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CASE REPORT

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Pre-splenectomy permanent tooth extraction in a child with hereditary spherocytosis: a case report and guidelines care

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Abstract

Hereditary Spherocytosis (HS) is a type of hemolytic anemia characterized by a defect in the membrane of red blood cells, which causes them to assume a spherical shape, become osmotically fragile, and be prone to early hemolysis. While it is relatively well understood in the medical field, there is limited evidence regarding the dental management of patients with this condition. Therefore, this report aims to present a case involving the extraction of permanent teeth before splenectomy in a child with HS who had been hospitalized. Additionally, treatment guidelines for these patients are proposed and developed. The patient was scheduled to undergo a total splenectomy due to splenomegaly resulting from severe hemolytic anemia. Prior to the surgery, the child was referred to the hospital's dental clinic with a chief complaint of dental pain. An intraoral examination revealed carious lesions with pulpal involvement in the first molars, and extraction was recommended to minimize the risk of post-splenectomy infection. Hematological support was provided during dental treatment since partial control of the anemia was necessary for the extraction procedure. Developing dental treatment guidelines for patients with HS is essential to ensure their safety.

Keywords

Hemolytic anemia; Hereditary spherocytosis; Dental care for children

1. Introduction

Hereditary Spherocytosis (HS) has been recognized as one of the most common hemolytic anemias globally, particularly in Northern Europe [1]. It can result from either an autosomal dominant or recessive chromosomal mutation or a single gene mutation that is not inherited from the parents [2]. HS is typically characterized by a defect in the structural proteins of red blood cell membranes, causing them to assume a spherical shape (instead of the usual biconcave shape), become osmotically fragile, and be more susceptible to early hemolysis [3, 4]. Additionally, the marked destruction of red blood cells by the spleen leads to an increase in its size, resulting in splenomegaly, which is a common clinical symptom in children with HS [1].

A diagnosis of HS can be made after conducting a thorough anamnesis and physical and laboratory examinations (*e.g.*, complete blood count, bilirubin count and blood smear) in patients exhibiting symptoms such as splenomegaly, jaundice and anemia. The diagnosis can be confirmed through a conventional incubated osmotic fragility test. While the diagnosis is relatively straightforward in most cases due to the presence of a positive family history and distinctive clinical and laboratory features, some cases may go undetected [5]. The clinical and therapeutic characteristics exhibited by the patient depend on the severity of hemolytic anemia, leading to the classification of HS as mild, moderate or severe [3].

Although HS is relatively well understood in the medical field, there is limited evidence regarding the dental implications of this condition. Therefore, this case report aims to summarize pre-splenectomy dental treatment in a child hospitalized with HS and proposes a set of guidelines for this type of care.

2. Case report

2.1 Patient information

A 10-year-old male patient with a brown complexion, residing in the countryside of Maranhão, was admitted to a public children's hospital in the city of São Luís, Maranhão, located in northeastern Brazil. His admission was due to severe hemolytic anemia, jaundice and an enlarged spleen. During his hospitalization, he received a diagnosis of severe HS with the typical laboratory profile, despite having no family history of the disease. However, there were reports from close relatives who had experienced recurrent anemias. Genetic analysis was

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not conducted to rule out a family history due to the patient's guardians being unable to cover the costs of this examination. Given the patient's clinical condition, a total splenectomy was recommended. However, the patient reported experiencing pain in his "back teeth" and was subsequently referred to the hospital's dentistry department. Prior to admission to the hospital, the patient had not received dental care.

2.2 Clinical findings

Before initiating dental care, the patient's vital signs were assessed and found to be within the normal range for their age. No significant changes were noted during the evaluation of the headgear, and an intraoral examination revealed extensive carious lesions that had compromised the crowns of all four first permanent molars (tooth #16, 26, 36 and 46). It's important to note that the patient had never previously received dental care. Despite none of the affected teeth responding to the cold sensitivity test, the patient reported experiencing pain during vertical percussion and digital apical palpation. Unfortunately, radiographic examination could not be performed due to the absence of the necessary equipment in the hospital's dental clinic, which prevented a definitive endodontic diagnosis.

2.3 Treatment and follow-up

Single-session endodontic treatment and prosthetic rehabilitation were considered unfeasible due to the urgency of the splenectomy and the family's financial constraints. This was because the state public service did not provide these procedures, leading to the decision to extract all four teeth in a single session under general anesthesia. This approach was chosen for the purpose of time optimization and the administration of hematological supplementation. The pre-surgical evaluation included a blood count, coagulogram, fasting glucose, urea, creatinine and a cardiological assessment. The blood count evaluation confirmed severe hemolytic anemia (hemoglobin (Hb): 6.6 g/dL; platelet count: 50,000/mm^3), while all other laboratory findings were within normal limits. The patient was classified as Goldman class I after evaluation by the cardiologist.

