CASE REPORT

Omnispective Dental Treatment Modalities in a Hemophiliac Child- A Case report

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Abstract

Hemophilia is the most common inherited bleeding disorder. It results from the deficiency of factor VIII (Hemophilia A) or factor IX (Hemophilia B). Hemophilic patients should be considered as ‘special’ patients. Since bleeding after dental treatment may cause severe or even fatal complications, people with hemophilia or congenital bleeding tendencies are a priority group for dental and oral preventive health care. Prompt treatment of bleeding episodes with hemophilia is essential to prevent long-term complications. Maintaining a healthy mouth and preventing dental problems is thus of great importance, not only for quality of life and nutrition but also to avoid the dangers of surgery. The present case report endows a comprehensive approach towards management strategy of a hemophilic Patient dictating extraction along with a preventive protocol for such patients.


Keywords- Haemophilia, Hemostasis, Extraction, Kidznteenz Pediatric Pentagon

Introduction

Haemophilia A, a X-linked recessive bleeding disorder, is the most common type of inherited bleeding disorder affecting approximately 1 in 10000 persons.1 Although transmitted as a sex-linked disorder largely affecting males, it has been shown that 25 per cent of all cases of haemophilia A, arise by spontaneous mutation.2 This disorder is attributable to decreased blood levels of properly functioning procoagulant Factor VIII. The severity of the disease depends on the level of circulating clotting Factor VIII and is characterized by prolonged clotting time and partial thromboplasin time. Whereas, the platelet count, platelet function tests and bleeding time are normal.3

The clinical presentation of the disease depends on the circulating levels of Factor VIII and is categorized as mild, moderate, and severe.

Table 1 shows the presentation of the disease clinically depending on the severity. Patients with haemophilia A often give a history of bruising,
joint swelling, and unusual bleeding associated with minor trauma or surgical procedures.  

Hemophilic patients should be considered as special patients. There are no contraindications to general dental treatment for hemophiliacs, as they generally do not involve bleeding. But surgical procedures involving local and general anesthesia must be carried out with caution. Such patients should always be managed in the setting of specialized units with appropriate clinical expertise. Novel approaches in the management of hemophilia have enabled many hemophilic patients to receive surgical dental procedures in an outpatient dental care on a routine basis. The purpose of this case report is to provide management strategies for a severely hemophilic patient requiring extraction and restorative treatment of teeth.

Table 1: Relation of Factor VIII concentration and severity of bleeding

<table>
<thead>
<tr>
<th>Factor VIII coagulant activity</th>
<th>Clinical features</th>
</tr>
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<tbody>
<tr>
<td>Less than 1% (severe)</td>
<td>Spontaneous bleeding into muscles and joints. Severe bleeding after trauma. Skeletal deformity.</td>
</tr>
<tr>
<td>1-5% (moderate)</td>
<td>Occasional spontaneous bleeding. Severe bleeding after mild trauma.</td>
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<tr>
<td>5-25% (mild)</td>
<td>Severe bleeding after major trauma.</td>
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<tr>
<td>25-30%</td>
<td>Tendency to bleed after major trauma.</td>
</tr>
</tbody>
</table>

Case report

A 6 year old male child reported to the Department of Pedodontics and Preventive Dentistry, Sardar Patel Post Graduate Institute of Dental and Medical Sciences, Lucknow who was referred by Sanjay Gandhi Post Graduate Institute, Lucknow.

Patient accounted for the chief complaint of mobility in relation to lower front teeth region and bleeding from the adjacent area (Figure 1).

The medical history was conclusive of Hemophilia A with factor VIII deficiency. The disease was confirmed on the basis of hematologic reports for a history of bruising and Injury for which he had been admitted at Sanjay Gandhi Post Graduate Institute, Lucknow, earlier. Thereafter few months later, he reported back with the complaint of mobility in his lower front teeth as well as persistent bleeding from the same area for which he was referred to our department. Additionally, the patient accounted for difficulty in speech as well as mastication due to the same. Family history yielded no relevant details. There was no other significant history of any medical disorder or exposure to anesthesia or surgical intervention.

On clinical examination, following findings were observed: grade II mobility- 71 & 81 and dental caries- 75, 85. Based on the clinical findings, the treatment plan was designed. Considering the age of the patient, complexity of the treatment, and compiliation in the coagulation, it was decided to carry out the treatment under careful supervision. The consent from medical, hematological and anaesthetical care centers was obtained before the treatment. The patient was scheduled for exodontia procedure, restorative treatment and put under chlorhexidine mouthwash for a week, to enhance the oral hygiene prior to the procedure.

