Mental Nerve Neuropathy: A Rare Manifestation in Sickle Cell Disease

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Abstract

Mental nerve neuropathy is a peripheral sensory neuropathy, characterized by acute numbness of the chin area. It is a rare entity with diverse aetiology including, among others, local odontogenic causes and malignancy. In rare cases, it might be associated with sickle cell disease, due to the combined presence of hyperviscosity and the sinuous course of the mental nerve and artery through the mental foramen. The authors present the case of an adolescent girl with numb chin symptoms during a multifocal sickle cell crisis. The aim is to briefly review the causes of numb chin syndrome, emphasizing the differential diagnosis in sickle cell patients.

Keywords: Adolescent; Anaemia, Sickle Cell/complications; Cranial Nerve Diseases/aetiology

Introduction

Mental nerve neuropathy or numb chin syndrome is a sensory peripheral neuropathy of the mental nerve, a terminal branch of the mandibular nerve, which is itself a branch of the trigeminal nerve. It is characterised by the sudden onset of hypoaesthesia or paraesthesia in its area of sensory distribution (chin, lower lip and the lower incisors and canine teeth's gingival tissue).[1-3]

The sinuous path of the mental nerve and artery through the mandibular canal predisposes to nerve compression and/or ischaemia,[1-4] which are associated with different aetiologies (Table 1).

Local benign mandibular/dental causes (for example, dental procedures, trauma, dental caries and dental abscesses) are the most frequent causes.[2] In their absence other conditions must be excluded, especially local or metastatic cancer.[2,4] In adults an association with solid tumours and lymphoproliferative disorders is described, caused by nerve compression and/or infiltration. In these cases, numb chin can be the presenting feature and is usually a poor prognostic indicator.[1-6]

The association between mental nerve neuropathy and sickle cell disease is rare, described for the first time in 1972 by Konotey-Ahulu.[1,3,4] The few reported cases in literature are of adult patients with predominantly unilateral symptoms.

Case Report

Seventeen-year-old female adolescent, born in Angola, with sickle cell diagnosis at 18 months (SS phenotype, Bantu/Bantu haplotype) and alpha+ thalassemia homozygosity.

She had a severe clinical course with multiple hospital admissions due to vaso-occlusive crisis, osteonecrosis of the humeral head and ischemic stroke at the age of 12 (as consequence of severe cerebral vasculopathy that progressed to Moyamoya disease). The patient underwent cranial revascularization surgery and was receiving monthly transfusions.

The patient was admitted to the emergency department with painful shoulders, left thigh and lower back over the previous twenty-four hours, consistent with a multifocal vaso-occlusive crisis. She also mentioned continued chin and lower lip hypoaesthesia, also frequent in previous episodes of vaso-occlusive crisis, devalued by the patient as due to its self-limiting and transient nature (minutes). The patient denied having fever, respiratory or gastrointestinal symptoms, or other neurological complaints, including headache, paresis or visual complaints. The last exchange transfusion dated back five weeks, with the one-week delay being due to school tests.

On admission, the patient was afebrile, pale and anicteric. She presented a grade II/VI systolic murmur in the left sternal border, and she did not present any articular inflammatory signs. Neurological examination

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showed an already known left brachial monoparesis and *de novo* sensory changes characterised by bilateral hypoaesthesia of the chin, lower lip and lower incisors' gingival tissue. The patient did not present other changes in physical examination, including the oral cavity and mandibular area. Laboratory evaluation revealed haemoglobin 9.5 g/dL (reference values 12.0-16.0 g/dL; basal baseline value 10 g/dL), haemoglobin S fraction of 60.6%, with stable usual haemolysis parameters and no elevation of inflammation/infection parameters. The patient was admitted with the diagnosis of vaso-occlusive crisis for intravenous fluid therapy and analgesia, requiring morphine infusion until the fourth day of hospitalisation due to persistent pain. Due to refractory pain, she underwent an exchange transfusion lowering haemoglobin S fraction to 26%. A general improvement was observed together with the resolution of pain, and the patient was discharged after nine days. Despite the favourable clinical evolution, she maintained chin and lower lip hypoaesthesia during hospitalisation, although the affected area was reduced. Only two months after discharge the described sensory changes were completely resolved.

