy that occurs from mental nerve or inferior alveolar nerve injury. It presents as paresthesia, hypoesthesia, or pain in lower lip or chin [1]. This syndrome was first observed by Charles Bell in 1830, in a patient with breast cancer. Later on, the term was coined in 1960 when five more cases were reported [2]. Numb chin syndrome is a manifestation of both benign and malignant conditions. In several cases, it has been misdiagnosed and led to late diagnosis of malignancy. The symptoms commonly caused by dental or surgical interventions, traumatic injuries, drugs, and extra cranial malignancies [3]. Although, few cases were reported NCS with sickle cell disease [4].

The case under discussion presented a unique picture of a young male with sickle cell disease, presented right sided neck chin syndrome during a vaso-occlusive crisis. After 2 months of follow-up, there is partial recovery of the syndrome with no crisis.

2. CASE PRESENTATION

A 20 year old male patient known case of Sickle Cell Disease presented to the emergency room with complaints of generalized severe, boney body aches and fever for 2 days. There was no history of cough or sputum production, no symptoms suggestive of an infectious focus, no history of travelling to high altitude, no new medications or substance abuse, no history suggestive of dehydration, positive history of chest pain without shortness of breath. He was taking folic acid and hydroxyurea for sickle cell disease. He reported good compliance with his medications.

On general physical examination, the patient was febrile 39, tachycardia recording 110 beats per minute, maintaining saturation on room air, and blood pressure recorded in the normal range. On Systemic examination, there was bilateral basal lung crepitation, the rest of the systemic examinations were unremarkable. His initial laboratory reports are the following (Table 1):

Table 1: Investigations upon admission to the hospital

Marker	Lab Values	Normal Range
WBC	14 - 20 K/ UL	3.7-10.1 K/UL
LYM	1.50 K / UL	1.5-4 K/UL
RBC	3.96 X 10^6 ML	4.06-5.58 X 10^6 ML
HGB	10.20 G/DL	12.5-17.7 G/DL
HCT	30.30 %	37.7-53.7 %
MCV	76.50 FL	81.1-96 FL
MCH	25.80 PG	27-31.2 PG

Marker	Lab Values	Normal Range
MCHC	33.70 %	31.8-35.4 %
RDW	23 K/ UL	11.6-14.8 K/ UL
PLT	240 K/UL	150-400 K/ UL
NEU %	80.10 %	39.3-73.7 %
LYM %	10.70%	18-48.3 %
AST	78 U/ L	5-34 U/ L
ALT	30 U/L	0-55 U/ L
CRP	85.30 mU/ L	0-10 mU/ L
Creatinine	0.90 mg/dl	0.72-1.5 mg/dl
Magnesium	2.10 mg /dl	1.8-2.4 mg/dl
Phosphorus	3.50 mg /dl	2.3-4.7 mg/dl
Potassium	3.60 MMol/L	3.5-4.7 MMol/L
Random Blood Sugar	127 mg/dl	79-140 mg/dl
Sodium	123MMol/L	136-145 MMol/L
Urea	12 mg/dl	12-20 mg/dl
Uric Acid	7.40 mg /dl	3.5-7.2 mg/dl

The complete blood count, liver function test, and renal function test were done and presented in Table 1. On chest X-ray (Image 1) and CT chest (Image 2 and 3), there was bilateral lower lobes lung opacification.

The patient was admitted to the ICU with a diagnosis of severe Vaso-occlusive sickle cell crisis with acute chest syndrome/community-acquired pneumonia. The management consisted of liberal hydration, empirical broad-spectrum antibiotics, and opioid for pain control, and planned for a blood transfusion. He responded well to this treatment.

