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 which 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 which caused by traumatic, inflammatory, infective, or compressive conditions on the facial nerve. However, in patients with SCA, stroke may occur as an acute clinical syndrome presenting with hemiplegia with unilateral facial nerve. Till now, there is no case has been reported in the literature with unilateral facial nerve palsy in SCA patient. Here, we present a rare case of idiopathic unilateral Bell’s palsy in a 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 which occurs include 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. Bell’s palsy was first depicted by Dr. Charles Bell in 1821. Trauma, inflammation, infection or compressive condition on the facial nerve is the common cause of Bell’s palsy (2). A number of cases with no identifiable etiologies of Bell’s palsy exist and are labelled as idiopathic. Any aetiology which causes the acute inflammation and oedema of the facial nerve can lead to compression and eventually ischemia of the nerve. So, 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 exclusion of all other causes. Bell’s palsy presentation in the paediatric age group is relatively rare, but they have a more favourable outcome as compared to adults (4). After performing a thorough literature search, we found no reports of. No case reports has been found in the literature mentioning the causes of unilateral lower motor neuron facial palsy in SCA child. Here we present a case of unilateral Bell’s palsy in a 14 year old female with SCA.

2- CASE PRESENTATION

A 14-year-old adolescent female, known case of SCA presented to the Pediatric Outpatient department, Acharya Vinabo bhave hospital, Sawangi Meghe, Wardha, India, with complaint of deviation of mouth to right side while chewing or swallowing since last seven 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. There was no history of previous blood transfusion. She is on oral medications like capsule hydroxyurea, tablet folic acid and syrup zinc since last 5 years. She was a product of non-consanguineous marriage and her both the parents are sickle cell trait (HbSA). Her vitals were temperature of 38.5°C, a pulse of 92beats/minute, a blood pressure of 110/70 mmHg, a respiratory rate of 20 breaths/ minute, and oxygen saturation 98% in room air. On 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 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 was no motor and sensory deficit. She had a normal gait and speech and co-ordination was normal. Signs of meningeal irritation were absent. Normal heart sound were auscultated on cardiovascular examination and 1 air entry was bilaterally equal on respiratory examination. Her abdomen was soft without hepatosplenomegaly.

Laboratory investigations revealed hemoglobin 10g/dl, white blood cell count 6,200 /mm³, platelets 1.6 lakh/mm³. She had normal serum electrolytes and renal function test. Parents were not willing for the neuroimaging study. A diagnosis of Left facial asymmetry with complete lower motor
neuron type left facial nerve paralysis i.e. Bell’s palsy was made and oral prednisolone was given for seven 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 a genetic disorder of the red blood cell but commonly associated with end organ complications if not diagnosed early. Its complications arise as a consequence of vasooclusive crisis, sequestration crisis, hyper-haemolytic crisis and aplastic crisis. The complications of the vasoocclusive crisis include acute chest syndrome, stroke, avascular necrosis of 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 complication occurs in the SCA children (5-7). Sudden acute onset of facial paralysis i.e. Bell’s palsy during childhood is less common than adults. In 1830, the first case of unilateral facial weakness was described by Sir Charles Bell. The approximate incidence of Bell’s palsy ranging 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 cause like birth trauma, genetic and acquired causes (like infection, inflammation, and trauma) of facial nerve paralysis need to be ruled out.
before the Bell’s palsy diagnosis is made. For the diagnosis of Bell’s palsy, there are no confirmatory laboratory tests available. Radiological and serological investigations are indicated in case of persistent facial weakness to exclude the neurological conditions like neuroma or lyme disease (9). Lumbar puncture are mainly avoided but should be considered for severe cases. To rule out the central nervous system (CNS) tumour, magnetic resonance imaging or computed tomography can be used. Most of the treating physician are depends on the symptoms and clinical signs for the diagnosis and management of the Bell’s palsy (9-11).

After excluding the other causes of acute peripheral palsy, the diagnosis can be made. Still there is ongoing debate about the role of antiviral and steroid in treatment of the condition. Though residual dysfunction may rarely see but the prognosis is good. Ogundunmade et al. (10) reported one adult case of sickle cell anemia with bilateral facial nerve palsy. Till date, unilateral facial nerve palsy in SCA child have not been reported in the literature. This is the first case of unilateral facial palsy associated with SCA to be reported.

4- CONCLUSION

In children, Bell's palsy is a self-limiting condition with a good prognosis. The diagnosis was made on the basis of clinical findings. So, 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.

6- REFERENCES


