Comprehensive Dental Management of a Patient Suffering With Christmas Disease: A Case Report

Padmakanth Mannava[1], Rajesh K. R. Juturu[2], Pooja Walia[3], Rakshit Sthapak[4]

[1]Department of Periodontics, Geetanjali Dental and Research Institute, Udaipur, Rajasthan, India.
[2]Department of Prosthodontics, Sri Sai Dental College, Srikakulam, Andhra Pradesh, India.
[3]Department of Periodontics, Guru Gobind Singh College of Dental Science and Research Centre, Burhanpur, Madhya Pradesh, India.
[4]Department of Orthodontics and Dentofacial Orthopaedics, Triveni Institute of Dental Sciences, Hospital and Research Centre, Bilaspur, Chhattisgarh, India.

Abstract

Background: Hemophilia is an X chromosome-linked hereditary bleeding disorder due to a deficiency of coagulation factor VIII (hemophilia A) or factor IX (hemophilia B) related to mutations of the clotting factor gene. This leads to a severely increased risk of prolonged bleeding from common injuries, or in several cases, bleeds may be spontaneous and without obvious cause. Proper dental and medical evaluation of patients is, therefore, necessary before any surgical procedure. Surgical treatment of patients with bleeding disorders requires careful planning. Administration of recombinant factor VIII and factor IX concentrates to increase the coagulation factor levels is a known treatment modality. In this case presentation, a 63-year-old patient with hemophilia B reported with the chief complaint of bleeding from gums. After evaluation by the surgeon and the hematologist, periodontal flap surgery and extractions were performed with administration of preoperative four units of fresh frozen plasma.

Keywords: Hemophilia, Periodontal flap surgery, Clotting factor

Introduction

Hemophilia is an X chromosome-linked hereditary bleeding disorder due to a deficiency of coagulation factor VIII (hemophilia A) or factor IX (hemophilia B) related to mutations of the clotting factor gene.[1] Hemophilia is considered severe when plasma activity is <1 IU/dL (normal range 50–100); moderate if it ranges between 2 and 5 IU/dL, and mild if it is between 6 and 40 IU/dL.[2]

Hemophilia B, also known as Christmas disease deficiency, is transmitted in a sex-linked recessive fashion. It is a bleeding disorder caused by a deficiency or defective factor IX within the intrinsic pathway of the coagulation system.[3] Not as prevalent as hemophilia A, hemophilia B accounts for 10–15% of all hemophiliacs. Patients with severe hemophilia (<2% factor VIII or IX) have spontaneous bleeding into muscles and joints that can lead to a crippling arthropathy. Patients with moderate (2–5%) and mild (>5%) conditions usually bleed only after trauma or surgery.[4]

Therefore, dentists must be aware of the impact of bleeding disorders on the management of patients. Proper dental and medical evaluation of patients is, hence, necessary before treatment, especially if an invasive dental procedure is planned. Patients should be queried about any previous unusual bleeding episode after surgery or injury, spontaneous bleeding, and easy or frequent bruising.[5]

The majority of guidelines recommend the use of clotting factor replacement therapy before invasive oral surgery. The dose of clotting factor used varies and this may be due to problems relating to both the availability and cost of factor concentrates in different parts of the world.[6]

Successful protocols are the result of cooperation between hematologists and dentists. These protocols suggest the use of factor concentrate combined with local hemostatic techniques and measures, like suturing, and oxidized cellulose, for example, surgical or fibrin glue in conjunction with

Access this article online

Website: www.jrad.co.in

DOI: https://doi.org/10.53064/jrad.2021.12.4.21

Address for correspondence: Pooja Walia, Guru Gobind Singh College of Dental Science and Research Centre, Burhanpur - 450 331, Madhya Pradesh, India.
E-mail: pwalia16dec@gmail.com

Received: Mar. 3, 2021; **Accepted:** Apr. 22, 2021; **Published:** Jun. 29, 2021

How to cite this article: Mannava P, Juturu RKR, Walia P, Sthapak R. Comprehensive Dental Management of a Patient Suffering With Christmas Disease: A Case Report. J Res Adv Dent 2021;12:4:103-105.

postoperatively administered antifibrinolytic agents.[6] This report discusses a case of dental management of a patient with partial deficiency of factor IX.

Case Report

A 63-year-old male patient with chief complaint of bleeding from gums came to the Department of Periodontics in Geetanjali Dental and Research Institute, Udaipur, Rajasthan. He stated that he had been diagnosed with hemophilia B or Christmas disease/factor IX deficiency 20 years back. He had received hospitalization once following an accident and bleeding got arrested subsequently after a unit of whole fresh blood transfusion. The patient also had a medical history of hypertension and was on nifedipine and aspirin for 10 years.