Pre operative investigation values were as follows: hemoglobin - 12.5 gm%, prothrombin time - 13 seconds and activated partial thromboplastin time (aPTT) - 39.88 seconds. Factor VIII concentration was found to be 1% of...
normal activity. Based on the discussion with the hematologist, it was decided to go for the factor VIII transfusion, in which 200 Units as a preoperative loading dose by slow infusion, half an hour prior to treatment and 200 Units as a maintenance dose 12 hours postoperative was done. Transfusion for factor VIII was carried out in Sanjay Gandhi Post Graduate Institute, Lucknow by slow intravenous infusion.

The presence of mobility which further interfered with speech and mastication made us [http://www.jisppd.com/viewimage.asp?img=IndianSocPedodPrevDent_2011_29_1_74_79954_f6.jpg](http://www.jisppd.com/viewimage.asp?img=IndianSocPedodPrevDent_2011_29_1_74_79954_f6.jpg) to opt for extraction. Before the commencement of extraction of the required tooth, topical anesthetic spray as well as local anesthetic infiltration was done to avoid any pain or discomfort during the procedure (Figure 2). The tooth was extracted atraumatically (Figure 3).

Care was taken to remove the granulation tissue underneath the socket, which may prolong the bleeding. After extraction, the sockets were irrigated with betadine thoroughly and local pressure was applied (Figure 4). As the bleeding was continuous, (Figure 5) the socket was packed with absorbable gelatin sponge (surgispon™) for further hemostasis (Figure 6). During the entire procedure, complete isolation was made by the saliva ejector by resting it on a moist condensed sterile cotton roll in the floor of the mouth carefully.

Furthermore the patient was counseled and given postoperative instructions:
- Soft diet for 24 hours;
- Salt-water mouthwashes (1 teaspoon of salt In a glass of warm water) should be used Four times a day starting the day after the Extraction and continued for 7 days
- Avoid using straw, metal utensils or any sharp objects.
- No strenuous activities for 24 hours.

The patient was reviewed after 48 hours with no evidence of bleeding from the extraction site (figure 7). Subsequently, the patient was kept on recall visits. Restoration of carious teeth by Glass ionomer cement and topical fluoride application was done. The patient is still under follow up for any dental concerns or complications.
Discussion

Amongst the various disorders of hemostasis the most well known and common is haemophilia A. It is caused by a deficiency of antihemophilic globulin (AHG or Factor VII) in the plasma. It is inherited as a sex-linked recessive character, appearing only in males and transmitted to them by clinically normal female carriers. The defective gene on the X chromosome causes a deficiency of Factor VIII, normal plasma contains one unit of a factor VIII per ml. or 100% level. Deficiency of factor is expressed as percentage (0.25 unit/ml=25%).6 Hemophilia is considered severe when plasma activity of factor VII 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.6 The patient reported to our department was diagnosed as severe hemophilic since his factor VIII level was less than 1%.

Since hemophilia is a disorder of hemostasis normal hemostatic mechanism needs to be comprehended appropriately.

Normal hemostasis

Normal hemostasis is regulated by a series of enzymatic reactions collectively referred to as the coagulation cascade. The coagulation cascade involves a number of proteins and enzymes, each having a critical role in hemostasis.7

Older explanations of hemostasis described 2 separate pathways for initiation of coagulation—intrinsic and extrinsic—which merged to a common pathway. However, contemporary research suggests that hemostasis is primarily initiated with the release of tissue factor (part of the extrinsic pathway) following vascular injury. Tissue factor then complexes with factor VII, with subsequent conversion of factor VII to activated factor VII (factor VIIa).8 This complex acts locally at the site of injury to activate other factors such as factors X and IX, resulting in the generation of a thrombin "burst." This small amount of thrombin activates platelets and other factors in the intrinsic pathway, including factors VIII and IX. Coagulation then becomes dependent on the actions of factor VIII and IX for additional thrombin generation.9 With sufficient thrombin and other coagulation factors, fibrinogen is converted to fibrin, eventually forming a stable fibrin clot at the site of injury.

