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PRE-OPERATIVE CONSIDERATIONS IN DENTAL MANAGEMENT OF AN 8-YEAR OLD FEMALE PATIENT WITH FANCONI ANEMIA

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ABSTRACT

Oral surgery in patients with bleeding disorders is associated with a high risk of bleeding during and after surgery. This article is aimed to present the case of an eight-year-old girl suffering from severe Fanconi anemia with pancytopenia who underwent a dental extraction.

Key Words: Fanconi anemia; extraction; bleeding

INTRODUCTION

Fanconi anemia (FA) is a rare autosomal recessive disorder characterized by congenital malformations, progressive bone marrow hypoplasia, and a high risk of malignancies. The existing literature describes occurrence frequency of FA of approximately 1:360,000[1] with a female-to-male ratio of 1:2.[2] In 90% of the cases, it is caused by a mutation in 3 genes: FNCA, FANCC, and FANCA. All the genes implicated in the development of FA DNA are involved in the crosslink repair pathway. Thus, patients with FA exhibit impaired capacity to repair the DNA

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crosslinks.[3] Fanconi anemia is named after the Swiss pediatrician, Guido Fanconi, who first described this disorder in 1927.[4] The diagnosis is usually made between the age of 5 and 10 years[5] and the life expectancy is 25 years, and may attain between 30 and 40 years of age.[6] Whereas, one third of the affected patients do not have any obvious abnormalities, in the majority of cases, physical signs, such as abnormal skin pigmentation, short stature, thumb and radial anomalies, structural renal defects, microcephaly, and delayed development are recognizable at birth and during early childhood.

The most frequent oral manifestations are gingivitis, periodontitis, rotated teeth and dental agenesis.[7] The high prevalence of periodontal diseases and gingivitis may be related to frequent immune system deficiencies, anemia and leukopenia. However, patients affected by FA often display deficits in their oral hygiene as well.[8]

Many FA patients experience bone marrow failure, which is generally characterized by an increasing pancytopenia, often initially with thrombocytopenia or leukopenia. In general, bone marrow transplantation is currently the most effective treatment to cure severe aplastic anemia or acute myeloid leukemia and the myelodysplastic syndrome.[9] All patients designated for transplantation should be examined for presence of any active infection. Dentists should carefully exam the oral condition within the pre-transplant evaluation. All the necessary dental treatment should be carried out before transplantation.

The aim of this paper was to report a case of an 8-year-old patient with FA presenting multiple carious lesions, as well as to discuss the multidisciplinary approach in the management and the treatment of this condition.

CASE DESCRIPTION

A known case of Fanconi anemia of an eight-year-old female patient was referred by a pediatrician to the Department of Pedodontics and Preventive Dentistry, D Y Patil School of Dentistry, Navi Mumbai for a dental examination. Patient complained of occasional pain in the right and the left side of the lower jaw. The family history was significant as the patient's parents had a consanguineous marriage and were carriers for FA. The patient had one younger brother. He had no medical history of Fanconi anemia. The parents' also had multiple carious lesions but presented no history of FA. The prenatal, natal, and past medical histories were significant. The patient's mother had taken antibiotics on account of systemic infection and fever in her seventh month of pregnancy, and the child was born through a cesarean operation after 34 weeks of

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pregnancy. The patient had low birthweight (1800 gms body weight at birth). The patient had recurrent episodes of severe pneumonia after birth. There was an atrial septal defect (ASD), which closed naturally at the age of 1.5 years. The patient's bone marrow was affected by thrombocytopenia with low hemoglobin (6g%).

The patient was conscious and fully cooperative with proper gait in general physical examination. The patient's height was less than normal for her chronological age. The skin was pigmented and hypertrichosis was noticed on her hands. (FIG)

On intraoral examination, multiple carious lesions were seen. Diagnosis of MIH was made, since all her permanent molars and incisors were hypomineralized. Oral hygiene maintenance was poor, leading to recurrent infections in the oral cavity.