On clinical examination, lower anteriors were found to be Grade III mobile. Generalized deep periodontal pockets (>7 mm) were present and orthopantomograph showed generalized horizontal bone loss (Figures 1 and 2).

On first appointment, only supragingival scaling was done. Periodontal flap surgery and extraction of lower anteriors were planned for next appointment.

To prevent possible bleeding during the periodontal treatment, the patient was referred to a hematologist for consultation before any procedure was performed. Hematologic data were as follows: Hemoglobin, 11.5 g/dl; bleeding time, >10 min; activated partial thromboplastin time, 34.10 s; abnormal platelet aggregation (with ADP, collagen, and epinephrine); and prothrombin time, 14.9 s.

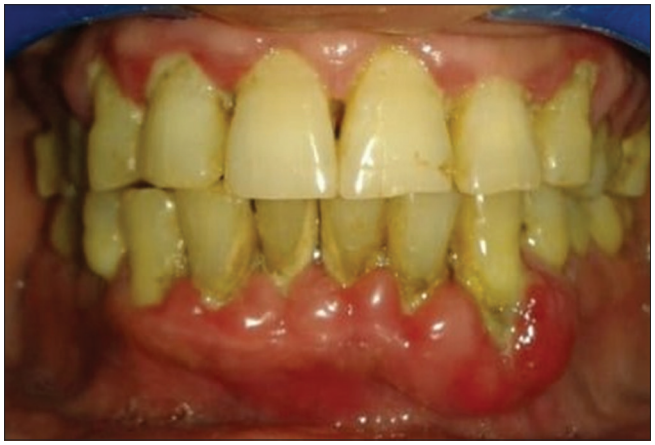

Figure 1: Pre-operative clinical photograph showing inflamed gingiva

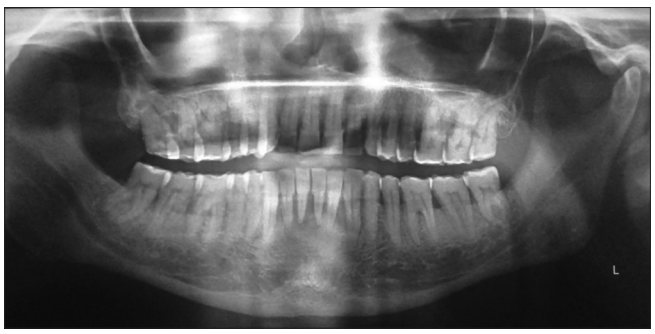

Figure 2: Orthopantomograph showing generalized horizontal bone loss

Pre-operative four units of fresh frozen plasma were given under the observation of hematologist. After local anesthesia, full mouth flap surgery and extraction of 31, 32, 41, and 42 were successfully done. After the surgery, resorbable sutures were placed using Vicryl 4–0 and non-traumatic needles (Figure 3). Immediately after surgery, again, fresh frozen plasma was given to the patient. To prevent mechanical trauma to the oral cavity, his diet was temporarily changed to include only soft foods. The patient was admitted to the hospital and was under physicians supervision for 2 days.

After 6 days of successful checkups, the 7th day patient complained of bleeding of gums on chewing hard food substance. Hence, four units of factor IX infusion were done leading to arrest of bleeding. The patient returned for reevaluation after 2, 7, and 10 days. Later, prosthetic crowns (zirconia crowns) for the anterior tooth in the lower jaw were placed. Crowns were placed with smooth margins to avoid impingement on the gingival sulcus. The patient was recalled for follow-up after 1 month and 3 months wherein healing was found to be satisfactory (Figure 4).

Discussion

Bleeding disorders are one of the most challenging medical conditions among dental health professionals. Dental practitioners should be aware that the management of these patients might require certain adjustments to treatment preoperatively and close post-operative follow-up.[7]

The decisions regarding clotting factor replacement therapy should be made in conjunction with the patient's hematologist. Successful dental treatment protocols for hemophilic patients presenting for invasive dental surgery include clotting factor replacement therapy combined with antifibrinolytic agents and local hemostatic measures.[8,9] Furthermore, it is important to

