Copper extraction: Dental consideration for Wilson’s disease – An uncommon case report

ABSTRACT

Wilson’s disease is a very rare and inherited autosomal recessive disease of copper metabolism. The cause of the disease is mutation of the Adenosine triphosphate 7B (ATP7B gene). The ATP7B gene is responsible for biliary excretion of copper and incorporation of copper into ceruloplasmin. The imbalance in the copper metabolism leads to copper toxicity which primarily involves the brain, liver, the kidney, and the skeletal system. Early diagnosis and intervention is needed to prevent the mortality and morbidity of the disease. In this article, we focus on the evaluation and dental management of patients with Wilson’s disease.

Keywords: Adenosine triphosphate 7B gene, copper metabolism, Wilson’s disease

INTRODUCTION

Wilson’s disease is a very rare and inherited metabolic disease with an incidence in India estimated at 30/1 million of population worldwide. The prevalence is higher in North Indian population. These diseases were first documented in 1912 by Sir S. A Kinnier Wilson.[1‑4] Dietary Copper is absorbed in the proximal small intestine, and it is taken into the liver where 90% of it binds into ceruloplasmin.[5,6] Ceruloplasmin is a serum glycoprotein synthesized and secreted by the liver. In Wilson’s diseases, the subsequent mutation of Adenosine triphosphate 7B (ATP7B) gene leads to abnormal accumulation of copper in hepatocytes due to failure of synthesis of ceruloplasmin. This leads to copper spillage into the various organs of the body.[6] As the diseases, progresses-free copper levels in the body may increase due to the reduction in binding of free serum copper into ceruloplasmin.[7,8] A low serum ceruloplasmin is the best and single laboratory clue for the diagnosis of Wilson’s diseases. The notable clinical feature is tremor, jaundice, dystarthish abnormal gait, abdominal distention, musculoskeletal symptoms, seizures, behavioral problems, dystonia, clumsiness, drooling of saliva, generalized weakness, altered sensorium, bleeding diathesis, dysphagia, chorea and poor vision, and scholastic performance.[9‑11] We present this rare case report and its preoperative evaluation and its dental consideration.

CASE REPORT

We are presenting a case of a 26-year-old female patient who reported to our department of oral and maxillofacial surgery for the extraction of upper right third molar. The patient was diagnosed with Wilson’s disease at a young age, and she had been undergoing treatment for the disease. On physical examination, she had symptoms of tremors and difficulty in walking. The patients magnetic resonance imaging revealed a giant panda appearance, which is T2 hypersensitivity in
bilateral, ventrolateral thalamus. The diagnosis of Wilson’s disease was confirmed by an increased level of ceruloplasmin. Liver function test (LFT) and ultrasound revealed no significant abnormality of the gastrointestinal tract. The patient’s status revealed asthenic habitus, dysarthria, and spastic ataxic gait with drooling of saliva.

Ophthalmologic evaluation reveals the presence of the characteristic Kayser-Fleischer ring which is a golden brown ring at the outer surface of cornea [Figures 1 and 2].

The patient’s laboratory results were as follows:
- **LFTs** reveals:
  - Total Bilirubin: 1.0 mg/dl; serum glutamic-oxaloacetic transaminase: 17 IU/L; serum glutamic-pyruvic transaminase: 24 IU/L; serum alkaline phosphatase: 57 IU/L; Albumin: 3.2 g/dl; Creatinine: 0.8 mg/dl; Hb: 11 g/dl, gamma-glutamyl transferase – 80 IU/L
- **Glucose** – 5.0; Total protein – 6.7 g/dl
- **Sodium** – 144 mmol/l, Potassium K – 3.5 mmol/l, DLC: 60,31,5 (neutrophils, lymphocytes, eosinophils); erythrocyte sedimentation rate: 14 mmos
- **Platelet**: 211,000 µl; BT: 3 min; computed tomography: 4 min; APTT: 32.

The medical examination reveals that the patient’s disease is moderate. The extraction was carried out under strict aseptic condition with a minimal dose of 1:200,000 of lignocaine with adrenaline. Hemostasis was obtained, and postoperative instructions are given. The patient reported back after 3 days, and the extraction socket was healing satisfactory.

**DISCUSSION**

The genetic mutation of ATP7B gene followed by impaired copper metabolism, due to low level of ceruloplasmin are the striking features of Wilson’s diseases. There is an increased Copper accumulation in the liver in patients with Wilson’s disease. This disturbs the metabolic function, protein synthesis, and drug metabolism that take place in the liver. Local anesthesia should be used cautiously at minimal dosage, as they are metabolized in the liver. Adrenaline should be used in very low concentration.[12,13] The patient of Wilson’s disease is highly susceptible to osteoporosis, and therefore, extraction should be done asatraumatically as possible without damaging the alveolar bone.[8,9] Patients with Wilson’s diseases are prone to increase in bleeding and clotting time due to impaired liver function. Therefore, before procedure, bleeding time and clotting time should be evaluated. Preoperative Vitamin K and fresh frozen plasma should be available chair side if necessary. A hemostatic agent should be used and suturing should be done to aid in healing. The use of postoperative drugs such as nonsteroidal anti-inflammatory drugs and metronidazole should be avoided as it increases hepatotoxicity, so low dose of paracetamol is advised.[13]

