

Management of a Maxillary Radicular Cyst in a Patient with Gilbert Syndrome: A Rare Case Report

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Abstract

Gilbert syndrome is a rare benign autosomal genetic disorder characterized by reduced activity of glucuronyl transferase, leading to decreased clearance of bilirubin and occasional mild jaundice in the absence of hepatic injury or hemolysis. Diagnosis involves clinical and laboratory investigations as part of differential diagnosis, with genetic analysis providing additional confirmation. Oral manifestations of Gilbert syndrome exist but often go unnoticed. Drugs metabolized by this enzyme need to be considered along with stress management in patients undergoing surgical procedures. This case report of a 24-year-old male diagnosed with a radicular cyst in the maxillary anterior teeth region, who incidentally exhibited Gilbert syndrome, also highlights perioperative management considerations for such patients.

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1. INTRODUCTION

Gilbert syndrome, first reported in 1901, is a genetic condition caused by a deficiency in the enzyme UGT1A1 (uridine diphosphoglucuronate glucuronosyl transferase)⁽¹⁾. This enzyme is essential for the conjugation of bilirubin, and its deficiency leads to elevated levels of unconjugated bilirubin in the blood. The prevalence of Gilbert syndrome in the general population ranges from 3% to 10%⁽²⁾.

The condition is typically asymptomatic but is mainly characterized by repeated episodes of jaundice, which can be aggravated by certain triggering factors⁽³⁾. Studies rarely report the prevalence of oral manifestations in Gilbert syndrome, such as yellow patches on the oral mucosa and altered taste sensation⁽⁴⁾. However, even if these oral symptoms are present, they are generally harmless. To achieve a definitive diagnosis of Gilbert syndrome, it is essential to assess bilirubin levels as part of a comprehensive liver function test.

Numerous anesthetic drugs that are metabolized in the liver through this pathway can accumulate, potentially resulting in adverse outcomes⁽⁵⁾. Optimal management of these patients necessitates the use of safe medications and careful avoidance of triggering factors during surgical procedures to ensure favorable outcomes.

This case report details a patient with a radicular cyst undergoing surgical intervention under general anesthesia, where an incidental discovery of Gilbert syndrome highlights the focus on oral manifestations and perioperative management of the condition.

2. CASE REPORT

A 24 year old male presented to our department of Oral and Maxillofacial Surgery complaining of pain in his upper front tooth region since 4-5 months. The patient's medical history was unremarkable, with no significant medical conditions, no relevant family medical history, and dental issues. The patient reported no known allergies to medications. The patient also gives history of occasional alcohol consumption and a history of smoking for the past year. On general examination, a generalized pale

yellowish hue of the skin was noted. On intraoral examination the patient's hard palate showed a deep hue of yellow, tapering off towards the soft palate. Additionally, a similar yellowish discoloration was noted on the floor of the mouth. The patient's teeth appeared normal, but there was noticeable paleness around both the free and attached gingiva, affecting all teeth uniformly. On local examination case was diagnosed as radicular cyst in relation to 21 region and diagnosis was confirmed with CBCT investigation. Therefore, surgical enucleation of the cyst with apicoectomy of tooth 21 under general anesthesia was planned. On routine blood investigation for the general anesthesia procedures, elevated unconjugated bilirubin levels (>2.1 mg/dl) were noted, but serum liver transferase levels and red blood cell count were surprisingly normal. Based on clinical and laboratory investigation patient was referred to Department of General Medicine Bapuji Hospital where further investigation was conducted, using ultrasound of the abdomen and pelvis, as well as urine examination, which revealed no abnormalities. These findings indicate no evidence of hepatic injury or hemolysis. Therefore, the patient was diagnosed with Gilbert syndrome. Following literature guidance, the patient underwent the procedure with specific precautions. Stress management protocols were implemented, and fasting was limited to 6 hours to avoid prolonged starvation. Pre-anesthetic medications such as midazolam and glycopyrrolate were administered, while long-acting opioids were avoided. Propofol was used as the inducing agent, followed by nasotracheal intubation. Maintenance with isoflurane instead of halothane gases was employed, and long-acting skeletal muscle relaxants like vecuronium were avoided. The surgical procedure was successfully performed, and the patient recovered without complications. There was no evidence of post-operative jaundice noted during the 1-month follow-up. **(Figure 1-3).**

