

Anaesthesia for a patient with Wilson's disease: A case report

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Abstract

Wilson's disease (WD), also known as hepatolenticular degeneration, is an autosomal recessive disorder and is due to reduction in the synthesis of copper transporter protein, Ceruloplasmin. Due to ceruloplasmin deficiency, there is failure of excretion of copper in bile and it accumulates in body tissues leading to major hepatic and neurological involvement. Hepatic involvement after excess copper deposition leads to chronic liver disease and cirrhosis, which may alter metabolism and excretion of anaesthetic agents. The neurological effect is a movement disorder, with abnormalities of speech, tremor, incontinence (lack of self control) and dystonia being common features. In spite of many known anaesthetic problems, there are very few reports of General Anaesthesia and Regional Anaesthesia in these patients, so we report a case of Wilson's Disease with anaesthetic management.

CASE REPORT

A 14 yr old girl from eastern Nepal presented to Kathmandu Medical College hospital for treatment of compound fracture of right shaft of femur. She was underweight, debilitated and diagnosed as Wilson's Disease two years ago in Christian Medical College (CMC), Vellore, India. Initially, there was jaundice and then she developed difficulty in standing, walking and speaking. Gradually there was abnormal posturing of limbs and tremors as well. When the patient presented in the hospital, all limbs were flexed with maximum rigidity except the fractured one (Fig 1). The patient was irritable, uncooperative & unable to speak. She was given Clonazepam, Pacitane & Tramadol by physicians.

Clinical features were suggestive of Wilson's disease with signs of chronic liver disease. Investigations showed deficient serum Ceruloplasmin and increased serum copper level. She was being treated with copper chelating agent: D-penicillamine initially and then Trientine and Zinc with avoidance of copper containing food.

Pre- anaesthetic investigations were within normal limits (including Electrocardiography and Echocardiography) except slightly deranged Liver Function Tests

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(Prothrombin Time-16 seconds, International normalized ratio, INR-1.25, slightly increased liver enzyme levels). Because the patient was very irritable and she had abnormal posturing and movement, regional anaesthesia was not possible and the case was planned under General Anaesthesia (GA).

Under standard monitoring & venous assess, GA was induced with Propofol, Pethidine and Vecuronium and maintained with 50% oxygen in air and Isoflurane with addition of Vecuronium as the surgery lasted for two hours. Her haemodynamics were within normal limits during the whole intraoperative period. The effect of vecuronium was expected to be prolonged due to liver dysfunction but the recovery was normal. She was reversed with neostigmine and atropine and extubated in the operating room. There was no deterioration of liver function or renal function in post operative period.



Figure 1: Photograph of the patient with flexion deformity of limbs.

DISCUSSION

In people with Wilson's disease, copper begins accumulating in the liver immediately after birth but signs and symptoms develop in first decade of life & sometimes not until third or fourth decade. The clinical manifestations vary amongst the patients, most common is neurological (69%), then hepatic (15%), and then behavioral/psychiatric (2%) & musculoskeletal (2% of cases)^{1,2}.

The main pathology is copper accumulation, due to deficient Ceruloplasmin which is necessary for excretion of copper from liver to bile. The excess copper causes damage primarily to liver and brain and the mentioned sign and symptoms occur. Once the disease comes into consideration, diagnosing steps should be carried out for Wilson's disease. These include urinary copper; blood Ceruloplasmin; slit lamp Kayser-Fleischer rings and clinical findings^{3,4}.

Currently there are four drugs being used as anti copper agents: Zinc blocks intestinal absorption of copper, Penicillamine and Trietine are chelators in blood which help in urinary excretion and tetrathiomolybdate which forms complex with copper and protein and blocks absorption from gut².

There are very few reports of administration of anaesthesia in patients with Wilson's disease. Some reports of regional anaesthesia for limb surgeries and general anaesthesia for dental extraction. Tanaka K et al

reported that there was no prolongation of Vecuronium during general anaesthesia with neuromuscular monitoring³. De Souza Hobaika AB and El Dawlatly AA et al reported regional anaesthesia in these patients without prolonging the effect^{1,5}. Baykal M, Karapolat S reported safe general anaesthesia with use of drugs which do not affect the liver function in these patients⁶.

Regarding which method is most appropriate in such cases, GA may be disadvantageous in that it may aggravate the already impaired liver function and problem in metabolism & elimination of drugs used. Inducing agents and inhalational agents may interfere with central nervous system function and may exacerbate neurological & behavioral problems postoperatively⁵.

These patients may be more sensitive to muscle relaxants due to reduced muscle function either from disease itself or from the use of D-penicillamine. Regional anaesthesia is safe if possible in these cases because of less use of drugs and minimal effects on vital organs⁶.

CONCLUSION

No patient is contraindicated for anaesthesia and surgery, provided we modify the techniques. Patients with Wilson's disease will have major hepatic and neurological involvement; therefore, according to site of surgery and disease extent, we have to choose the techniques with careful evaluation and perioperative monitoring.

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