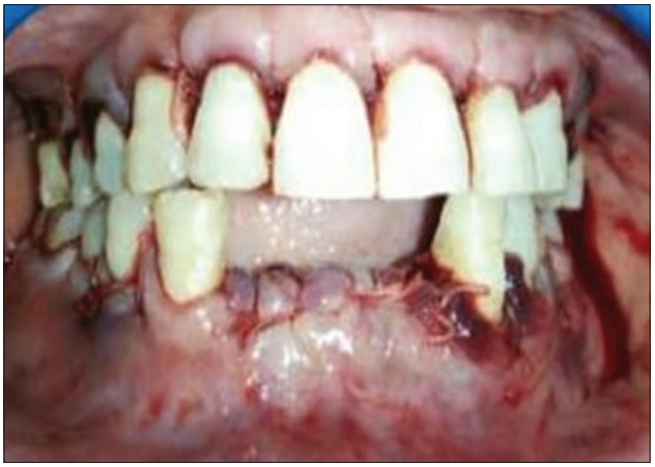

Figure 3: Immediate post-operative photograph with resorbable sutures in place

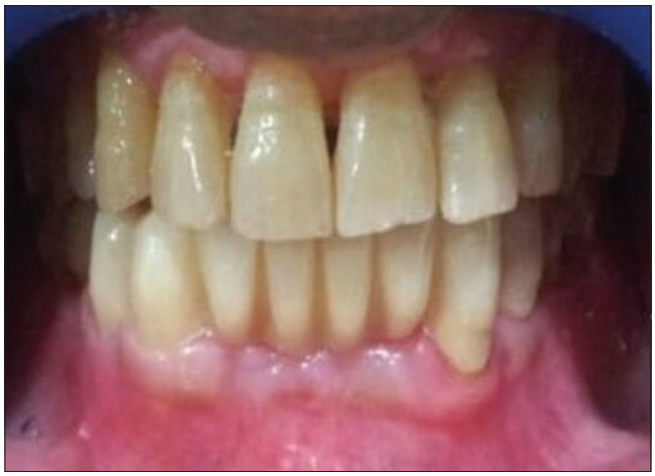

Figure 4: Post-operative clinical photograph at 1 month with prosthesis showing satisfactory healing

manage the tissues atraumatically and the lingual tissues should be left undisturbed to prevent blood from tracking down the mediastinum.

In the present case report, the patient had been diagnosed as having partial deficiency of hemophilia B or Christmas disease/ factor IX deficiency.

Periodontal flap surgery and extraction of lower anteriors were performed in single visit to avoid excessive blood loss as well as repeated administration of fresh frozen plasma and resorbable sutures were used to avoid bleeding associated while removal.[10]

Antifibrinolytic agents such as tranexamic acid and aminocaproic acid were used in conjunction with clotting factor replacement therapy. These drugs prevent post-operative bleeding by inhibiting the activation of plasminogen to plasmin, which works to degrade fibrin clots.[11] The patient described in the present case report was provided with tranexamic acid mouthwash.

Aspirin and nonsteroidal anti-inflammatory drugs, which affect platelet function, should be avoided for pain management in patients with hemophilia. Acetaminophen or combination drugs may be a suitable alternative. Hence, paracetamol was prescribed to the patient for pain management.

Conclusion

Invasive dental care procedures for patients with moderate-to-severe hemophilia ideally should be conducted in a specialized setting. The decisions should be made by the dentist, in cooperation with the patient's hematologist, based on the planned dental procedure and the severity of the patient's disease. In general, laboratory tests, proper treatment planning with regard to controlling bleeding, and careful manipulation of tissues during the surgical procedure are significant in successful management of dental patients with hemophilia.

References

1. Srivastava A, Brewer AK. Guidelines for the management of hemophilia. Hemophilia 2013;19:e1-47.
2. Brewer AK, Roebuck EM, Donachie M, Hazard A, Gordon K, Fung D, *et al*. The dental management of adult patients with haemophilia and other congenital bleeding disorders. Haemophilia 2003;9:1-5.
3. Anderson J, Brewer A. Guidance on the dental management of patients with haemophilia and congenital bleeding disorders. Br Dent J 2013;215:497-504.
4. Hampton KK, Preston FE. Bleeding disorders, thrombosis, and anticoagulation. BMJ 1997;314:1026-9.
5. Gupta A, Epstein JB, Cabay RJ. Bleeding disorders of importance in dental care and related patient management. J Can Dent Assoc 2007;73:77-83.
6. Brewer A, Correa ME. Guidelines for dental treatment of patients with inherited bleeding disorders. In: World Federation of Hemophilia; 2006. p. 1-8.
7. Mannucci PM, Franchini M. Present and future challenges in the treatment of haemophilia: A clinician's perspective. Blood Transfus 2013;11:s77-s81.
8. Franchini M, Lippi G. Prothrombin complex concentrates: An update. Blood Transfus 2010;8:149-54.
9. Farrkh A, Garrison E, Clossman J. Dental surgical management of the patient with hemophilia. Gen Dent 2016;64:14-7.
10. Abed H, Ainousa A. Dental management of patients with inherited bleeding disorders: A multidisciplinary approach. Gen Dent 2017;65:56-60.
11. Estcourt LJ, Desborough M, Brunskill SJ, Doree C, Hopewell S, Murphy MF, *et al*. Antifibrinolytics (lysine analogues) for the prevention of bleeding in people with haematological disorders. Cochrane Database Syst Rev 2016;3:1-55.