Fig 1. Extra oral view showing short stature, abnormal skin pigmentation and hypertrichosis of the patient with known history of Fanconi Anemia.

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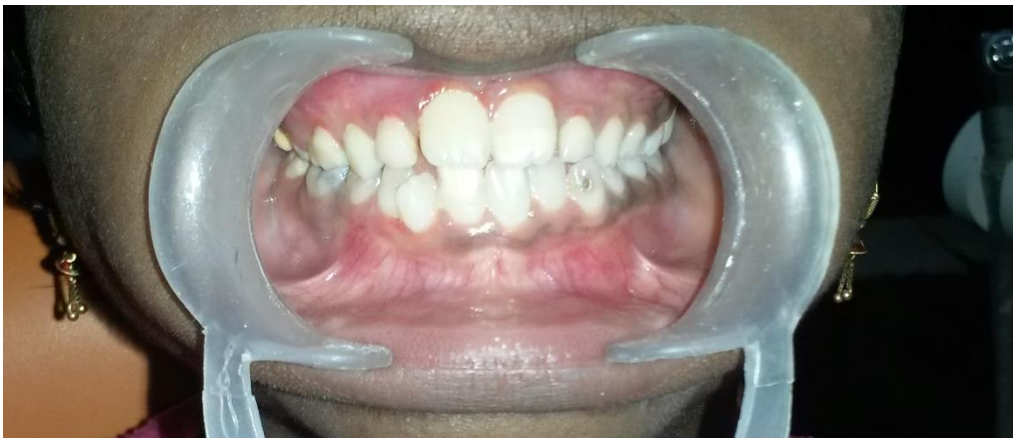

