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Surgical management in a patient with Bernard-Soulier syndrome: case report

Manejo cirúrgico em paciente com síndrome de Bernard-Soulier: relato de caso

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ABSTRACT
Bernard-Soulier syndrome is a rare inherited disease characterized by thrombocytopenia, presence of extremely large platelets and prolonged bleeding time. **Objective:** This paper aims to present an alternative management of surgical dental procedures in a female patient, faioderma, 27 years old, carrier of Bernard-Soulier syndrome. **Case Description:** On clinical examination and history, the patient reported a history of recurrent pericoronaritis associated with units 38 and 48, and a history of bleeding associated with menstruation. Imaging examination revealed impacted and impacted dental units. The treatment plan included exodontia of elements 18, 28, 38, and 48 under general anesthesia, transfusion of platelet concentrates (01U apheresis or 07U pool) two hours before surgery, fresh frozen plasma reserve in case of transoperative bleeding, and RBC concentrate reserve, according to the guidelines prescribed by the hematologist. At the moment, in a seven-month postoperative follow-up, the patient evolves without signs and symptoms of coagulation disorders associated with exodontia. **Conclusion:** Thus, it is essential that dental professionals are aware of the multidisciplinary management strategies for this group of patients in order to minimize postoperative complications.

Keywords: Perioperative Care; Oral Surgery; Bernard-Soulier Syndrome

1 INTRODUCTION

Bernard-Soulier syndrome (BSS), also known as thrombocytic hemorrhagic dystrophy, is a disease that is associated with autosomal recessive inheritance, characterized by the presence of extremely large platelets and prolonged bleeding time[1] . This syndrome was first described in 1948 by Bernard and Soulier, where they report the case of a male patient who died at the age of 28 due to intracranial hemorrhage after a fight[2].

This syndrome is inherited in an autosomal recessive manner, reported in individuals whose parents are close relatives, and is grouped with other disorders associated with large platelets, such as in Velocardiofacial Syndrome, Grey Platelet Syndrome, May-Hegglin Anomaly, Sebastian Syndrome, and Epstein Syndrome[1,3].

Macrothrombocytopenia and increased bleeding time are the two main characteristics of this syndrome[3]. The clinical manifestations usually include purpura, tendency to spontaneous bleeding, epistaxis, menorrhagia, gingival bleeding and bleeding after exodontia. Bleeding episodes are mainly associated with trauma and surgical procedures. [4]

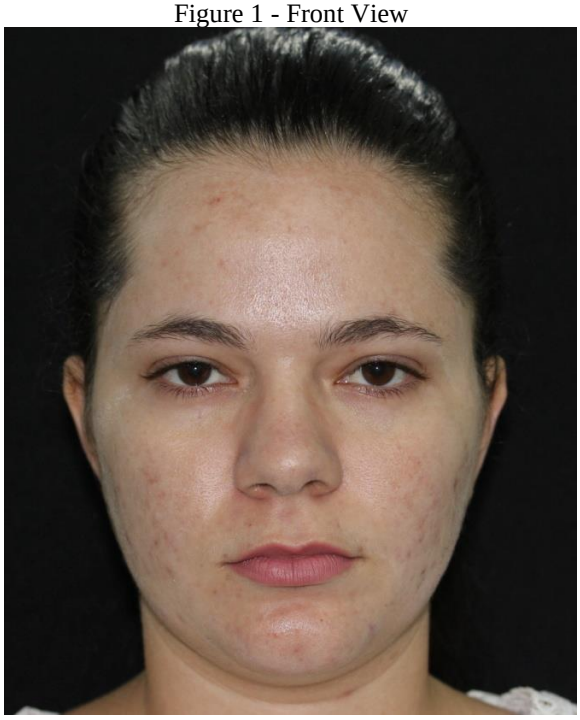
In patients with bleeding disorders, routine dental extraction can cause life-threatening complications[2]. Thus, while in congenital disorders associated with platelet dysfunction, the most frequently employed therapies in this group of patients are platelet transfusions in more severe cases, and the use of desmopressin and/or antifibrinolytic drugs in mild cases. Sometimes, the application of alternative factors and therapies is necessary, both in the preparation prior to the treatment of the patient, and in rescue therapies after intense or prolonged bleeding, for example, the reserve of fresh frozen plasma if bleeding in the transoperative period[1,3, 5].

This paper aims to present an alternative management of surgical dental procedures in a female patient with Bernard-Soulier Syndrome.

2 CASE REPORT

A female patient, brown skin, 27 years old, came to the Oral and Maxillofacial Surgery outpatient clinic of the Federal University of Bahia School of Dentistry - Salvador/BA due to pain complaints in the lower third molar regions (Figure 1).

