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Case Report

Anaesthetic Management of Scoliotic Deformity with Wilson's Disease

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Abstract
Wilson's disease is an inherited autosomal recessive disorder of copper metabolism. As a consequence copper accumulates in liver, brain, kidney and red blood cells. Patients may develop chronic liver disease, hemolytic anaemia and neurological symptoms. Hepatic involvement may interfere with metabolism of various anaesthetic drugs. Anaesthesia for deformity correction of scoliosis itself is challenging and associated with various complications. Very few cases has been reported of anaesthetic management in patients with wilson's disease associated with scoliosis.
Keywords: *Wilson's disease, scoliosis, General Anaesthesia.*

Introduction

Wilson disease is an inherited autosomal recessive disorder characterized by reduced synthesis of ceruloplasmin-a major copper transport protein in blood. As a result, copper accumulates in brain liver and various other tissues of the body causing mainly hepatic and neuropsychiatric insult. The clinical features vary among patients and most common is neurological (69%) followed by hepatic (15%), behavioral or psychiatric (2%) and musculoskeletal (2%) of cases.[1]

Hepatic involvement may alter the metabolism and excretion of many anaesthetic drugs. There are very less reported cases of GA in wilson's disease. Anaesthetic management of scoliotic deformity itself is very challenging. Therefore we present a case report for anaesthetic management of deformity correction of scoliosis associated with wilson's disease.

Case Report

A 22 year old male with Wilson disease was admitted in AIIMS, NEW DELHI, for corrective surgery of kyphoscoliotic deformity. C3 to D11 vertebral level were involved with cobb's angle of 70 degree. No neural deformity was present.

History

Patient was apparently normal till 16yrs of age, after which he started complaining of pain in

dorsolumbar area associated with deformity which was gradually increasing. 4yrs later patient developed abdominal pain and distention with abnormal movement of right upper limb. Opthalmological examination revealed bilateral KF ring in both the eyes. USG abdomen showed features suggestive of liver cirrhosis with portal hypertension and hypersplenism. His serum ceruloplasmin was 7.8 mg/dL and 24 hrs urinary copper was 253.05mg. family screening showed positive history of wilson's disease in his brother. The patient was started on D-Penicillamine initially and then Zn 50mg TDS. He had to undergo splenectomy under GA which was uneventful.

Pre-Operative Evaluation

There was no other associated co-morbidities. Effort tolerance was > 4 Mets. There was no anticipated difficult airway. Per-operative investigation included complete hemogram, coagulation profile, renal function test, serum electrolytes, liver function test, Chest X-Ray, PFT, ECG, echocardiography. All the parameters were within normal limit.

Intraoperative Management

Induction

Standard ASA monitors were attached. Large bore i.v access secured. Patient was induced with 2microgram/kg fentanyl, propofol with titrated doses and atracurium 0.5 mg/kg. Airway was secured with 7.5mm ETT. Arterial, central line, urinary catheter and nasopharyngeal temperature probe was inserted. Surgery was planned in prone position, so all pressure points including eyes were carefully padded.

Monitors

- Standard ASA monitors
- Invasive blood pressure
- Temperature (hypothermia was prevented using fluid warmers and warming blankets)
- Urine output per hour
- Neurophysiological monitoring (SSEP and MEP)
- BIS (for depth of anaesthesia)- values maintained within 40-60 throughout the surgery.

Maintainence of Anaesthesia

Propolof infusion at the rate 50-150microgram/kg/min and fentanyl infusion at the rate of 1-2 microgram/kg/hr was run continuously to maintain the depth of anaestheia with target BIS value of 40-60. Oxygen and air was used. Muscle relaxant was not used post induction.

Other Drugs

- Tranexamic acid-10mg/kg loading dose followed by1mg/kg/hr infusion as maintenance dose throughout the surgery to prevent blood loss
- Ondansetron- to prevent post operative nausea vomiting
- Dexamethasone- to prevent airway edema and post operative nausea vomiting
- Ketorolac- for multimodal analgesia
- Morphine and paracetamol were avoided

Intra-Operative Event

Total duration of surgery was 5 hours. There was about 1.5L of blood loss. 4 unit of each PRBC, FFP and PC were transfused. A total of 2L of stereofundin isotonic solution crystalloid was transfused. Urine output was about 0.5-1mL/kg/hr. All hemodynamic were within normal limit with MAP>60mm of Hg was maintained throughout the surgery.

Post-Operative Management

Patient was extubated in the operating room. Vitals were stable and he was shifted to HDU for further monitoring and pain management. Multimodal analgesia was used for pain. Fentanyl 1microgram/kg/hr infusion with diclofenac 75mg BD and tramadol 50mg BD was given.

Post-operative investigations included CBC, coagulogram, CXR, RFT, LFT, serum electrolytes. LFT was monitored daily for 48hrs. There was no deterioration of any parameters or development of any neuropsychiatry symptoms. Patient was satisfactorily discharged after few days.

Discussion

Hepatic blood flow is decreased during general anaesthesia due to the effects of anaesthetic agent. Scoliosis surgery is associated with substantial blood and heat loss which may lead to hemodynamic instability. This hemodynamic instability may further aggravate hepatic insult.

Impaired hepatic function can affect the metabolism and excretion of muscle relaxant, analgesics and sedatives. Hypnotics and sedative drugs interfere with CNS and may precipitate or exacerbate the neuropsychiatric symptoms post-operatively.[1] A case of WD diagnosed by occurrence of acute neuropsychiatric symptoms after general anesthesia has been reported.[3] Massive blood loss may decrease cerebral perfusion which may further trigger neuropsychiatric dysfunction. Hence fluid balance and blood transfusion must be meticulously managed and beat to beat monitoring by using invasive arterial monitoring must be done to prevent hemodynamic instability. If required vasopressors can be used.

There is increased sensitivity to neuromuscular relaxants in wilson's disease either from disease itself or from the use of D-penicillamine. Atracurium can be used safely as its metabolized mostly by hoffman's degradation. Use of neuromuscular monitoring is advocated. In our case inhalational agents were not used as it interferes with SSEP and MEP monitoring. Only single dose of atracurium was used for intubation and once the effect was weared off, the SSEP and MEP monitoring was started.

Pain management is challenging in scoliosis surgery as pain is severe due to extensive procedures. A multimodal approach of analgesia is recommended. Hepatotoxic drugs must be avoided. In our case, we avoided PCM and morphine. However fentanyl is a narcotic agent and can be safely used.[4] NSAIDS can be used to supplement analgesia. Regional techniques can also be used like administration of epidural opiods.

Whenever there is a choice between regional and general anaesthesia, regional techniques should be preferred as GA may aggravate already impaired hepatic dysfunction. Regional techniques also has the advantage of opioid sparring effect.[5] However general anaesthesia can be safely administered with meticulous approach. Tanaka et al reported that vecuronium can be used in these patients with neuromuscular monitoring.[6] Baykal et al concluded in his report that GA can be administered in Wilson disease patients by using anaesthetic agents that are least toxic to liver.[7]

Conclusion

Though regional is preferred over General anaesthesia but GA can be safely administered in asymptomatic patients with Wilson disease. A proper pre-operative evaluation with judicious use of anaesthetic agents must be ensured. Hepatotoxic drugs should be avoided. A meticulous monitoring and follow-up is required both intra-operatively and post-operatively to secure a successful outcome.

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