Fig 2. Intra oral frontal view

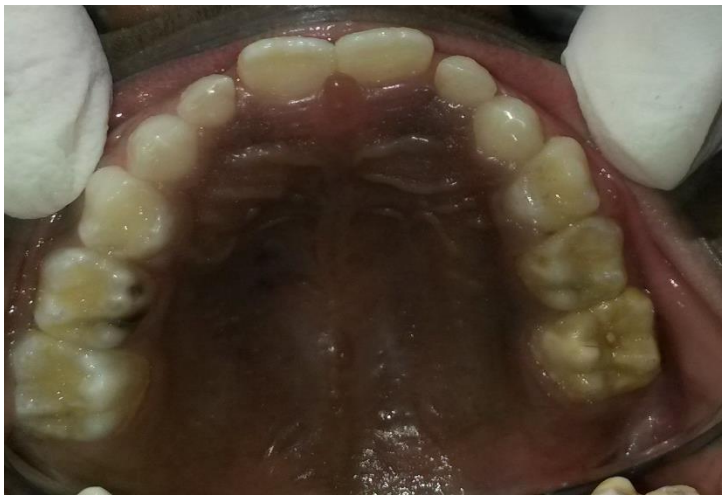

Fig 3. Hypoplastic maxillary molars with occlusal and palatal caries

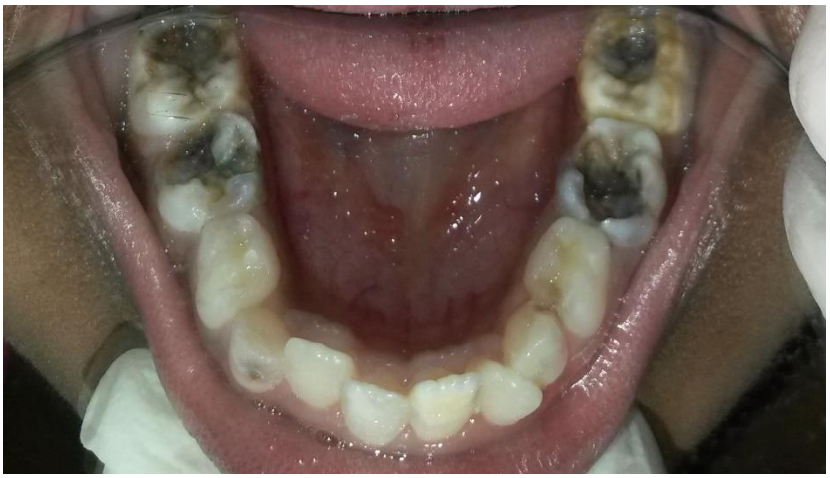

Fig 4. Grossly decayed 75,85,46,36

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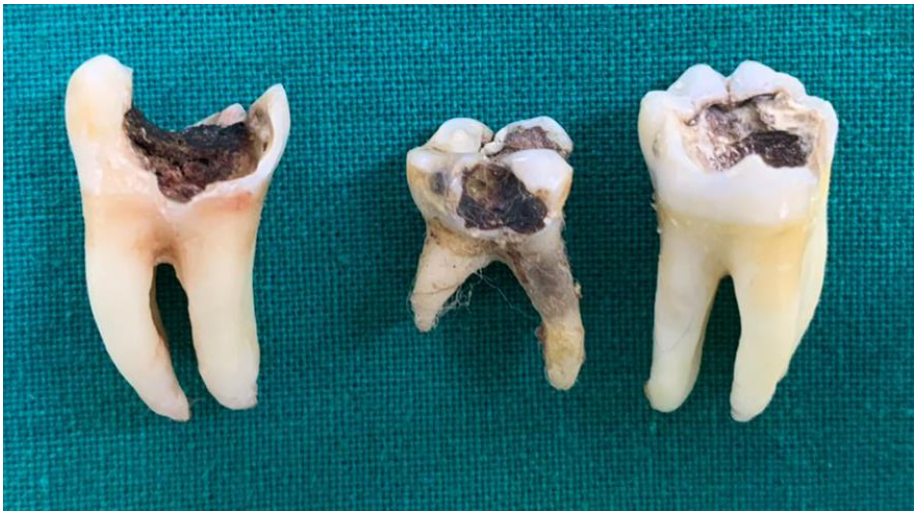
Fig 5. Extracted teeth



Treatment plan of restoration and extraction with most affected teeth was decided. However, she was chronically pancytopenic, with her platelet count less than 3×10^4 cells/lL and hemoglobin level at 6.0 g/dL. In addition, operative bleeding due to pancytopenia was expected while performing dental extractions. Due to these reasons, rendering dental treatment was extremely challenging. Her hematologist was consulted and it was decided that treatment would be carried out under general anesthesia. Over the next several weeks, many discussions were held in regard to the risk-benefit ratio of undergoing the necessary surgical procedures. Parents and patient understood that surgery in her case was a potentially life-threatening situation. Pre operative laboratory examination 7 days prior revealed pancytopenia. No abnormal findings were observed on the chest X-ray and electrocardiogram. Prior to the procedure, the patient's hematologist was again consulted for assistance in managing her hematologic condition. It was recommended that Tranexamic acid (TXA) to be used prophylactically during and after the procedure. The results of preoperative laboratory tests were reviewed and found to be consistent with her anemia. She received 1 U of single donor platelets pre-operatively to increase the platelet count. Other medications given were cefazolin, 1 g intravenously (IV) for prophylactic measure to prevent

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infection and dexamethasone, 8 mg IV. Due to the FA related severe thrombocytopenia, a platelet concentrate was necessary to raise the platelet count above 50,000/μL. A platelet count of 75,000/μL was achieved.

The treatment was performed under general anesthesia. Restorative treatment was done for 55 and pit and fissure sealants were applied on 16, 26. Following that, 75, 36, 85 and 46 were extracted. The sockets were immediately covered with tranexamic acid (TA). Two units of fresh frozen plasma (FFP) was advised post-operatively as a prophylactic measure on the advice of the hematologist. It was also advised to give cefazolin 1 g IV every 8 hours until the time of discharge, at which time cefadroxil 500 mg orally was prescribed to complete a 10-day course.

It was advised to keep the patient in ICU (Intensive Care Unit) post operatively for three days after the surgery to monitor the post-operative bleeding as well as other complications if arise any.