Disorders of hemostasis can manifest either as excessive bleeding or excessive blood coagulation. Excessive bleeding can occur when proteins or other components critical to coagulation are functionally absent due to either qualitative or quantitative deficiencies.10

As the age increases, the child's physical activity naturally increases, which results in more exposure to trauma, especially in hemophiliacs, as stated by Scully and Cawson.11 The case reported to our department was male with no family history of hemophilia, this can be well explained by that Hemophilia generally affects males on the maternal side. In hemophiliacs, family history of bleeding is commonly obtained. However, both FVIII and FIX genes are prone to new mutation and as many as one-third of all patients may not have a family history of these disorders.12

In the evaluation and treatment of hemophiliacs, the psychosocial aspect should never be underestimated. As there are some reports that the emotional disturbance is a contributing cause of spontaneous hemorrhage in hemophiliacs, the dentist must be alert to the emotional problems of the young hemophiliac as well as to the over-concern of the mother.2

Preoperative management:

Historically, international guidelines for oral surgery recommended the administration of clotting factor concentrates both before and after surgery. Doses are calculated to increase the level of factor VIII or IX to 50-100 IU/dL for a period of at least seven days.13 Ublansky et al. (1992) advised an increase of factor VIII or IX to 50% for
either regional or infiltration local anesthesia. At present, replacement of missing factor is commonly achieved with porcine Factor VIII or recombinant Factor VIII. One unit of Factor VIII concentrate per kilogram of body weight elevates the Factor VIII level by 2%. The amount of factor VIII to be infused was discussed and decided with the hematologist.

Anesthetic and operative procedure

The mandibular block should be avoided, because this form of injection may cause hemorrhage into the lateral pharyngeal spaces, where it is difficult to apply controlling measures. Therefore, topical anesthesia administration followed by local anesthetic infiltration was conducted before extraction. Extraction of the required teeth was accomplished asatraumatically as possible. Careful postoperative mouth toilet was carried out in our patient to avoid postoperative bleeding or infection as suggested by Brewer A, Correa ME (2010). Post surgery, the local hemostatic measures recommended is Surgicel®, Oxidized cellulose, and fibrin glue. Tranexamic acid, collagen, cyanoacrylate, and fibrin glues can also be helpful. Local use of fibrin glue and swish and swallow rinse of tranexamic acid before and after the procedure is also suggested as a cost-effective alternative. Based on the previous discussions with hematologist, we opted for gel foam (surgisponTM) which was used in our patient as a local hemostatic measure for achieving hemostasis. Patient was kept on recall for postoperative evaluation.

Restoration and prevention

The prevention and restoration of dental problems is an essential component of oral care in a hemophiliac. A successful regimen will reduce the need for treatment and should reduce the number of emergency visits. Every restoration in a hemophiliac eliminates a potential extraction. The dentist need not compromise the standard of dental service because the patient is a hemophiliac. Evans and Aledort (1978) suggested that high-speed vacuum aspirators and saliva ejectors can cause hematomas. Trauma from the saliva ejector was minimized by resting it on a gauze swab in the floor of the mouth in our patient. The necessity for good oral care and prevention of dental disease for the hemophiliac cannot be overemphasized, since preventive dentistry for these children minimizes the need for the hazards of restorative treatment.

Dental prevention depends on a number of different factors. Some of these may not be available in developing countries but are included to demonstrate the ideal situation.

- Brushing twice daily with a fluoride toothpaste. The use of fluoride toothpaste depends on the fluoride concentration in the water supply as well as the use of additional fluoride supplements. It should not be used if fluoride supplements are taken or if the water supply has a fluoride content of 1 ppm or more.
- The toothbrush should have medium texture bristles because hard bristles can cause abrasion of the teeth and soft bristles are inadequate to remove plaque.
- Interdental cleaning aids, such as floss, tape, and interdental brushes, should be used to prevent the formation of dental caries and periodontal disease.
- Fluoride supplements may be used, but are not recommended if the water supply has a fluoride content of 1 ppm or more.
- Artificial sweeteners can be used as an alternative to sugars in food and drinks. Examples are aspartame, sorbitol, acesulfamate etc.
- Regular dental visits, usually every 6 months, will help identify problems early. Reinforce prevention and emphasize the importance of reducing the intake of food and drink containing high levels of sugar or acid.

Conclusion

The care of people with hemophilia often requires a multidisciplinary team to address different aspects of their problems. This concept of comprehensive care is recommended by the World Health Organization (WHO) and the World Federation of Hemophilia (WFH). It comprises Hematologist, Anesthetist, and the Pediatric dentist, with the ultimate focus on the child. So, keeping the
special child in the center, the "Kidzteenz Pediatric Pentagon" [Figure 8] can be drawn to represent the above conclusion. This will ensure the effective and efficient management of the special child with the bleeding disorder. Moreover, a pediatric dentist and physician should treat the child in a least traumatic modality consequently posing the minimum amount of risks.

![Kidzteenz Pediatric Pentagon Diagram]

Figure 8: Kidzteenz Pediatric Pentagon

References