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Sickle Cell Anemia Child presented with Bell's palsy: A Rare Case Report

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Abstract

Sickle cell anemia (SCA) is an inherited red blood cell disorder that results from the replacement of a valine residue for glutamic acid at position 6 in the beta-subunit of haemoglobin. This can lead to tissue ischemia, microcirculation obstructions, infarction, and acute stroke while Bell's palsy is a neuropathy caused by traumatic, inflammatory, infective, or compressive conditions on the facial nerve. In patients with SCA, however, stroke may occur as an acute clinical syndrome presenting with hemiplegia with the unilateral facial nerve. Until now, no case has been reported in the literature with unilateral facial nerve palsy in SCA patients. Here, a rare case of idiopathic unilateral Bell's palsy is presented in an SCA patient.

Key Words**:** Child, Bell's palsy, Facial nerve, India, Sickle cell anemia.

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1- INTRODUCTION

Sickle cell anemia (SCA), an autosomal recessive disease, results from the replacement of valine for glutamic acid at position of the beta-globin gene. Neurological complications, mainly ischemic stroke, are a major cause of morbidity and mortality in SCA. The crisis that occurs includes vasoocclusive crisis, sequestration crisis, acute chest syndrome, and aplastic crisis (1). The most frequent neurological complication associated with SCA is hemiplegia, which is frequently accompanied by unilateral facial nerve palsy. Dr. Charles Bell first depicted Bell's palsy in 1821. Trauma, inflammation, infection, or a compressive condition on the facial nerve are the common causes of Bell's palsy (2). There are a number of cases with no identifiable etiologies of Bell's palsy and are labelled as idiopathic. Any aetiology that causes the acute inflammation and oedema of the facial nerve can lead to compression and eventually ischemia of the nerve. Thus, it manifests as the paralysis of the facial expression muscles. Furthermore, it is not a life-threatening disorder, but it has significant functional and psychological effects on the child (2-3). The diagnosis of idiopathic facial nerve palsy is always considered after the exclusion of all other causes. Bell's palsy presentation is relatively rare in the paediatric age group, but they have a more favourable outcome as than adults (4). After performing a thorough literature search, no case reports were found in the literature mentioning the causes of unilateral lower motor neuron facial palsy in SCA children. Here, a case of unilateral Bell's palsy is presented in a 14-year-old female with SCA.

2- CASE PRESENTATION

A 14-year-old adolescent female, a known case of SCA, presented to the Pediatric Outpatient department, Acharya Vinabo bhave hospital, Sawangi Meghe, Wardha, India, with a complaint of deviation of the mouth to the right side while chewing or swallowing since last 7 days. It was sudden in onset and non-progressive in nature. There was facial asymmetry, especially when she laughed and inability to close her left eye completely. There was no history of fever, cough, vomiting, ear discharge, head trauma, convulsion, headache, unconsciousness, or weakness in the limbs, as well as previous blood transfusion. She was on oral medications, such as hydroxyurea capsules, folic acid tablets, and zinc syrup for the last 5 years. She was a product of non-consanguineous marriage and both of her parents had the sickle cell trait (HbSA).

Her vitals were a temperature of 38.5 ℃, a pulse of 92 beats/minute, a blood pressure of 110/70 mm Hg, a respiratory rate of 20 breaths/ minute, and oxygen saturation of 98% in the room air. General examination revealed mild pallor, no icterus, and cyanosis, or clubbing. There was no cervical lymphadenopathy. Her tonsils were not inflamed. Facial nerve examination revealed that she had a loss of nasolabial fold, inability to blow her cheeks, incomplete closure of eyelids, and complete weakness on the left side of the face (**Figure. 1**).

There was no loss of sensation or taste. The higher function and the examination of the other cranial nerves were normal. There were no motor and sensory deficits. She had a normal gait and speech and her coordination was normal. Signs of meningeal irritation were absent. Normal heart sound was auscultated on cardiovascular examination and l air entry was bilaterally equal on respiratory examination. Her abdomen was soft without hepatosplenomegaly. Laboratory investigations revealed haemoglobin (10 g/dl), white blood cell count (WBC, 6,200 /mm^3), and platelets (1.6 lakh/mm^3). She had normal serum electrolytes and renal function tests. Parents were not willing for the neuroimaging study. It was diagnosed as a left facial asymmetry with complete lower

motor neuron type left facial nerve paralysis, i.e. Bell's palsy, and oral prednisolone was given for 7 days. The patient had a remarkable improvement without any residual weakness.