The patient who has an acute Wilson’s disease may show parkinsonism, neurological impairment, dementia, and athetosis.[14,15] We can see cases of rigidity and orthostatic hypotension in these patients. Special care should be taken to the patients to avoid aspiration of dental materials or water during the procedure.[16] The chair should be inclined slowly at the end of each procedure for reequilibration. As seen in Parkinson’s diseases, patients of Wilson’s disease also show excessive salivation, show increased muscular activity and tremor, This leads to problem during the administration of local anesthesia and while providing other treatment. Antimuscarinic anti-parkinsonism drugs reduce the salivation but can cause taste distortion, therefore, it might increase the risk of caries in these patients. Often patients have difficulty in maintaining their oral hygiene because of their physical disability, so frequent recall is necessary after dental treatment.[15,16] The dentist should minimize the use of copper containing material such as amalgam and medicines. If orthodontic treatment is carried on these patients, the use...
of fixed appliances should be avoided because it may lead to fast bone resorption.[17]

CONCLUSION

In the era of modern medicine, medically compromised patients tend to visit the dental office more frequently for their dental problems. Sound knowledge in the field of medicine is necessary for the management of medically compromised patients. Wilson's disease is very rare and uncommon disease. According to the reported medical literature till date and to best of our knowledge, this is the first reported case of Wilson's disease in the south Indian population and which was successfully treated for the tooth extraction without any complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Taly AB, Meenakshi-Sundaram S, Sinha S, Swamy HS, Arunodaya GR. Wilson disease: Description of 282 patients evaluated over 3 decades. Medicine (Baltimore) 2007;86:112-21.

2. Wilson SA. Progressive lenticular degeneration: A familial nervous disease associated with cirrhosis of the liver. Brain 1912;34:295-507.

3. Dastur DK, Manghani DK, Wadia NH. Wilson’s disease in India. I. Geographic, genetic, and clinical aspects in 16 families. Neurology 1968;18:21-31.

4. Compston A. Progressive lenticular degeneration: A familial nervous disease associated with cirrhosis of the liver, by S. A. Kinnier Wilson, (From the national hospital, and the laboratory of the national hospital, Queen Square, London) brain 1912: 34, 295-509. Brain 2009;132:1997-2001.

5. Chu NS, Hung TP. Geographic variations in Wilson’s disease. J Neurol Sci 1993;117:1-7.

6. Taly AB, Prashanth LK, Sinha S. Wilson’s disease: An Indian perspective. Neurol India 2009;57:528-40.

7. Roberts EA, Schilsky ML: American Association for Study of Liver Diseases (AASLD). Diagnosis and treatment of Wilson disease: An update. Hepatology 2008;47:2089-111.

8. Fox C, Lombart M, Emma L, editors. Gastroenterology. Spain: Elsevier Health Sciences; 2004. p. 132.

9. Brewer GJ. Wilson disease. In: Fauci AS editor. Harrison’s Principles of Internal Medicine. Vol. 2. New York: McGraw-Hill Medical Publishing Division; 2008.

10. Ghah WA. Wilson’s disease. In: Goldman L, Bennett JC, Gill D, Kokko G, editors. Cecil Textbook of Medicine. Vol. 1. India: W. B. Saunders Company, a Harcourt Publishers International Company; 2001. p. 1130-1.

11. Ala A, Walker AP, Ashkan K, Dooley JS, Schilsky ML. Wilson’s disease. Lancet 2007;369:397-408.

12. Little JW, Falace DA, Miller CS, Rhodus NL, editors. Liver disease. In: Dental Management of the Medically Compromised Patient. St. Louis: Mosby Elsevier; 2008. p. 152.

13. Greenwood M, Meechan JG. General medicine and surgery for dental practitioners. Part 5: Liver disease. Br Dent J 2003;195:71-3.

14. Lohe VK, Kadu RP, Degwekar SS, Bhowate RR, Wanjari AK, Dangore SB, et al. Dental considerations in the patient with Wilson’s disease. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2011;111:20-3.

15. Chapman RW, Collier JD, Hayes PC. Liver and biliary tract disease. In: Boon NA, Colledge NR, Walker BR, Hunter JA, editors. Davidson’s Principles and Practice of Medicine. Philadelphia: Churchill Livingstone; 2002. p. 871-2.

16. Ciarrocca KN, Greenberg MS, Garfunkel A. Neuromuscular diseases. In: Greenberg MS, Glick M, editor. Burket’s Oral Medicine Diagnosis and Treatment. India: BC Decker Inc.; 2003. p. 597-8.

17. Davidson R, Walters S. Basal ganglia disorders and related conditions. In: Rose L, Kaye D, editor. Internal Medicine for Dentistry. Philadelphia: The CV Mosby Company; 1990. p. 721-3.