Temporomandibular joint arthritis in sickle cell disease: a case report

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We report a rare case of aseptic arthritis in the temporomandibular joint of a patient with sickle cell anemia. A 22-year-old woman with sickle cell disease, in the 18th week of gestation, was referred by her hematologist to investigate a sudden mouth opening limitation and severe pain on her left cheek. The patient received a standard pain assessment protocol, clinical examination, and complementary exams (complete blood count, hemoglobin electrophoresis, blood solubility test, panoramic radiograph, and magnetic resonance imaging [MRI]). The blood results were consistent with a sickle cell crisis and the MRI showed an inflammatory process around the left temporomandibular joint. Treatment with opioid analgesics and blood transfusion provided good results. Sickle cell anemia is a disease that can cause arthritis of the temporomandibular joint, and although it is rare, clinicians should be attentive to the differential diagnosis in patients with this disease. (Oral Surg Oral Med Oral Pathol Oral Radiol 2013;115:e31-e35)

Sickle cell anemia is an autosomal recessive disease with high prevalence in the world as well as in Brazil (1 in every 200 births)[1] and consists of the clinical expression of a homozygous hemoglobin S gene, where polymerized hemoglobin leads to chronic hemolytic anemia.[2] It is characterized by vaso-occlusive crises that affect many organs and manifests primarily by crises of acute pain.[3] The triad of ischemia, infarction, and inflammation contributes to the pathophysiology of pain in this condition.[4]

The interaction between erythrocytes and the vascular endothelium is critical in the pathogenesis of painful crises that occur during sickle cell anemia. This process involves hemoglobin S polymerization, subsequent red cell distortion, interaction of erythrocytes with adhesion proteins of the vascular endothelium, and an ensuing inflammatory response that further increases cellular adhesiveness. In sickle cell disease, erythrocytes upregulate the expression of intercellular adhesion molecule-1 and vascular cell adhesion molecule-1 in vascular endothelial cells in vitro. Heme and heme oxygenase, the heme-degrading enzyme, are also potent inducers of inflammation and endothelial cell adhesion, as is phosphatidylserine when exposed on the surface of red cells. Increased adhesion and the subsequent inflammatory response decrease blood flow, leading to further sickling in an increasingly hypoxic and acidic environment. These vaso-occlusive crises are typically manifested by pain. Repeated episodes of decreased blood flow can lead to the impaired nourishment of critical structures (e.g., femoral head, vertebral bodies, or joint), leading to bony lesions with loss of trabeculae (e.g., avascular necrosis, vertebral collapse).[5-13]

Nearly half of sickle cell patients (49%) experience orofacial pain. Sixty-eight percent experience pulpal necrosis without apparent cause and 77% experience headaches.[14] Complications such as mandibular osteomyelitis and neuropathy, fibrous ankylosis, and asymptomatic pulpal necrosis have been reported.[15-18] Limb joint pain is common, but temporomandibular joint (TMJ) pain is rare.[19] Here, we report a patient who had aseptic arthritis of the TMJ and sickle cell anemia, focusing mainly on a multidisciplinary assessment and the treatment outcome of the patient.

CASE REPORT

A 22-year-old woman in the 18th week of pregnancy was admitted to the hematology ward after experiencing a throbbing pain for 2 days in the left preauricular region. The pain was constant and severe, scoring 10 on the Visual Analog