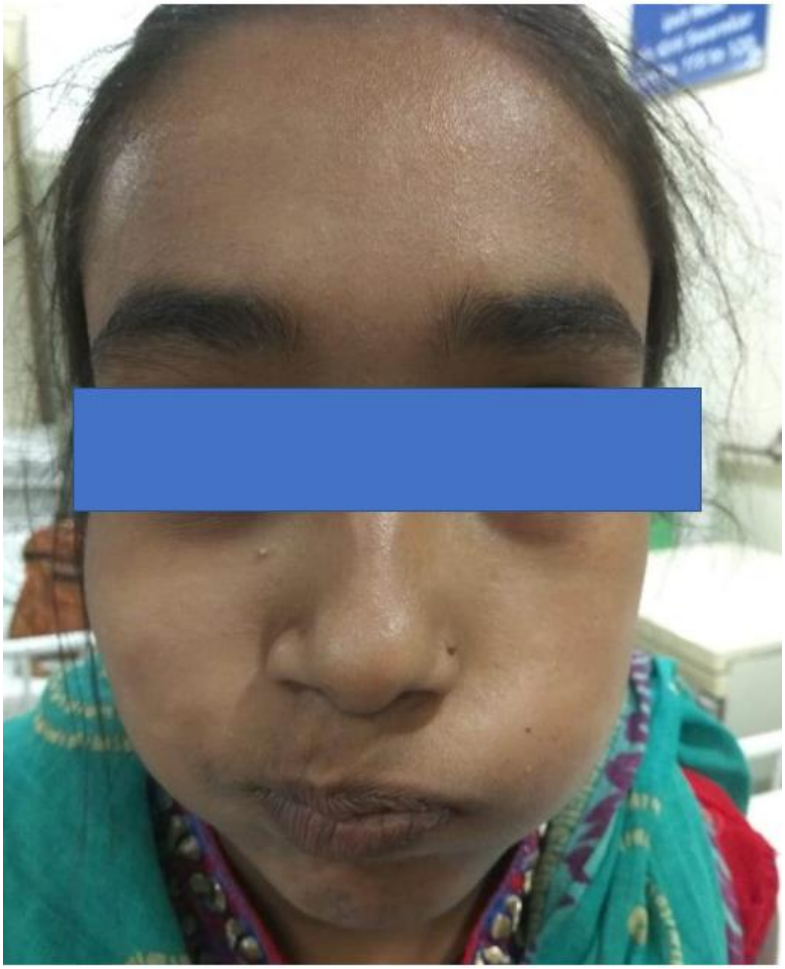

Fig.1: Facial weakness on the left side of the face.

3- DISCUSSION

SCA is a genetic disorder of the red blood cell but is commonly associated with end-organ complications if not diagnosed early. Its complications arise because of vasooclussive crisis, sequestration crisis, hyper-haemolytic crisis, and aplastic crisis. The complications of the vasoocclusive crisis include acute chest syndrome, stroke, and avascular necrosis of the femur, osteomyelitis, priapism, renal insufficiency, and retinopathy. Cerebral infarction is the most common neurologic complication that occurs with SCA. Transient ischaemic attack, cerebral haemorrhage, or seizures are the other neurological complications that occur in SCA children (5-7). Sudden acute onset of facial paralysis, i.e. Bell's palsy, is less common during childhood than adulthood. In 1830, Sir Charles Bell described the first case of unilateral facial weakness. The approximate incidence of Bell's palsy ranges from 11.5 to 53.3 /100,000 person-years in different populations (8). The aetiology of Bell's palsy is not completely understood; however, congenital causes, such as birth trauma, genetic, and acquired causes (e.g. infection, inflammation, and trauma) of facial nerve paralysis need to be ruled out before the diagnosis of Bell's

palsy, for which there are no confirmatory laboratory tests available. Radiological and serological investigations are indicated in the case of persistent facial weakness to exclude the neurological conditions, including neuroma or lyme disease (9). Lumbar puncture is mainly avoided but should be considered for severe cases. Magnetic resonance imaging or computed tomography can be used to rule out the central nervous system (CNS) tumour. Most of the treating physicians are dependent on the symptoms and clinical signs for the diagnosis and management of Bell's palsy (9-11). The diagnosis can be made after excluding the other causes of acute peripheral palsy. There is still ongoing debate about the role of antiviral and steroids in the treatment of the condition. Though residual dysfunction may rarely be seen, the prognosis is good. Ogundunmade et al. (10) reported one adult case of SCA with bilateral facial nerve palsy. To date, unilateral facial nerve palsy in SCA children has not been reported in the literature. This is the first case report of unilateral facial palsy associated with SCA.

4- CONCLUSION

In children, Bell's palsy is a self-limiting condition with a good prognosis. The diagnosis was made based on the clinical findings. Therefore, this is the first case report of Unilateral Bell's palsy seen in a 14-year-old female adolescent with SCA.

5- CONFLICT OF INTEREST: None.

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