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Radicular Cyst Enucleation in a Moderately Affected Hemophilia A Patient: A Case Report

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Abstract:
Background: *Hemophilias are a group of blood disorders and subdivided into Hemophilia A and B subtypes. These disorders come with a deficiency of clotting factors. This deficiency results in no or delayed clotting mechanism. Therefore, it becomes difficult to perform surgical procedures causing uncontrolled bleeding and thus hemorrhage. So, it becomes important to follow a management protocol, during pre operative, operative and post operative stages.*
Case Presentation: *A 33 year old male patient reported to the Outpatient department of our institute with a chief complaint of swelling on the right side of lower jaw. He was known case of hemophilia A. On clinical and radiographic assessment, the swelling was found cystic and provisional diagnosis of radicular cyst was given. Since the patient was a known hemophiliac a management protocol was used for preoperative and post operative stages. This protocol was established after thorough literature search and discussion with hematologists.*
Conclusion: *Hemophilia is a bleeding disorder which can lead to serious complications and such patients when undergoing any surgical procedure require a planned protocol for pre as well as post operative periods.*
Keywords: *Hemophilia, Radicular cyst, Malmo Protocol, Oral Surgery.*

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I. Background

Hemophilias are a group of bleeding disorders which are known always X-linked. They are subdivided into Hemophilia A (HA) which is due to deficiency of clotting factor VIII. The other variant Hemophilia B (HB) , which is caused due to deficiency of Factor IX or Christmas Factor.[1]Deficiency of these factors result in insufficient production of thrombin in intrinsic pathway of coagulation cascade resulting in bleeding. HA accounts for 80-85% of cases and HB in 15-20% of cases.[1] An uncommon type, Hemophilia C (HC) is an autosomal recessive defect that results in deficiency of factors XI and is characterized by bleeding in mucous membrane.

Genes of factor VIII AND IX are located on X chromosome.[2] The disease primarily affects males, but female carriers may be symptomatic.[3] Daughters of affected males are obligate carriers. One third of HA is due to spontaneous mutations and affected patients have no family history. However, all cases due to Factor VIII inversions are inherited for a mother carrier. The risk that a mother of an affected male is a carrier of is about 80%.[4]Process of coagulation is elaborated in two parts: primary and secondary hemostasis.[2] Primary hemostasis is the procedure where aggregation of platelets at the injury site takes place. It is followed by formation of more mature fibrin clot which forms during secondary coagulation cascade.[5]During the secondary hemostasis, a series of reactions takes place, which eventually forms a stable fibrin clot.[4] Factor VIII and Factor IX are both part of the intrinsic pathway of the coagulation cascade and are necessary to convert Factor X to Factor Xa, the first step of the common pathway. A deficiency or defect in either Factor VIII or Factor IX decreases the activation Factor Xa, impairing subsequent reactions necessary to create fibrin clots.[2]
Based on the levels of circulating procoagulants, hemophilia is divided into three categories:[6]

- Mild – Circulating clotting factors are >5% but less than normal 40%
- Moderate – 1-5% of normal circulating clotting factors

- Severe - <1% of clotting factors.

Hemophilia patients depending on the levels of clotting factors develop signs and symptoms accordingly. Patients with severe form are diagnosed shortly after birth. These patients commonly present with large cepahlohematomas and may also bleed profusely after circumcision.[7] Whereas, patients with milder forms present at a later stage of life. Presenting symptoms usually include swelling in the weight bearing joints like knee, ankle and hip joint.[8]Other presentations include hematoma formation following immunizations, minor trauma followed by excessive bleeding.[7]

Hemorrhage from many sites in the oral cavity is a common finding in hemophilia, and gingival hemorrhage may be massive and prolonged.[9] Even the physiologic processes of tooth eruption and exfoliation may be attended with severe prolonged hemorrhage.Along with these oral cavity is a very common site for multiple routine oral hygiene procedures. But in cases of hemophilia it becomes imperative to follow precautious measures in order to prevent excessive bleeding.Therefore, we present a case of radicular cyst enucleation in a moderate HA patient. Moreover we will also elaborate on the pre operative measures taken to prevent hemorrhage.