As a pre-operative measure, hematological supplementation was requested from a pediatrician, and a transfusion of three units of fresh frozen plasma and two units of platelet concentrate was administered. Subsequently, the blood count evaluation was repeated, showing a decrease in the severity of hemolytic anemia (Hb: 10.1 g/dL; platelet count: 71,000/mm^3).

Once the patient was deemed suitable for extraction, antibiotic prophylaxis was administered using 500 mg intravenous cefazolin (as per the hospital's protocol) before the procedure commenced. The teeth were then extracted with the assistance of forceps, and no local anesthetic injections were required. Gelatin sponges were subsequently placed in the alveolar sockets, and the Chompret maneuver was employed. Surgical closure was achieved using absorbable "X" sutures (Fig. 1).

After the procedure was completed, the patient received an additional transfusion of three units of fresh frozen plasma, two units of platelet concentrate, and one unit of compact cell volume under medical prescription. A repeat blood count evaluation showed that the severity of the hemolytic anemia remained mild (Hb: 9.9 g/dL; platelet count: 65,000/mm^3). No postoperative complications were observed, and the patient underwent a successful total splenectomy 15 days later, followed by discharge from the hospital three weeks after that. Throughout the patient's entire hospitalization, the dentistry team visited him every two days to manage and provide guidance on the oral hygiene of the child and the guardian.

Hereditary Spherocytosis (HS) was first reported in 1871 by Vanlair and Masius, who observed spherical microcytic red blood cells and marked splenomegaly, without hepatomegaly, in a patient with jaundice. Initially named Minkowski-Chauffard Disease, it wasn't until 1900 that Oskar Minkowski and Antonele Marie Émile Chauffard defined HS as we know it today [1]. This disease is generally observed in the Caucasian population, with a prevalence of 1 in 2000. Being a genetic condition, it is necessary to understand the patterns of its transmission. Thus, this pathology presents itself in 75% of cases as autosomal dominant, with the remaining 25% of cases associated with recessive patterns [6].

The primary clinical feature of HS, hemolytic anemia [6], can increase the risk of osteomyelitis and interfere with invasive dental treatment procedures that induce inflammation, as it can lead to vascular stasis, hypoxia, ischemia, necrosis and secondary infections. The mandible is usually affected to a greater extent due to its poorer blood supply compared to the maxilla [7]. HS patients are also predisposed to heart failure (HF), which can either cause anemia or occur as a consequence of it [8, 9]. This emphasizes the importance of a thorough anamnesis and clinical examination for proper dental treatment planning.

Patients with hemolytic anemia are more likely to develop skin and mucous membrane pallor, lingual papillae atrophy, dental dyschromia, microdontia, hypomineralization and pulpitis [10]. Physical fatigue associated with systemic diseases can also reduce the frequency of oral hygiene practices and their effectiveness, although a direct relationship between the two has not yet been established. Combined with the constant intake of sucrose-containing medications, this can contribute to the development of caries and periodontal disease [11]. In this case, the patient's oral condition also reflected the lack of basic sanitation in the interior regions of the city where he resided.

The patient presented with a chief complaint of dental pain and discomfort, leading to the decision to carry out extractions under general anesthesia rather than outpatient endodontic treatment. This decision, made in collaboration with the patient's family, was based on the urgency of the total splenectomy, the family's financial constraints, and the distance between the patient's residence and the hospital. The duration, discomfort and costs associated with travel were likely to result in treatment delays or abandonment. Thus, despite endodontic intervention being the more conservative treatment option, extraction under general anesthesia was recommended due to the urgency of treatment, financial constraints, and the lack of support for endodontic treatment at the hospital.

Considering the patient's severe anemia (Hb: 6.6 g/dL), the dental procedures were carried out in a single session to minimize the extent of necessary hematological manipulation. In this case, only two instances of hematological support under

medical prescription were required, one pre-surgical, which decreased the severity of the hemolytic anemia (Hb: 10.1 g/dL), and the other post-surgical to aid in healing.

Despite its benefits for extensive dental procedures that induce stress, general anesthesia in patients with HS can be challenging. A thorough evaluation by an anesthesiologist and a hematologist [12], a complete blood count assessment, and consideration of the hematological support required based on the patient's overall health are essential [13]. Thus, direct collaboration between the dentist and the multidisciplinary medical team responsible for the patient's care is crucial [14].

It is worth noting that hyposplenic patients, such as the one reported here, or splenectomized patients, are immunosuppressed and should receive antibiotic prophylaxis following the guidelines of the American Heart Association [15]. Although current evidence on antibiotic prophylaxis in these patients is debated, Okabayashi *et al.* [16] found that individuals with immune deficiencies were more susceptible to infections from any microorganism. Additionally, Pasternack *et al.* [17] recommended the use of antibiotic prophylaxis in patients younger than 16 years or older than 50 years to effectively reduce the risk of infectious complications.