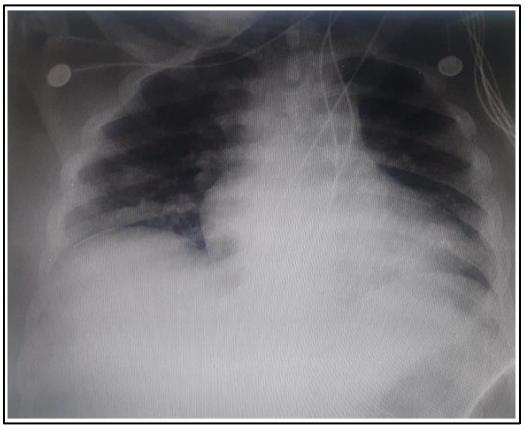

Image 1: PA view Chest X-ray

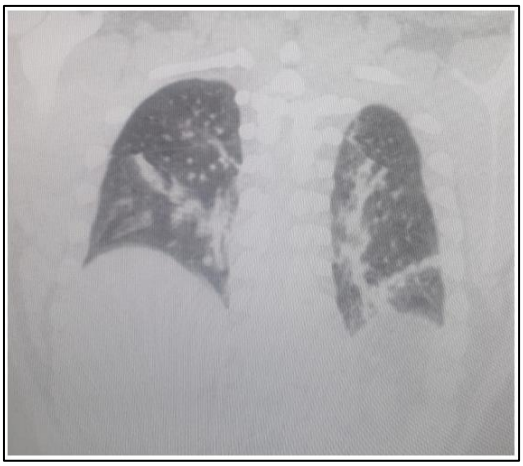

Image 2: Coronal view Chest CT showing bilateral lung infiltrates

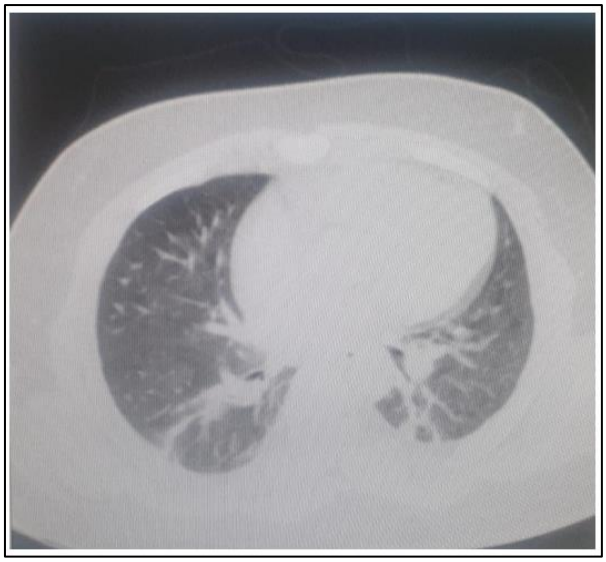

Image 3: Transverse view CT Chest showing bilateral lung infiltrates

On the 4th day of admission, the patient reported right chin area numbness and loss of sensation. It started around the time of the attack. He had no history of trauma, dental or facial procedures, no previous dental problems, and no other CNS complaints. Complete CNS Examination at that time only revealed loss of sensation over the right chin area. Dental consultation and examination were unremarkable. MRI of the brain and temporomandibular joint and bone were negative for stroke, bone, or soft tissue acute pathology. And the patient was diagnosed with numb chin syndrome secondary to the vaso-occlusive crisis.

The patient was discharged from the hospital on the 7th day with a resolution of the crisis, but the numbness and loss of sensation did not improve.

The patient was seen on follow-up after 2 months and reported partial resolution of the numbness and partial recovery of the sensation. He did not report any other CNS symptoms and did not experience other Sickle cell crises.

3. DISCUSSION

Mental nerve neuropathy or numb chin syndrome is an important clinical sign presenting as numbness or altered sensation over mental nerve anatomical distribution. It has been observed that patients with this neuropathy have unilateral involvement commonly between ages of 61 to 70 years reporting as 100 % numbness, 17 % pain and 18% paresthesia [5]. It is not presented with taste or motor disturbance. The common causes are trauma, malignancy, dental intervention [6]. In this case, there was neither significant local cause, nor history of trauma or malignancy.

There is a loss of sensory distribution in the right chin area and in this case study it is presented how sickle cell disease presents clinical manifestations other than hematological presentations. Cerebral infarcts and stroke are common in sickle cell disease complications [7]. In this case, the CNS examination was unremarkable. In sickle cell disease, vaso- occlusion leading to tissue ischemia occurs due to presence of hemoglobin S. The crescent shaped red blood cells lead to hemolysis and ischemia in blood vessels [8].

In this case of numb chin syndrome, vaso occlusion anatomical location is in mandibular bone. The passage of mental nerve and inferior alveolar artery is through mandibular foramen and mental foramen through narrow mandibular bony canal leading to infarction and ischemia [9]. Sickle cell disease leads to inflammatory state resulting in vascular compromise [10]. NCS occurs due to disturbance in mental nerve distribution. Theoretically, in this case NCS pathophysiology exists due to hyper viscosity caused by sickle cell disease [11].

The presentation of NCS secondary to vaso-occlusive crisis suggests that trigeminal nerve (mental nerve) connections could have been affected due to ischemia caused by the crisis. The precise mechanism that caused possible transient ischemia needs to be understood. During follow-ups, partial resolution depicts some degree of recovery with time. Due to limited literature, existence of vaso-occlusive crisis along NCS needs more research.

4. CONCLUSION

This case study showed complex manifestations of neurological numb chin syndrome and vaso- occlusive crises in sickle cell disease. It is important for healthcare providers to understand the potential complications of sickle cell disease. It is important to consider these neurological complications as differential diagnoses

before and after vaso- occlusive crisis in patients with sickle cell disease.

Further clinical studies and research are required to get a better understanding of the underlying pathways of the numb chin syndrome in sickle cell disease and developing right management strategies for such complications.

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