

## Case Report

## MENTAL NERVE NEUROPATHY IN SICKLE CELL DISEASE: REPORT OF FIVE CASES

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العديد من التغيرات الوراثية الشاذة شوهدت مع بعض الشكايات السرية مصحوبة بفقر الدم المنجلي خاصة أذية العصب الذقني قُدرت بخمس حالات حيث أن مريضين منهم شوهدا قبل خلع الأسنان والمريض الثالث ما بعد خلع السن والرابع أنثى تلو ولادة قيصرية وآخرهم يشكو من ضرر غير معروف السبب .

المعالجة كانت الخيار الوحيد وقد استنتج من هذا أن أذية العصب الذقني مع أنها غير شائعة ولكنها قد تحدث قبل أو بعد خلع الأسنان وفقر الدم المنجلي كجزء من ظاهرة انسداد الأوعية الدموية .

Several genetic abnormalities and clinical problems are associated with sickle cell disease, particularly mental nerve neuropathy. These abnormalities are exemplified in the five cases in this report. The five cases focused on two patients who were presented before dental extraction, one patient after teeth extraction, one patient following caesarian section and the last one with idiopathic numbness. Conservative and supportive management seemed to be the only treatment protocol employed. It was concluded that mental nerve neuropathy, although uncommon, can occur before or after tooth extraction in sickle cell diseased patients as a part of the vaso-occlusive phenomenon.

## Introduction

Sickle cell disease is a hereditary condition which is common in Africa, the Mediterranean countries, and India and their descendants who have migrated to Europe and South, Central and North America and Bahrain. Sickle cell disease is a result of the alteration in the globin portion of the hemoglobin.<sup>2</sup> When only one of the B-chains is affected, the condition is referred to as sickle cell trait or heterozygous sickle cells disease - HbAS. The patient may have

between 60 to 80% of normal hemoglobin and exhibits virtually no symptoms of sickle cell disease. When both chains are affected, the condition is known as homozygous sickle cell disease - HbSS and gives rise to the very severe symptoms of sickle cell anemia. In this condition, 75 to 100% of the hemoglobin is the S type, and the rest are fetal hemoglobin.

In the deoxygenated state, the red cells become extremely fragile, undergo massive premature hemolysis giving rise to severe aplastic anemia, aggregate, slow down the circulation and very often occlude small blood vessels. The vaso-occlusive episodes cause infarction, severe pain and necrosis in various organs. Among the organs frequently affected is the bony skeleton.<sup>4</sup>

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Anemia and the vaso-occlusive crisis are the cardinal signs of sickle cell disease. Frequent hemolysis and anemia demand increased erythropoiesis by the expansion of the bone marrow. The jaws exhibit radiological features of increased radiolucency, thinning of the inferior border of the mandible and presence of areas of increased radiopacity. Profound anesthesia following dental extraction under local anesthesia in sickle cell diseased patients is a rare complication. Few cases of numbness have been reported in the literature before and after dental extraction. This study focuses on five cases of mental nerve neuropathy in sickle cell patients.

history of repeated admission for treatment of sickle cell crisis. The initial clinical examination showed a moderately swollen lower lip and organized clot in the socket of both mandibular first molars. There was a complete profound anesthesia of the lower lip and those areas supplied by the mental nerve felt like wooden blocks, as demonstrated by the needle-prick test (Fig. 1). Orthopantomograph (OPG) taken at this examination demonstrated bilateral multiple radiolucencies and decreased trabeculations in the mandible (Fig. 2).

