

ABSTRACT

Hemophilia A is a bleeding disorder caused by a congenital deficiency in Factor VIII, a blood-clotting protein which plays an essential role in the coagulation cascade. It is classified as mild, moderate, or severe based on the level of coagulation factor present in circulation, with severe measuring as less than 1% factor activity. Therefore, these patients are at significantly increased risk for bleeding. In dentistry, this is a major concern as bleeding can be triggered by physiological events such as tooth eruption, exfoliation, and mobile tooth roots, treatment of dental caries and periodontal diseases, invasive procedures such as extractions, suturing, fixed and removable orthodontic appliances, endodontic therapies, and administration of local anesthetic via inferior alveolar nerve blocks or lingual infiltration. This case report details management of facial cellulitis in a 4 year 1-month-old male with severe hemophilia A.

INTRODUCTION

Hemophilia is a bleeding disorder caused by a congenital deficiency in clotting factor. There are several types, including hemophilia A, a Factor VIII deficiency, and hemophilia B, a Factor IX deficiency. As both coagulation factors are part of the intrinsic pathway of the coagulation cascade, these deficiencies result in dysfunctional clot formation. Hemophilia is inherited as an X-linked recessive trait, and therefore predominantly affects males.

Hemophilia is classified based on severity, which is related to the level or activity of coagulation factor present in circulation. The normal range of coagulation factor level in circulation is 0.5-1.5 IU/mL. In mild hemophilia, this falls between 5% and 40% of normal levels, between 1% and 5% of normal levels in moderate hemophilia, and less than 1% of normal levels in severe hemophilia. Diagnosis of hemophilia A is confirmed by prolonged partial thromboplastin time.

In dentistry, bleeding disorders are of particular concern, as bleeding can be triggered by physiological events such as tooth eruption, mobile tooth roots, and tooth exfoliation, treatment of dental caries and the periodontium, and invasive procedures like tooth extraction, administration of local anesthesia, specifically inferior alveolar nerve block or lingual infiltration, fixed or removable orthodontic appliances, and endodontic therapies. There are several complications associated with hemophilia A, including increased bleeding risk secondary to prolonged bleeding time from clot instability, bleeding complications ranging from simple oozing at an extraction site to widespread intraoral hematomas and life-threatening hemorrhages or airway obstructions, and development of neutralizing IgG antibodies (inhibitors) to Factor VIII.

Due to the serious nature of the complications associated with bleeding disorders in a dental care setting, medical consultation with the patient's hematologist is essential to identify the type and severity of the bleeding disorder, the medications used, and whether pre-treatment with factor concentrate, nasal desmopressin, or antifibrinolytic agents is required. This case report details the dental management of a pediatric dental patient with severe hemophilia A who presented with a chief complaint of facial cellulitis secondary to an odontogenic infection.

Patient Introduction

- 4 year 1-month-old male presents to Hurley Medical Center Emergency department with a chief complaint of left facial swelling and abscess present in the upper left quadrant

History of Present Illness

- Swelling was first noticed 1 day prior
- Pain began at the same time and is constant and nocturnal in nature
- No history of fever, trauma, or pus drainage from the area

Medical History:

- Severe hemophilia A
- Eczema
- Seasonal allergies

Medications:

- Hemlibra
- Amicar
- Advate
- Triamcinolone cream
- Claritin

Allergies:

- Adhesive tape- silicones
- Strawberries
- Milk
- Aspirin and NSAIDs contraindicated due to bleeding disorder

Surgical History:

- Circumcision with no complications

Social History:

- Lives with mother, father, and no pets
- Passive exposure to tobacco smoking (father smokes outside the home)
- No exposure to alcohol or recreational drugs

Family History:

- Mother: mild hemophilia A, asthma, mental illness
- Maternal grandfather: severe hemophilia A
- Maternal grandmother: cancer, diabetes
- Father has no pertinent medical history