DISCUSSION

FA is a rare autosomal recessive heterogeneous disorder of hematopoietic stem cells causing pancytopenia and bone marrow hypoplasia with the depletion of all blood cell lines.[10] Bone marrow failure is the leading cause of death in FA, followed in frequency by leukemia and solid tumors. Poor oral hygiene and multiple caries before chemotherapy might increase the risk of developing systemic complications from dental infections, which may prove fatal.[11,12] Dental treatment is essential to minimize morbidity and improve the general condition of the patient.[13]

The main purpose of FA treatment is to increase the survival rate of the patients and establish a better quality of life. Patients with FA need a close follow-up with an interdisciplinary team, due to the complexity of the clinical manifestations. The oral manifestations of FA also require thorough investigation due the high chances of malignancy.

The predominant symptom of FA is the progressive bone marrow failure usually during early childhood with increasing pancytopenia, often associated with thrombocytopenia or leukopenia. Because of the important role of platelets in hemostasis in forming the initial plug in vessel injuries, severe thrombocytopenia presents a high risk for a postoperative hemorrhage after dental extraction in these patients.[14] In this case patient received 1 U of single donor platelets pre-operatively to increase the platelet count. It is a common practice to increase the platelet count to at least 50,000/μL. for procedures in critical regions, such as the brain or eyes and for major surgery, the platelet count should be raised to at least 100,000/μL.[15],[16]Henderson et al.[16]

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reported three treatment protocols for patients with thrombocytopenia undergoing dental extraction. Platelet transfusions were combined in one case with gelatin sponge for local hemostasis. In all cases, the surgical sites were closed primarily. One patient presented postoperative hemorrhage, which was managed systemically with multiple transfusions of packed red blood cells and platelets and topically with thrombin soaked gauze over the surgical sites as an adjunctive hemostatic measure. This is an indication that postoperative bleeding after oral surgery in patients exhibiting thrombocytopenia is difficult to predict, even though defined treatment protocols are used. Postoperative monitoring of these patients is required to prevent excessive hemorrhage.

In order to prevent secondary bleeding, resorbable suture material is advisable used to avoid suture removal. Non-steroidal, anti-inflammatory drugs and aspirin, which inhibit the platelet function, must be avoided for pain control. Paracetamol is a safe alternative to prevent postoperative pain. There are successful reports in the literature for patients affected by thrombocytopenia and Fanconi Anemia. Peisker et al.[14] described a case of present the case of an eight-year-old girl suffering from severe Fanconi anemia with pancytopenia who underwent a dental extraction. The hemostatic effect of local administration of tranexamic acid in combination with a primary suture was used to reduce the necessity of blood products and the risk of postoperative bleeding.

Patients with FA need a close follow-up with an interdisciplinary team, which should include a hematologist for the control of anemia; a pediatrician for managing systemic problems of children; an endocrinologist for the assessment and treatment of developmental disorders; an ENT specialist for the examination of nasopharynx, oropharynx, and larynx; a cardiologist for heart diseases; and an oncologist for the management of tumors. A pediatric dentist should also be a part of the multidisciplinary team. A highly planned preventive phase is important to prevent future radical treatment, which can be provided by a pediatric dentist. Treatment of acute dental infections should be carried out immediately and oral sources of infection should be eliminated, making sure that there are no foci of infection in order to ensure the overall health of the patient. Genetic testing should be done in all cases of suspected FA, siblings, parents, and close blood relatives. This will help in appropriate treatment planning and also in selecting a suitable donor for hematopoietic stem cell transplantation and to plan for genetic counseling.

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CONCLUSION

The topical administration of TA in combination with primary suture and perioperative transfusion of platelets seems to be effective in the prevention of postoperative hemorrhage in patients with severe thrombocytopenia undergoing dental extraction. It is essential to follow defined surgical protocols in the management of patients with bleeding disorders. A specialized dental service should perform these procedures in close cooperation with a hematology oncology treatment center. The high risk of developing an oral squamous cell carcinoma for patients affected by FA, requires a frequent oral examination by a dental practitioner.

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