II. Case Presentation

A 33 year male patient reported to the Department of Dentistry, Sardar Patel Medical College and Associated Group of Government Hospitals. He presented with a chief complaint of swelling on the right side of lower jaw since 3-4 months. On his medical evaluation it was found that he is a known HA patient. He takes Tab.Cartigen-Duofor pain and swelling in left knee. Another finding was that patient undergoes prophylactic factor replacement therapy every 1-2 months.

Oral cavity examination showed that the swelling was diffuse extra orally and was tender. While intraoral examination showed that there was a grossly decayed 46 and distally carious 45. Intra orally there was an obvious expansion of the buccal cortical plate obliterating the buccal vestibule indicating it to be a cystic lesion. The patient was prescribed with Cap.Clindamycin 300mg three times a day, with Tab. Ibugesic and paracetamol when required, alongwith Tab. Lactobacillus three times a day as a supportive medication.An Orthopantomogram was advised for further radiological evaluation.Radiologically, a unicystic well defined radiolucency present at the periapical region of 46 and 45. Therefore it was decided to enucleate the cyst along with the surgical extraction of 45 and 46.

The patient underwent necessary preoperative routine blood investigations. Along with these a panel of necessary clotting factor assay was ordered. This included Prothrombin Time (PT), Partial Thromboplastin Time activated, PT with International Normalized Ratio and Factor VIII essay was performed. On evaluation of the blood investigations it was established that the patient had 1% activity of Factor VIII which according to clinical classification made him a moderate hemophiliac. Other blood investigation parameters were within the normal range.

The prophylactic factor replacement therapy is intravenous injections of Factor VIII concentrate in order to prevent anticipated bleeding. Replacement therapy is useful in cases where the Factor levels are not maintained above 1.0 IU/dl, especially in preoperative cases.

There are few dosing schedules to calculate the amount of factor required for preoperative prophylaxis. These include:[10]

- ***Malmö* Protocol**- 25-40 IU/kg per dose administered three times a week for those with hemophilia A, and twice a week for those with HB.
- ***Utrecht*Protocol**- 15-30 IU/kg per dose administered three times a week for those with HA, and twice a week for those with HB.

There are various other protocols for prophylaxis, but a most favorable regimen is yet to be defined. Therefore using the Malmö protocol, along with the consideration of the complexity of procedure, patient was given 2800 IU of 1 unit Factor VIII through intravenous route 12 hourly one day before surgery. Also 2800 IU of Factor VIII concentrate was given one hour prior to the surgery.

The patient was operated under local anesthesia. The solution used for local anesthesia was 2% lignocaine with adrenaline due to its vasoconstrictor effects. Tranexamic Acid 30-40 mg/kg intravenously was given during the operative period. After the induction of local anesthesia the mucoperiosteal flap was raised in relation with lower right quadrant of body of mandible region. Involved teeth 45 and 46 were extracted as atruamatically as possible and the cyst was enucleated, examined and sent for histopathology. The area was closed with 3-0 vicryl absorbable sutures.

During the post operative period the patient was kept under observation of a Hematologist. This period lasted for 7 days and went uneventful. The patient was given Tranexamic acid 30-40 mg/kg intravenously twice a day. Along with Factor VIII replacement therapy 12hourly.

The surgical specimen was preserved in 20% neutral buffered formalin. It was soft, grayish red in color with smooth borders and measures around 2.5 x 2.4 cm.

Histopathological evaluation showed presence of irregular non keratinized cystic epithelium which was of variable thickness (2-8 cell layered). The underlying stroma was densely packed with inflammatory cells including lymphocytes and plasma cells. Few slit like areas were present in the stroma suggestive of cholesterol clefts. There were areas of hemorrhage present. Endothelial lined blood vessels were noted. These features were suggestive of radicular cyst.

III. Discussion

Hemophilia patients, are very much prone to acquire dental diseases due to their inadequacy in performing routine oral hygiene procedures.[11] Moreover, they have higher susceptibility for oral bleeding slight vigorous cleaning action e.g., toothbrush trauma. Therefore, these patients require multiple visits to achieve definitive dental treatment. In the present case, the patient presented with radicular cyst which occurs as a sequelae of dental caries which in turn occur in the absence of proper oral hygiene measures.