Discussion

This case report describes mental nerve neuropathy associated with sickle cell disease.

Mental nerve neuropathy is a rare complication of a vaso-occlusive sickle cell crisis, present in approximately 4% of cases.[7] It results from microinfarcts in the mental nerve due to the hyperviscosity inherent to the underlying disease and the sinuous path of the mental nerve and artery through the mandibular canal.[1-4] In general, the hypoaesthesia is unilateral and preceded by local bone pain; however, cases of bilaterality (15%) and absence of previous pain, as in the case mentioned hereinabove, were described.[1-3] The mandibular magnetic resonance imaging may reveal high signal in T2, which is not observed in T1 sequences, indicative of local bone microinfarcts.[3] Nonetheless, mental nerve neuropathy is a clinical diagnosis,[1] as only 35.7% of patients in a vaso-occlusive crisis show these lesions in the magnetic resonance imaging.[8] Treatment is supportive, with intravenous fluid therapy and analgesia.[1] Sensory recovery of the mental nerve distribution area is indolent, and it may take up to several months after the resolution of the vaso-occlusive crisis.[1,3,4]

Mandibular osteomyelitis as an infectious cause for this clinical condition was considered unlikely in this patient, given its early presence during the vaso-occlusive crisis, the absence of fever, local inflammatory signs and laboratory parameters of infection. Also, self-report of similar complaints during previous painful crisis excludes the diagnosis.

On the other hand, the presence of *de novo* neurological changes, in a patient with history of severe cerebral vasculopathy, associated with the delay of the last exchange transfusion (haemoglobin S 60.6%), require a careful neurological assessment to exclude a cerebrovascular cause. However, this hypothesis was also

Table 1. Several aetiologies of mental nerve neuropathy[2,5]

Malignant causes
Breast cancer
Lymphomas and leukaemia
Bronchial and lung cancer
Prostate cancer
Thyroid cancer
Renal cancer
Melanoma
Gastrointestinal cancer
Multiple myeloma
Head and neck tumours
Non-malignant causes
Odontogenic causes:
Dental abscess
Periapical dental infection
Odontogenic cysts
Traumatic and/or iatrogenic causes:
Facial trauma
Dental procedures / anaesthesia
Drugs (bisphosphonates, mefloquine)
Infectious causes:
Mandibular osteomyelitis
Human immunodeficiency virus infection
Lyme Disease
Syphilis
Inflammatory and/or autoimmune causes:
Vasculitis
Sjögren Syndrome
Giant cell arteritis
Diabetes *mellitus*
Systemic lupus erythematosus
Demyelinating diseases
Other causes:
Sickle cell disease
Sarcoidosis
Amyloidosis
Hypertriglyceridaemia
Aneurysms

unlikely as the clinical manifestations were bilateral, and the rest of the neurological examination was unremarkable. In addition, the thorough delimitation of the affected area – limited to the chin, lower lip and lower teeth's gingival tissue –, was crucial to identify the exclusive involvement of the mental nerve, in contrast with the absence of complaints in areas asociated with more proximal branches of the trigeminal nerve. As such, no further studies were performed.

Considering other aetiologies for mental nerve neuropathy, non-related with sickle cell disease, such as a local odontogenic cause, was not found. Moreover, the hypothesis of an underlying tumour was unlikely due to the age group and past history.

WHAT THIS CASE REPORT ADDS

- Mental nerve neuropathy is characterised by acute hypoaesthesia and/or paraesthesia in the mental nerve sensory distribution area, with multiple possible aetiologies.
- Mental nerve neuropathy is often associated with local odontogenic or mandibular causes. In their absence, malignant aetiology or other rarer systemic causes should be considered.
- In sickle cell disease, despite being rare, mental nerve neuropathy may occur in vaso-occlusive crisis. The diagnosis is clinical, and treatment includes analgesia and intravenous fluid therapy. Its resolution is spontaneous and variable, and may occur after several months.
- In contrast, other neurological manifestations of sickle cell disease should raise clinical suspicion of major disease complications due to the high risk of stroke.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

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Protection of human and animal subjects

The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Confidentiality of data

The authors declare that they have followed the protocols of their work centre on the publication of patient data.

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