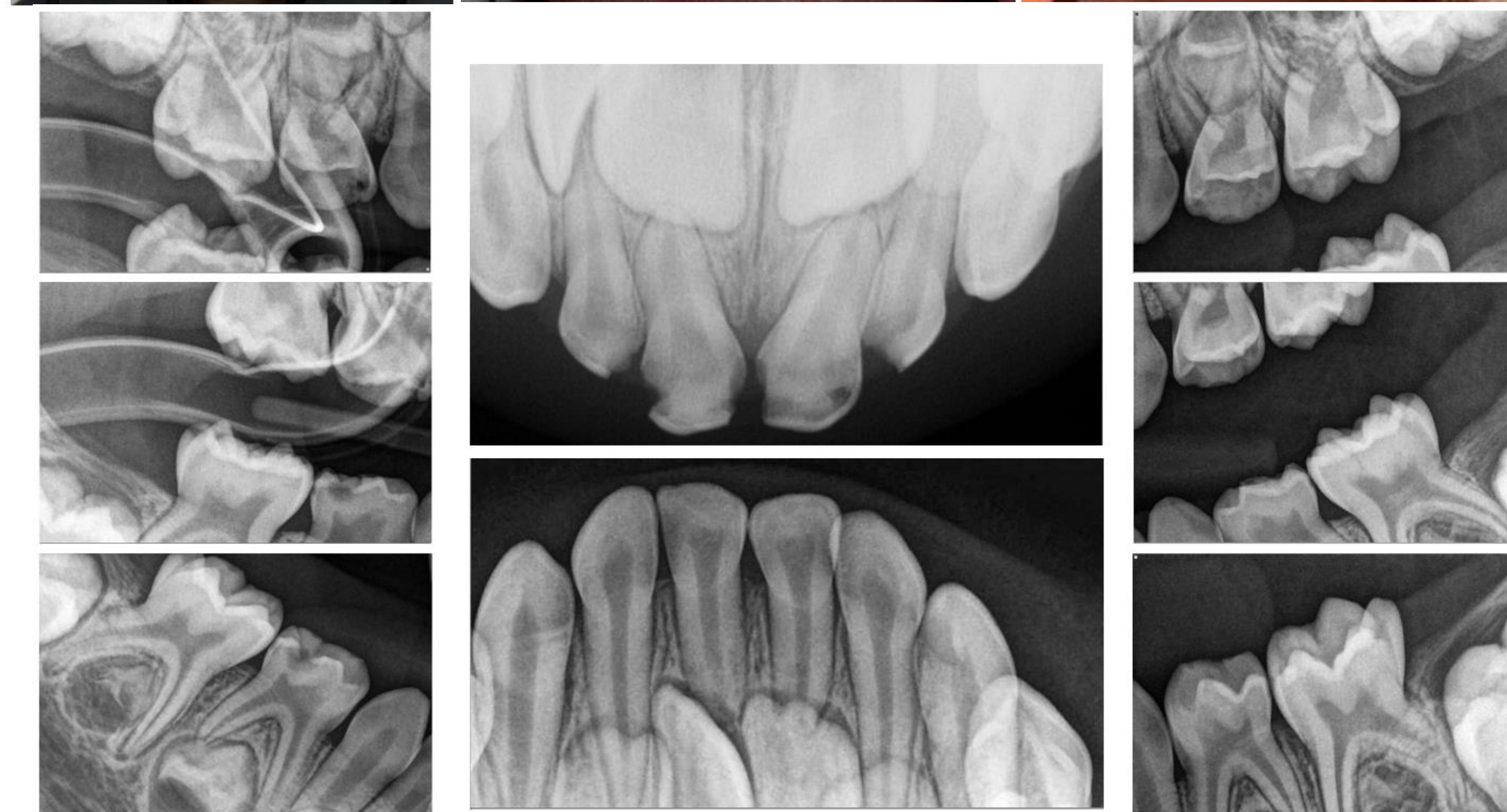
Clinical and Radiographic Findings:

- Caries on #A-MO, #B-MODB, #D-MFL, #E-MFLD, #F-MIFL, #G-MIFL, #I-B, #J-O, #L-D, #S-DOL, #T-O
- Fractures on #O-I enamel, #P-IFL enamel-dentin
- Deep, plaque retentive grooves #K-OB
- Hypoplasia #B-B, #I-B
- Facial decalcification #C-F, #H-F

Diagnosis:

- Facial cellulitis secondary to odontogenic infection

CASE REPORT



Treatment Plan:

- Prescribed oral Augmentin to address facial cellulitis
- Medical consultation completed with patient's hematology team, which indicated that invasive procedures must be completed under general anesthesia with pre- and post-procedural interventions to minimize bleeding risk
- Patient referred for full mouth dental rehabilitation under general anesthesia due to extensive and invasive treatment needs and complex medical history

Treatment Rendered:

2 BW, 6 PA, recall exam, dental prophylaxis, topical fluoride varnish; EXTs #D, #E, #F, #G; SSCs #A, #B, #I, #J, #L, #S, #T; IPTs #B and #S; sealant on #K-OB, and polish #C-F, #H-F, #P-IFL

HEMOSTASIS PLAN

Coordination with patient's hematology team was a key component to safe management of the patient's dental care. Medical consultation rendered the following hemostasis plan for the patient:

- The smallest gauge needle should be used for local injections and patient should not receive nerve blocks, mandibular blocks, or deep injections unless it is critical to his well-being
- Procedures requiring deep injections or considered very invasive may need to be completed under general anesthesia to minimize risk of bleeding
- No SBE prophylaxis indicated
- Amicar 50 mg/kg (max dose 2 g) should be taken the night prior to procedure continuing every 6 hours for up to 7 days
- NSAIDs and aspirin contraindicated; acetaminophen or narcotics only for pain management
- Patient will need an infusion of recombinant factor VIII prior to his procedure
- Patient will have PIV left in place and mother will be instructed how to infuse additional doses of recombinant factor VIII as needed; please advise mother as to expected amounts of post-operative bleeding
- Planned admission is not recommended unless patient has more bleeding than expected in the perioperative or postoperative phase
- Intubation method is up to the discretion of the surgical team as recombinant factor VIII given should increase patient's factor VIII activity level to normal range

DISCUSSION

Patients with hemophilia often require restorative treatment in a tertiary care center with a multidisciplinary approach. Children with hemophilia should be considered at high risk for dental caries, and the focus should be on preventing initiation and progression of caries. Extensive counseling about oral health should start when primary teeth begin to erupt, including diet, oral hygiene, the need for regular follow-up, and exposure to fluoride.

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