The hemoglobin level was 9.9 gm/100ml and the hematocrit reading was

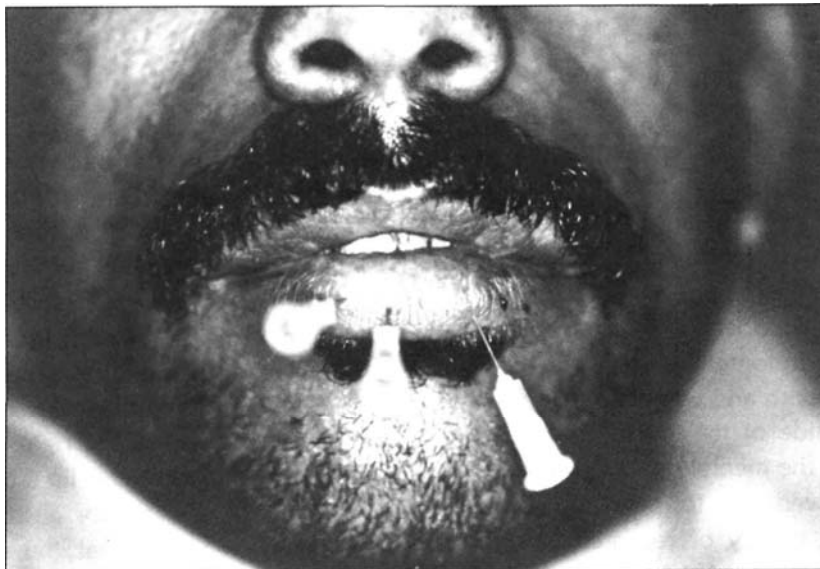


Fig. 1. Needle-prick test of the lower lip demonstrating profound anesthesia.

### Case 1

A 28 years old man was referred by his private dental practitioner, complaining of complete numbness of the lower lip for a two-week duration. He reported that the lower first molars were extracted under local anesthesia successively in two weeks time. His past medical history showed that he is a known case of sickle cell disease with

29%. The white blood cell count was 10,200 with a normal differential. The patient was admitted in the hospital and received supportive therapy consisting of intravenous fluids, orally administered analgesics and oxygen. Bone scans were undertaken and showed increased uptake in both sides of the mandible. The patient has been followed up for the past 6 months in the outpatient department and seems to have improved significantly with the



Fig. 2. Panoramic radiograph demonstrating multiple radiolucencies and decreased trabeculations in the mandible.

gradual return of normal sensation to the lower lip.

#### Case 2

A 30 years old female patient was referred from the orthopedic department due to her inability to open her mouth for a one week duration. Clinical and radiological examination revealed severe limited mouth opening measuring 3 mm due to a periodontally involved lower left first molar tooth and complete paraesthesia of the left side of the lower lip and areas supplied by the mental nerve. Her hospital record showed her to have a sickle cell disease and history of repeated admission for the treatment of sickle cell crisis. She had total hip replacement for avascular necrosis of the femoral head one year ago. Hematologically, there was a hypochromic microcytic anemia, the hemoglobin being 9.5 g/dl and the blood film revealed many nucleated erythrocytes. Oral antibiotics was given and mouth opening improved significantly after one week. Under local anesthesia, the lower left first molar tooth was extracted and the patient was kept on a regular follow up for the evaluation of the paraesthesia of the lower lip. One year later, there was neither improvement in the

paresthesia of the lower lip nor in the structures supplied by the mental nerve.

#### Case 3

A 32 years old sickle cell diseased male patient was referred from the department of internal medicine for the evaluation of a toothache of a one week duration. The patient's medical history showed multiple admissions for painful crisis. Clinical and radiological examination revealed a decayed lower right molar tooth and complete numbness of the right side of the mandible and lower lip along the distribution of the inferior alveolar nerve. The patient reported that the episode of numbness started a few days before the commencement of the toothache. Technetium 99m bone scan (Fig. 3) was obtained and revealed increased gallium uptake in the mandible. The findings were consistent with bone marrow infarction of the mandible. The tooth was extracted under local anesthesia. The patient is now 9 months post extraction with gradual recovery from the numbness.

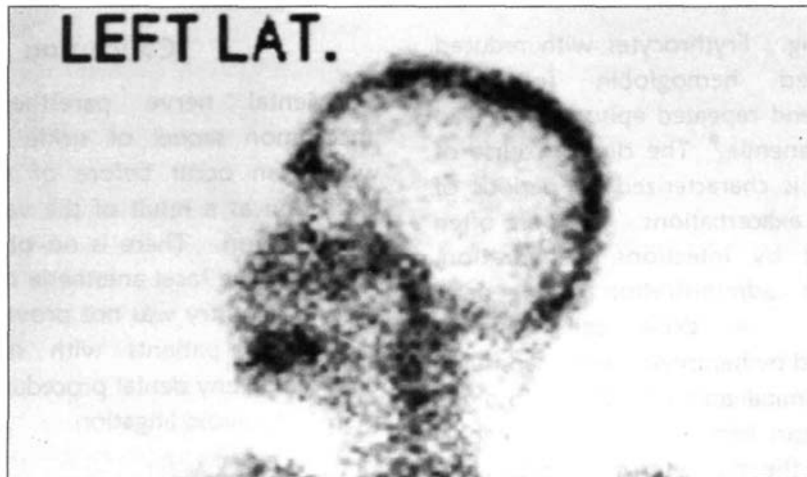


Fig. 3. Tc99m bone marrow scan reveals bone marrow infarction of the mandible.

