Hemophilia comprises a group of hereditary disorders caused due to the deficiency of one or more clotting factors leading to prolonged clotting time and excessive bleeding tendencies. It is broadly divided into Hemophilia A, B and C which occur due to deficiency of factors VIII, IX, or XI (F VIII, F IX, F XI) respectively. Hemophilia A is an X linked recessive hereditary disorder and is the most common of the three, accounting for 80-85% of the cases. Understanding this complex entity is very important for a dentist to provide appropriate dental treatment and to avoid undesirable consequences. The aim of this article is to report a case of Hemophilia A with literature review highlighting the importance of restorative treatment in salvaging the teeth and preventing complications anticipated from the surgical procedures.

Keywords: Endodontic treatment, Factor VIII, Hemophilia A

INTRODUCTION

Hemophilia comprises a group of hereditary disorders due to the deficiency of one or more clotting factors leading to prolonged clotting time and excessive bleeding tendencies that may be fatal.1 It is broadly divided into Hemophilia A (Deficiency of F VIII), Hemophilia B or Christmas disease (Deficiency of F IX) and Hemophilia C or Rosenthal Syndrome (Deficiency of F XI). The aim of this article is to report a case of Hemophilia A with literature review highlighting the importance of restorative treatment in salvaging the teeth and preventing complications anticipated from the surgical procedures.

Hemophilia A, which occurs due to the deficiency of F VIII is the most common of the three, accounting for 80-85% of the cases. It is an X-linked recessive hereditary disorder characterized by a deficient or defective F VIII coagulant (factor VIII C or Anti-Hemophilic Globulin). It is ordinarily carried through females and affects males. The incidence of Hemophilia A is approximately 1 in every 10,000 persons. However, 30% cases are caused by new mutations and hence may not be associated with a family history.2-4

CASE REPORT

A 26-year-old male patient reported with severe, sharp and continuous pain in a decayed tooth in the lower left back tooth region since 2 days that increased on chewing food on the left side. His medical history was significant as he suffered from Hemophilia A; diagnosed when he was 2 years old and suffered from spontaneous bleeding from gums, epistaxis, and bleeding from right ear. He had less than 1% activity of F VIII and had subsequently received multiple transfusions (whole blood and F VIII) over the years. The patient’s 8-year-old nephew (sister’s son) also suffered from this disorder. His general physical examination revealed a limping gait and swelling of both knees (Figure 1). On examination, it was found that his left mandibular permanent first molar (36) was deeply carious with slight obliteration of the buccal vestibule opposing this tooth (Figure 2). A provisional diagnosis of acute periapical abscess was given and the investigations advised included an intra-oral periapical (IOPA) radiograph, complete hemogram, prothrombin time, activated partial thromboplastin time (APTT), F VIII assay, international normalized ratio (INR), enzyme linked immunosorbent assay (ELISA) for human immunodeficiency virus (HIV) and HsBAg (since he was at a higher risk of these infections due to multiple transfusions), random blood sugar and knee radiographs (both antero-posterior and lateral). The IOPA of tooth 36 revealed proximity of deep carious lesion to the pulp with widening of periodontal ligament space and discontinuity of lamina dura suggestive of acute periapical abscess (Figure 3). His complete hemogram picture was within normal limits except an increased clotting time. His
DISCUSSION

Hemophilia A is caused due to F VIII deficiency, the gene for which is located on the long arm of the X chromosome at Xq28. Hemophilia has been classified into three forms: Severe form where factor level is less than 1% of normal (<0.01 IU/mL), moderate form where factor level is 1-5% of normal (0.01-0.05 IU/mL) and mild form with factor level more than 5-40% of normal (>0.05-0.40 IU/mL).

Severe cases may manifest with massive intra-uterine hemorrhage leading to still birth. In toddlers, oral ulcerations and ecchymosis involving lips and tongue are common. Tendency towards easy bruising, massive hemorrhage after trauma or minor surgical procedures are commonly encountered. Bleeding into the joints can lead to hemarthrosis of the joints. However, hemarthrosis of temporomandibular joints is unusual. Tissue hemorrhage forms tumor like masses termed as “Pseudotumors of Hemophilia”. Complications in hemophiliacs include...
chronic hemophilic arthropathy, development of inhibitors against F VIII and most importantly transfusion-related infections such as HIV, Hepatitis B virus, Hepatitis C virus, Hepatitis A viruses.2,10

The dental management of patients with Hemophilia A depends on the severity of the condition (mild, moderate or severe) and the invasiveness of the planned dental procedure.11,12 Restorative treatment is of utmost importance as the advanced dental conditions and subsequent treatments are more complicated and risky. However, restorative as well as endodontic treatment should be carried out bearing some considerations in mind.

Pulpal pain can usually be controlled with a minor analgesic such as acetaminophen. The use of any non-steroidal anti-inflammatory drug must be discussed with the patient’s hematologist because of their effect on platelet aggregation. Penicillin is the first line of drug used to control dental infection in conjunction with metronidazole to give good coverage of both the aerobic and anaerobic bacteria present in the oral cavity. Erythromycin and clindamycin can be prescribed to patients who are allergic to penicillin.13

Local anesthetics may not be needed in initial restorative phase, thus reducing the potential for serious hemorrhage. Care must be practiced while using matrix bands, rubber dam or wooden wedges during restorative treatment to avoid injuries to adjacent tissues. A rubber dam should be used to prevent soft tissue lacerations. High-speed suction can injure the mucosa in the floor of the mouth and cause hematomata or ecchymosis.

Endodontic therapy is preferred over extraction whenever possible. Working length of the root canal should be calculated precisely to avoid over instrumentation. Although there are no restrictions with respect to the type of local anesthesia used, those with vasoconstrictors may provide additional local hemostasis. Buccal infiltration and intra-lingual injections are preferred techniques over nerve blocks. Intra-canal injection of local anesthetic solution containing adrenaline may be useful to minimize intra-canal bleeding. Surgical endodontics requires F VIII replacement up to 50-75%. The need for post-operative maintenance of factor levels should depend on the type of surgery and severity of hemophilia.14,15

CONCLUSION

Hemophilic patients form a priority group for dental and oral health care, since bleeding after dental treatment may cause severe or even fatal complications. Restorative and endodontic treatment can avoid invasive procedures at a future date. In the case reported by the authors, endodontic treatment helped the patient to save the tooth as well as minimize the unnecessary post extraction complication.

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