The dental management of hemophilia patients depends on the clinical severity of this blood dyscrasia.[11] Therefore, with our case we did a thorough literature search to decide on the method of factor VIII replacement therapy to be used during pre and post-operative periods. Hence, based on the literary search we found Malmö protocol to be most suitable for factor VIII replacement. Avoiding robust dental procedures and achieving local as well as systemic heamostasis is of utmost importance.[11]Using intraoral-periapical radiographs can mutilate the oral mucosa producing bleeding therefore, we advised an orthopantomograph for radiological evaluation.Hemophilia patients frequently undergo factor replacement therapy and may be infected with HIV or Hepatitis B.[11] Therefore a complete viral marker panel was ordered for our case which included Anti-HIV, HBsAg and HCV which were non-reactive.

It is very much advisable to be very cautious during the operative procedure to avoid any iatrogenic injury to the oral mucosa.[12]We took utter care in our patient to extract 45 and 46 as atraumatically as possible, followed by thorough enucleation of the cyst to prevent recurrence or or any other future complication. Closure of the surgical site was done using absorable suture materia so as to avoid the need of suture removal. Analgesics that interfere with Platelet degradation should be avoided.[12] Hence, we preferred a selective pain management criteria, where Tab. Ibugesic and Paracetamol combination was given to the patient whenever needed in pre as well as post operative period.

During the literature search, we could not find any definitive guidelines for dental management of hemophilia patients. We came across a conglomerate of guidelines that state prevent of bleeding during any surgical procedure. The World Federation For Hemophilia, has given some guidelines to followed before any surgical procedure on Hemophiliacs.[13]

- Use of factor concentrates to cryoprecipitate or fresh frozen plasma for replacement therapy in patients with hemophilia.
- Patient's hematologist must be consulted before treatment initiation regarding the factor levels, factor replacements, type of surgery, and the need for systemic hemostatics.
- Replacement therapy comprising coagulation factor VIII is adviced.
- Surgery must be performed with caution to reduce trauma to soft tissues, and also measures to reduce intraoperative and postoperative hemorrhage must be undertaken.
- Proper closure of surgical site using absorbable sutures is indicated.
- Anti-fibrinolytic agents such as tranexamic acid (adult dose 1 g three times a day) and epsilon aminocaproic acid (50 mg/kg four times a day) are used at the start of the surgery and should be continued for a total of 7 days.
- Postoperative Factor replacement therapy is indicated after consultation with hematologist.

Hemophilic patients form a priority group for dental and oral health care, since bleeding after dental treatment may cause severe or even fatal complications. Moreover, maintenance of oral hygiene and prevention of dental diseases is of great significance to improve the quality of life and avoid the dangers of surgery. The prevention of large cystic lesions causing bone destruction can be achieved by regular dental checkups with prophylactic oral hygiene procedures. Proper X-ray evaluation before extraction and post extraction curettage of the periapical lesion is recommended.[14]

The close cooperation between hematologists, general physicians, oral physicians and surgeons, and general dentists will help to provide utmost care and appropriate treatment for patients with hemophilia. Genetic counseling is an important part of hemophilia care to help people with hemophilia, carriers, and their families make more informed choices about having children where there is a possibility of having a child with hemophilia or risk of having another affected child and the options available. Dentists can not only provide

complete oral care for hemophilia A patients but also contribute in the genetic counseling through wide range of tests for diagnostic and carrier detection, as well as individual counseling.

IV. Conclusion

In conclusion it is very important to prevent formation of large odontogenic pathologies in hemophilia patients and for this it is very important to perform thorough clinical oral examination in every hemophilia patient. Patients should be educated regarding oral hygiene procedures, without causing any oral mucosal mutilation leading to bleeding. Emphasis should be given on raiographic evaluation using extraoral radiographs. Another important point for consideration in these patients is to decide on an adequate factor replacement therapy protocol which will make any surgical procedure on these patients easier.

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