



Case Report

Anaesthesia challenges in Wilson's disease with multisystem involvement

Pratika Pradeep Bhokare¹, Viral Girish Parekh^{2,*}

¹Bai Jerbai Wadia Hospital for Children, Mumbai, Maharashtra, India

²Dept. of Anaesthesia, Lokmanya Tilak Municipal General Hospital, Mumbai, Maharashtra, India



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ABSTRACT

Wilson's disease is a rare genetic disorder with a prevalence rate of 1:10000-1:30000.¹ It is a multi-system disorder with a defect in the copper transporter protein; thereby resulting in extortionate copper deposition in organs of the body; mainly liver, brain and cornea. Osteo-muscular involvement may also be seen, presenting as dystonia and muscle rigidity.² The case discussed below is that of a 13 year old patient, ASA III E, diagnosed with Wilson's disease 7 years back and now is posted for emergency tracheostomy under anaesthesia in view of chest rigidity, difficulty in breathing and repeated episodes of bronchospasm. The patient presented in late stage of the disorder with associated derangements of multiple systems of the body. This case report provides details of successful administration of Anaesthesia to a patient with Wilson's in late phase. The management protocol can be used as a guide in other surgeries of patients with this disorder, as precise titration of anaesthesia is needed.

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1. Introduction

Wilson's disease (hepatolenticular degeneration) is named after Dr. Samuel Wilson, a British neurologist. It is a rare genetic autosomal recessive disorder with a mutation in ATP7B Gene. This mutation leads to the paucity of the copper transporter protein (Ceruloplasmin). As a result, there is copper deposition in the organs of the body- mainly liver.³

Here, we present the successful management of a young patient with Wilson's disease with involvement of multiple systems of the body along with bronchospasms posted for emergency tracheostomy under Anaesthesia. This case can work as an aid in the administration of Anaesthesia in patients with this disease posted for other minor or major surgeries.

2. Case Report

A 13 year old patient diagnosed with Wilson's disease 7 years back, presented with complaints of gradually progressive generalized dystonia, dysarthria and severe muscle wasting. The patient had recent history of frequent episodes of severe bronchospasms and oro-mandibular muscle stiffness since 2 months for which he was posted for emergency tracheostomy. The patient took Penicillamine for 5 years and has been non-compliant since last 2 years. On general examination, the condition was guarded, the patient was vitally stable (Pulse -110/min, BP- 120/80mm Hg, SpO₂ – 98% on room air). On auscultation, the respiratory system revealed bilateral basal crepitations and wheeze; chest wall muscles had rigidity. The patient had pallor and Kayser-Fleischer rings in cornea along with bilateral upper and lower limb dystonia and atrophy. Patient was mentally retarded for his age. Since the patient had jaw stiffness, Inj. Botox was given 2 days prior to relax the jaw muscles and open the jaw. On airway evaluation, the mouth opening was 2.5 fingers with Mallampati Class II with restriction of neck flexion and extension. The patient was unable

* Corresponding author.

E-mail address: vgparekh2020@gmail.com (V. G. Parekh).

to close the mouth. The patient had raised liver enzymes (AST/ALT- 150/226IU/L) with decreased protein levels (Total protein: 4.8g%) and low serum calcium (7.8mg/dl). For anaesthetic management of this patient for the surgery, the difficult airway cart was kept ready. Standard monitors (ECG, NIBP, Pulse Oximeter, EtCO₂) were attached. Premedication was given with Inj. Glycopyrrolate 0.2mg (0.005mg/kg) and Inj. Midazolam 0.5mg. Fentanyl was avoided due to chest rigidity. Injection dexamethasone 8 mg (0.2mg/kg) and injection hydrocortisone 100 mg were given prophylactically. Induction was done with injection Propofol 1.5 mg/kg and without muscle relaxant. Patient was intubated with McCoy laryngoscope and anaesthesia was maintained on O₂, N₂O and Propofol aliquots. Salbutamol puffs were given through endotracheal tube. Injection Dexamethasone and Hydrocortisone, and Salbutamol puffs were given as the patient had multiple episodes of bronchospasm with wheezing in the last few days and an impending episode of the same had to be avoided perioperatively.

Patient was vitally stable and a saturation of 100% was maintained throughout the procedure and no event of bronchospasm or laryngospasm occurred. Post procedure, the position of the tracheostomy tube was confirmed and patient was ventilated through the same until spontaneous respiration resumed. Patient was put on T-piece post procedure with oxygen supply and monitored till he was fully awake.

3. Discussion

The clinical features⁴ of the Wilson's disease are associated with:

1. Liver injury- hepatitis, steatosis and cirrhosis.
2. Neurological – dysphagia, gait abnormality, dystonia, dysarthria and Parkinsonism.
3. Psychiatric – behavioral changes, mood disorders and depression.

Clinical manifestations of each patient of Wilson's disease is variable and anaesthesia care is to be tailored accordingly. The patient had multisystem involvement and care had to be taken to avoid further deterioration due to Anaesthesia. The patient had deranged liver function tests, causing hampered absorption, distribution and metabolism of anaesthetic drug; aggravating the already impaired hepatic function. Sedatives may exacerbate the neurological problems. The sensitivity to neuromuscular relaxants is increased in these patients in view of decreased muscle function which may be attributed to the disease process itself or as an adverse effect of prolonged Penicillamine use.⁵ Penicillamine is also known to cause hypersensitivity pneumonitis, myasthenia and membranous glomerulopathy.⁶ The hyper reactive

airway associated with intermittent bronchospasm, chest rigidity and oro-mandibular muscle stiffness pose difficulty in managing airway. In this case, patient was intubated due to hyper reactive airway and impending spasms. Supraglottic airway can also be used in patients of Wilson's disease without any airway influence. Corticosteroids and Salbutamol have been known to attenuate the perioperative bronchoconstriction.⁷ Action of skeletal muscle relaxant could get prolonged and thus, to be avoided in liver pathology depending on the procedure. The patient is to be restarted on medications for Wilson's disease as soon as possible.

4. Conclusion

Being a rare genetic disorder, information about anaesthetic management in a patient of Wilson's disease poster for surgery is not widely available. These patients may present with numerous complications. Challenges like difficult intubation altered drug metabolism, bronchospasm are usually seen in patients of this disease. The above case report will serve as a useful aid in the management of these patients.

5. Declaration of Patient Consent

The authors certify that they have obtained the appropriate consents for recording the data. The patient's guardians understand that their names will not appear or be published in any of the documents and complete anonymity will be maintained.

6. Source of Funding

None.

7. Conflict of Interest

There are no conflicts of interest.

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Author biography

Pratika Pradeep Bhokare Specialist Medical Officer

Viral Girish Parekh Assistant Professor

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