3. DISCUSSION

Gilbert's syndrome, a mild unconjugated hyperbilirubinemia, is usually benign and arises from either increased bilirubin production or impaired conjugation⁽¹⁾. Distinguished from hemolysis by Powell in 1972, it was first described by Gilbert and Lereboullet in 1901⁽⁶⁾. Kornberg's 1942 study suggests its prevalence might be around 7% in large populations. The condition is more common in males and may follow an autosomal dominant inheritance pattern. Symptoms, if present, include nonspecific abdominal pain or lethargy, but many cases are asymptomatic. Mild jaundice is the primary sign, with unconjugated bilirubin levels typically below 6 mg per 100 ml, and other liver function tests remain normal⁽⁷⁾. The exact mechanism is

unclear, but studies show reduced hepatic bilirubin clearance, potentially due to defective conjugation and decreased activity of bilirubin UDP-glucuronyl transferase. Gilbert's syndrome does not affect life expectancy⁽⁸⁾. While phenobarbitone can reduce bilirubin levels, long-term treatment is generally unnecessary.

In our case study, the patient displayed a yellowish tint in the hard palate, floor of the mouth, gingiva with no abnormalities in teeth structure. The patient also had yellow discoloration of skin⁽⁴⁾. Proper diagnosis is essential, as Crigler-Najjar Syndrome can present similar symptoms to Gilbert's syndrome.

Patients with Gilbert's syndrome often miss oral symptoms. In dental procedures, patients with Gilbert's syndrome usually don't experience discomfort. However, some cases report jaundice after oral surgery⁽⁸⁾. Research suggests potential challenges with regional and general anesthesia due to liver dysfunction impacting drug metabolism, necessitating careful selection of anesthetic agents⁽⁵⁾.

Any stress can exacerbate symptoms of Gilbert's syndrome, such as fasting, surgery, infection, exercise, fatigue, alcohol intake, and menstruation. These stressors can lead to symptoms ranging from clinical jaundice to nausea, malaise, discomfort in the right hypochondrium, or even abdominal pain. Prolonged fasting can induce symptoms because fatty acids compete with unconjugated bilirubin in the liver. Postoperative jaundice following oral surgery has been reported, attributed to stress and reduced caloric intake⁽⁹⁾.

To prevent prolonged fasting, we prioritized this patient for early surgery scheduling. Alprazolam was prescribed the night before to alleviate pre-surgery stress, despite lower clearance rates in Gilbert's syndrome patients compared to controls⁽¹⁰⁾. Early administration of 5% dextrose on the morning of surgery aimed to prevent dehydration and hypoglycemia-induced stress. Propofol was chosen over thiopentone or ketamine due to its dual metabolism by the liver and kidneys, ensuring a safer administration. Thiopentone and ketamine can affect liver function based on dosage, complicating the diagnosis of postoperative jaundice. Atracurium was chosen due to its Hofmann degradation and ester hydrolysis properties, despite limited evidence in the literature of other muscle relaxants causing prolongation in Gilbert's syndrome. Isoflurane was considered safer for its minimal liver metabolism (less than 0.2%), which helps preserve liver blood flow⁽⁵⁾⁽¹⁰⁾. Postoperative pain relief included diclofenac sodium and pentazocine, while paracetamol and morphine were avoided due to concerns about potential toxicity in Gilbert's syndrome patients⁽¹¹⁾. Gilbert's syndrome, being the most common hereditary cause of elevated bilirubin,

allows for safe anesthesia administration if the implications of glucuronyl transferase deficiency on drug metabolism and excretion are well-managed.

4. CONCLUSION

Evaluating a patient with Gilbert's syndrome (GS) and a radicular cyst requires recognizing GS symptoms, such as intermittent jaundice triggered by factors like fasting and stress, and examining oral manifestations for effective management. Understanding GS's pathophysiology and triggers is essential for the safe administration of anesthesia.



Figure 1: Extra-oral Image



Figure 2: Intraoral palate showing yellow patches



Figure 3: Labial gingiva of maxilla showing paleness

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