Following extraction, gelatin sponges were placed in the alveolar sockets, followed by the Chompret maneuver and the placement of "X" sutures. These measures were taken to prevent trans or postoperative hemorrhage, always taking into account the techniques and the patient's clinical condition.

Therefore, it is crucial for dental professionals to have a clear understanding of the specific characteristics of patients with HS. Table 1 summarizes general guidelines for dental treatment in this population [18].

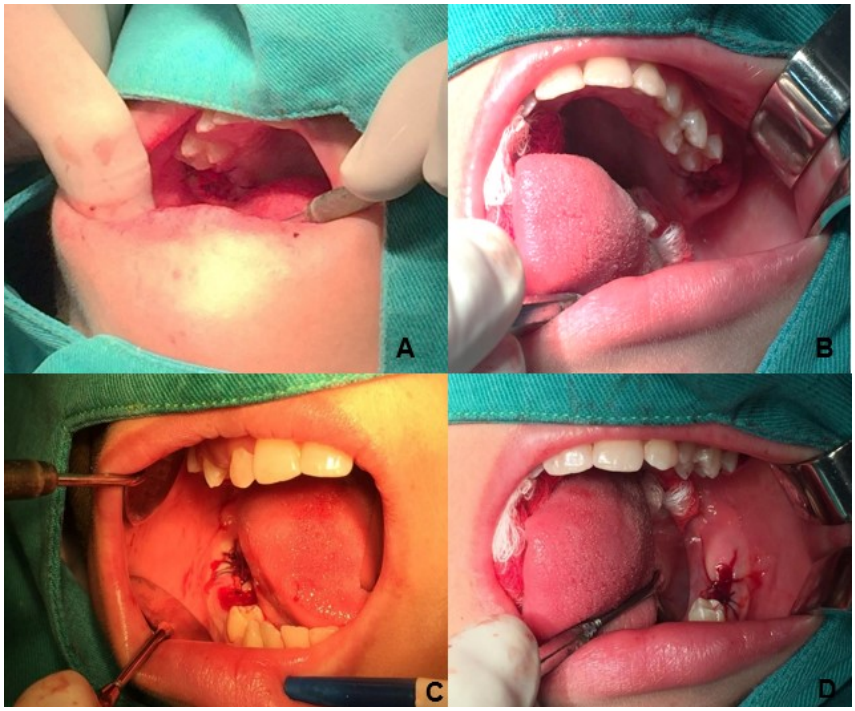

FIGURE 1. Alveolar region after tooth extraction #16 (A), tooth #26 (B), tooth #46 (C) and tooth #36 (D).

TABLE 1. Dental care for patients with hereditary spherocytosis.

Dental care	Goal
Comprehensive medical history	Appropriate, individualized dental treatment planning based on the severity of HS and factors that may interfere before, during and after the procedure.
Complete blood count	Patients with HS typically present with hemolytic anemia, and a blood count evaluation must be conducted, with the findings taken into consideration when planning treatments.
Cardiovascular assessment	In anemic patients, the cardiovascular system is strained due to an elevated cardiac output, thereby increasing the risk of heart failure.
Hematological support	This should be evaluated in coordination with the attending physician to enable compensation for hemolytic anemia and improve the patient's prognosis.
Antibiotic prophylaxis	This is important because hyposplenic or splenectomized patients are immunosuppressed.
Hemostatic techniques	These maneuvers aid in the regeneration of cortical bones, cessation of bleeding, and prevention of hemorrhaging.

HS: Hereditary Spherocytosis.

3. Conclusions

In conclusion, direct interaction between dental professionals and the medical teams caring for patients diagnosed with HS, and requiring pre-splenectomy dental surgical intervention to eliminate infection foci, is crucial. Furthermore, proper pre-splenectomy treatment planning, continuous monitoring during the trans- and postoperative phases, and comprehensive laboratory examinations are equally significant in reducing the risk of complications in patients with severe disease. The establishment of dental treatment guidelines for HS patients is imperative to ensure their safety.

AVAILABILITY OF DATA AND MATERIALS

All the datasets on which they rely, as presented in the manuscript, are already included in the article.

AUTHOR CONTRIBUTIONS

CPSC and RRJT—designed the research study. CPSC, LMMP and WJNA—conducted the research. CPSC, RRJT and PPG—wrote the manuscript. All authors contributed to editorial changes in the manuscript. All authors have read and approved the final manuscript.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

The study protocol was approved by the Ethical Committee at Ceuma University, Brazil (Approval no. 1.819.524). The authors obtained consent forms from the patients' parents. In the form, the patients' parents provided their consent for the publication of images and other clinical information related to their child's case in the journal. They also acknowledged that their child's name and initials would not be disclosed.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

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