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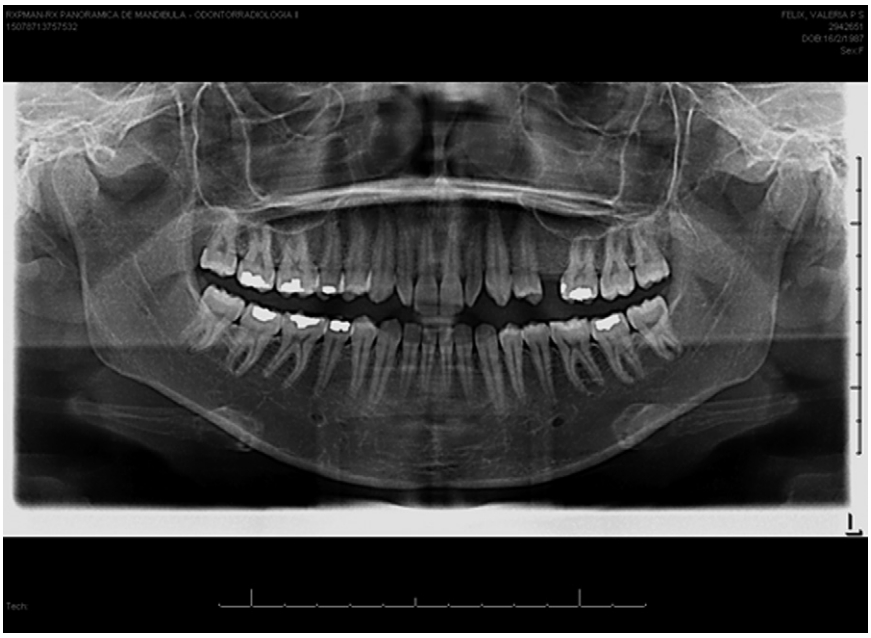

Fig. 1. Panoramic radiographs show enlargement of the trabecular bone in the symphyseal region, bilateral mandibular body, and apices of the left and right mandibular third molars.

Scale, which ranges from 0 (no pain) to 10 (worst pain). The pain increased when she reclined, chewed, or talked and only subsided with parenteral opioid analgesia. She denied fever or trauma.

Her medical history at the time of her pregnancy included various complications associated with sickle cell anemia: frequent hospitalizations caused by painful crises and episodes of pneumonia since childhood; ischemic stroke at 10 years old without sequelae; cholecystectomy for cholelithiasis; femoral and humeral head osteonecrosis; transfusional iron overload; and a recent splenic sequestration crisis. Apart from folic acid supplementation and iron chelation therapy, she had been treated with hydroxycarbamide for 3 years.

Findings at physical examination included mild edema, allodynia, and hyperalgesia in the left preauricular region, hyperalgesia of the left masseter muscle, limited interincisal mouth opening (11 mm), and pain upon mandibular movement. Only 1 tooth was missing, a maxillary left premolar. No other dental or periodontal abnormalities were observed during the visual examination, with thermal or percussion testing, or on panoramic radiograph (Figure 1).

The following complementary tests were performed: complete blood count, biochemical tests, hemoglobin electrophoresis (Tables I and II), and magnetic resonance imaging (MRI; Figure 2). Arthritis in the left condylar head secondary to a sickle cell crisis was diagnosed after the analysis of the laboratory tests and images. The final diagnosis rendered was aseptic arthritis of the left mandibular condyle secondary to a sickle cell crisis.

Treatment

The patient was admitted to the hematology ward and received standard analgesia recommended for treatment of severe pain: parenteral opioid analgesia and nonsteroidal anti-inflammatory drugs (NSAIDS; naproxen). She also received a blood transfusion because of worsening anemia.

The pain gradually subsided after 2 days of hospital stay, allowing progressive weaning from the analgesic treatment. Mouth opening amplitude improved substantially to 28 mm interincisal opening (initially 11 mm). After 1 week, there was resolution of facial pain and edema; thus, the analgesic and anti-inflammatory medications were discontinued at this time. The patient was followed for 6 months without relapse of facial pain.

DISCUSSION

This rare case of arthritis of the left condylar head illustrates an uncommon situation of joint pain of systemic origin, although vaso-occlusive crises associated with sickle cell anemia often cause pain. The pain lasted 10 days and was accompanied by leukocytosis. The clinical signs and symptoms were consistent with the diagnosis of sickle cell crisis.[20,21]

The patient had a systemic disease that affects bones and joints, including TMJ. According to the diagnostic criteria of the American Academy of Orofacial Pain, the diagnosis requires the presence of a clearly documented disease or event associated with osteoarthritis, functional pain, local tenderness, limited TMJ movements and deviation to the affected side, crepitation or multiple joint noises,[22] and manifestations of arthritis and necrosis in other joints. The present case met all these criteria. Although the classification of polyarthritis does not contemplate sickle cell disease, it should be