#### Case 4

A 30 years old Bahraini lady was referred by her physician for the evaluation of lower lip numbness of three weeks duration following cesarean section. The patient is a known case of sickle disease and Glucose-6 Phosphate Dehydrogenase deficiency with frequent admissions for acute painful vaso-occlusive crisis. Clinical examination showed completely numb lower lip along the distribution of the mental nerve. Radiological examination showed generalized radiolucencies and decreased trabeculations in the mandible. The hematological status of the patient was hemoglobin level of 10.2 gm/dl. Supportive therapy was instituted, and the patient reported complete recovery from numbness nine months later.

#### Case 5

A 20 year old sickle diseased male patient was referred by his physician for the evaluation of idiopathic numbness of the left side of the lower lip and chin of three months duration. The patient reported frequent admissions for sickle cell vaso-occlusive crisis, splenectomy and frequent blood transfusion. Clinical examination

revealed complete profound numbness of the lower lip confluent with area supplied by the mental nerve as demonstrated by the needle prick test. The hemoglobin level was 8 gm/dl, and the white blood cell count was  $19 \times 10^9/l$  which is significant of infection. Supportive therapy was instituted. Six months later, gradual recovery from the numbness was noted.

#### Discussion

Sickle cell anemia is a hereditary chronic hemolytic anemia. In the homozygous person, the erythrocytes contain 70 to 100% of altered pigment that may undergo sickling even within the physiologic range of oxygen tension.<sup>7</sup> Sickling is a process in which the altered hemoglobin pigment with a low coefficient of solubility forms a semisolid gel. Tactoids, the crescent-shaped rods composing the gel, distort the shape of the normally biconcave red blood cells and render it fragile. The cells are unable to pass through small blood vessels and are less able to withstand the mechanical trauma caused by the circulation through the organs of the body. When sickled red blood cells become trapped in the smaller blood vessels, erythrosthiasis occurs. Plugs of sickled erythrocytes become solid and occlude the vessel, a thrombus forms and infarction follows. Deoxygenation and a reduced pH

favor sickling. Erythrocytes with reduced and altered hemoglobin frequently hemolyze, and repeated episodes will lead to severe anemia. The clinical course of the disease is characterized by periods of latency and exacerbations. Crisis are often precipitated by infections, dehydration, trauma or administration of general anesthesia. A sickle cell crisis is characterized by hemolysis, fever, sepsis and severe abdominal and joint pain. No organ system appears immune from the potential damage of the micro-thrombotic ischemic process of the disease. Possible neurologic signs may include coma, convulsions, cranial nerve palsies and headaches. The lesions in the central nervous system are most often due to thrombi precipitated by the sickling process. Peripheral neuropathies associated with sickle cell crisis have been reported.

Konotey-Ahulu described five cases with symptoms of anesthesia of the mental nerve. He found that 4% of patients with repeated sickle cell crisis have moderate to severe pain in the mandible. An undetermined portion of these patients developed altered sensation of the lower lip followed by numbness. The anesthesia was unilateral and along the distribution of the mental nerve. The mental nerve has probably become infarcted during sickle cell crisis as it exits the mental canal. Recovery of the paresthesia is usually slow lasting as long as 18 months.

In cases 2 and 3, paresthesia was present at the same time of referral, i.e. before dental extraction. It can be hypothesized that the paresthesia was part of the vaso-occlusive episode of the sickle cell disease. Under these circumstances, it is important for the dentist to be aware of this sign and to document the above sign before dental extraction or he could face a medico-legal complaint of producing the paresthesia iatrogenically during dental extraction.

## Conclusion

Mental nerve paresthesia is an uncommon sequel of sickle cell disease which can occur before or after dental extraction as a result of the vaso-occlusive phenomenon. There is no obvious cause for it and the local anesthesia or any drugs used in dentistry was not proven to be the cause, but patients with such disease undergoing any dental procedure should be warned to avoid litigation.

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