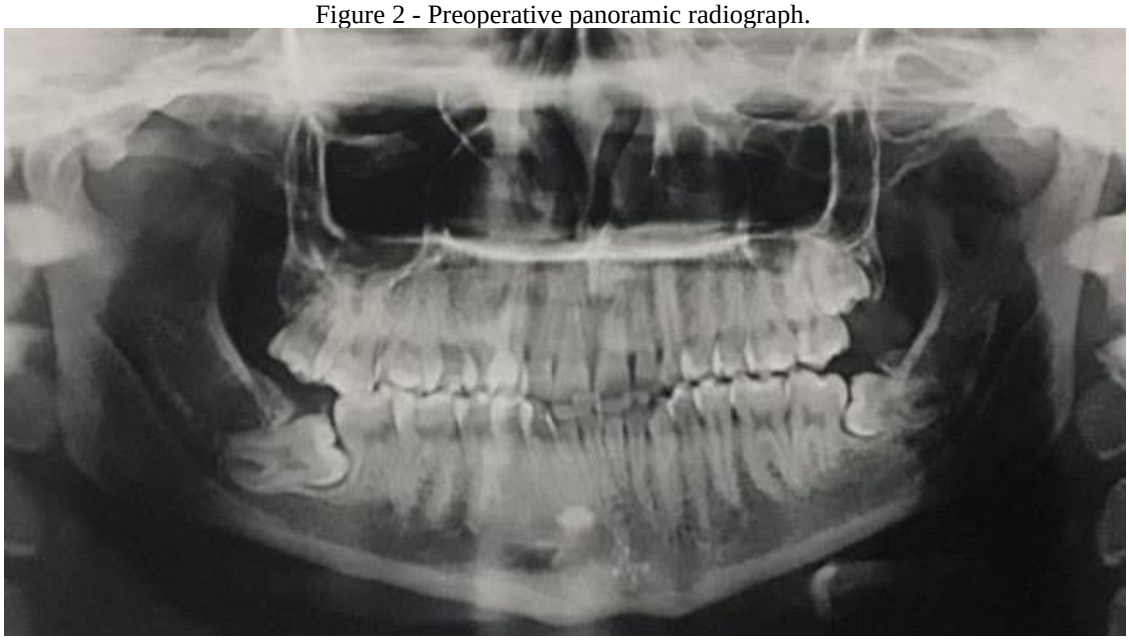
Figure 1 - Front View



The clinical analysis and anamnesis reported a history of recurrent pericoronaritis associated with units 38 and 48, in addition to a history of bleeding associated with the menstrual period. Based on the information gathered, laboratory tests were ordered, and the results were within normal limits (white blood cell count: 7190/mm3; red blood cells: 12.4 million; hematocrit:

37.9%; platelets: 329/mm3). Due to the incompatibility of the clinical and complementary findings, an evaluation with a hematologist was requested. The hematologist confirmed the diagnosis of Bernard-Soulier syndrome due to qualitative platelet deficiency. Imaging exam showed impacted and impacted dental units (38 and 48 in Pell and Gregory position 2B), mesioangulated (Figure 2).

Figure 2 - Preoperative panoramic radiograph.



The treatment plan included exodontia of elements 18, 28, 38 and 48 under general anesthesia, platelet concentrate transfusion (01U apheresis or 07U pool) two hours before surgery, fresh frozen plasma reserve in case of transoperative bleeding, and RBC concentrate reserve, according to the guidelines prescribed by the hematologist. During the surgery, the aforementioned patient did not present abnormal bleeding and evolves without bleeding in the immediate/mediate postoperative period. At the moment, in a seven-month postoperative follow-up, she evolves without signs and symptoms of coagulation disorders associated with exodontia.

3 DISCUSSION

Bernard-Soulier syndrome is characterized by a rare platelet disorder. It is more often misdiagnosed as immune thrombocytopenia, which leads to unnecessary interventions such as splenectomy[6,7]. Its prevalence, according to the literature, can be estimated at less than 1 in 1 million[1]. It involves a biochemical defect that proposes low or no amount of the glycoprotein Ib-IX-V complex[6]. This complex has two important functions, such as mediating adhesion to the

blood vessel wall and facilitating the platelet activation capacity of thrombin at lower levels, which explains the longer bleeding time observed in patients with BSS, a condition that corroborates the present report[1,7].

Among the clinical manifestations of BSS, include mucocutaneous bleeding, observed most commonly during early childhood, gingival bleeding, epistaxis, and heavy menstrual bleeding are common presentations[8]. The impact in women may be more severe due to gynecological and obstetric manifestations[9]. Menorrhagia is variable in premenopausal women and in some cases can be controlled by oral contraceptives. The orientation about the indispensability of avoiding any kind of trauma or antiplatelet medication is indispensable. [1,6]

If platelet deficiency or other bleeding dyscrasias are suspected, immediate referral for hematological evaluation is indicated, as in this case[4]. The management of this group of patients should consider the severity of the disease, the invasiveness of the planned dental procedure, and the experience of the professional responsible for the treatment[1,5]. In addition, they require local and systemic care managed together with the responsible hematologist, in a hospital environment[5,6,8].

Due to the rarity of the disorder, there are no well-defined protocols for the treatment[9,10]. However, antifibrinolytic therapies, platelet transfusions and hormone therapy have been used for prophylaxis in patients with BSS, the latter in women[8]. In this article, an alternative of satisfactory management is presented, through platelet transfusion and local measures according to the surgical act.

4 FINAL CONSIDERATIONS

Therefore, it is essential that dental surgeons are aware, above all, of the multidisciplinary management strategies for this group of patients in order to minimize postoperative complications and facilitate the treatment of patients with this rare but life-threatening hemorrhagic syndrome.

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