

CASE REPORT

Spinal anesthesia in a case of Huntington's Disease

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ABSTRACT

Huntington's disease (HD) is an autosomal dominant degenerative disorder. This disease has been reported to have a number of anesthetic implications; however, no firm recommendations are available regarding the use of general versus regional anesthesia in these patients. A fifty two year old female with a three year history of HD presented with right intertrochanteric fracture and was posted for dynamic hip screw fixation. Preoperative and routine laboratory investigations were unremarkable except for gross choreiform movements, dysarthria and cachexia. However, with adequate support spinal anaesthesia was given using 0.5% hyperbaric bupivacaine and a sensory level of T8 was achieved. Choreiform movements were completely abolished in the lower limbs and motor power began to recover after ninety minutes of spinal anesthesia. The reported anesthesia experience in these patients is anecdotal and consists of a limited number of case reports and letters, of which majority of reports are of general anesthesia.

Key words: Huntington's disease; Choreiform movements; Spinal anesthesia

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INTRODUCTION

Huntington's disease [HD] is an autosomal dominant neurodegenerative disorder that affects about 5-7 per 100,000 individuals. Clinical onset is usually between 30 and 50 years of age. The disease has a strong family history, is progressive and usually leads to a fatal outcome within 15 to 20 years.

The initial symptoms may be abnormal movements or intellectual changes but ultimately both occur. The earliest mental changes are behavioural but subsequently dementia develops. At the outset, a mild dyskinesia is often present which initially may be no more than an apparent fidgetiness or restlessness but eventually choreiform movements occur.

Essentials of diagnosis include gradual onset and progression of chorea and dementia or behavioural changes and a family history of the disorder. Patients develop a wide, swaying gait, and falls are common.

There is no cure for Huntington's disease. The treatment is largely symptomatic. The biochemical changes suggest a relative under activity of neurons containing GABA and acetylcholine or a relative over activity of dopaminergic neurons. Patients are usually on dopamine antagonists or vesicular monoamine transport inhibitors (VMAT).

Huntington's disease has been reported to have a number of anesthetic implications; however no firm recommendations are available regarding the use of regional versus general anesthesia in these patients². We present the successful anesthetic management of a patient with Huntington's disease under sub-arachnoid block.

The case report may help alleviate the unfound fear from the anesthesiologists' minds about giving spinal to patients with neurological diseases.

CASE REPORT

A 52 year old woman, with a 3 years history of HD was admitted after a fall, resulting in right

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intertrochanteric fracture. She was posted for Dynamic Hip Screw (DHS) fixation.

There was a positive family history of a similar disorder in her father and a younger sister.

Preoperative examination and routine laboratory investigations were unremarkable except for gross choreiform movements in all the four limbs, mild dysarthria, and cachexia. The only medication the patient was receiving was tab tetrabenazine 25 mg orally thrice daily.

The procedure was explained to the patient during the pre-anesthetic visit. Informed consent for anesthesia and surgery was taken. The patient was premedicated with alprazolam 0.5 mg orally. After an eight hour pre-operative fasting period, she was taken up for surgery.

Inj. midazolam 1 mg intravenous was administered after securing the intravenous line on the operating table. Routine monitoring was performed with NIBP, ECG and Pulse oximeter. She was co-loaded with 500 ml of crystalloid solution.

It was initially difficult to position the patient for spinal anesthesia (SA) due to her choreiform movements. However, she could be placed in sitting position with support, and SA was administered with 2.5 ml of 0.5% of hyperbaric bupivacaine (12.5 mg) using 25G Quincke spinal needle on first attempt. The patient was placed in the supine position and a sensory level of T8 was achieved after 8 min of drug administration. The effect of the SA was adequate for surgery, 10 min after the administration of the drug. Choreiform movements were completely abolished in the lower limbs till motor power was regained. Motor power in the lower limbs began to recover after 90 min of administration of SA.

Intra-operatively all vital parameters remained stable and no anesthetic complication occurred. Administration of vasopressors was not required. Optimal surgical conditions were provided by the SA for entire duration of the surgery which lasted for 50 min. Complete recovery of motor and sensory blockade occurred after around 150 min of administration of SA.

In the postoperative period, analgesia was provided with administration of intramuscular diclofenac

sodium and tramadol.

On third postoperative day she developed a mild headache with typical features of post-dural puncture headache in the occipital region which was relieved on lying flat. However, this responded to simple analgesics and subsided within hours. Thereafter, her postoperative course was uneventful and she was discharged on eighth postoperative day.

DISCUSSION

Experience with the management of anesthesia in patients with Huntington's disease is too restricted to recommend a specific anesthetic drug or technique.² The reported anesthesia experience in these patients is largely anecdotal and consists of a limited number of case reports and letters. The majority of case reports describe general anesthesia (GA).

A number of complications including prolonged apnea, prolonged recovery, generalized tonic spasms, postoperative fever, increased sensitivity to barbiturates and benzodiazepines and postoperative shivering have been reported⁵. The increased likelihood of pulmonary aspiration must be considered if pharyngeal muscles are involved.² However all reported complications have been in cases which received GA.

A literature search of English language journals revealed only five case reports of SA in patients with HD.^{3,4} None of the other case reports involving SA in HD patients reported any adverse intra operative or postoperative event. The mild postspinal headache in our case, can be attributed to the use of the Quincke needle (the non-availability of a pencil point needle precluded its use).

Patients with HD, who do not have marked psychiatric symptoms, can co-operate sufficiently to undergo SA. These patients may be difficult to position for SA, but if this difficulty is overcome it appears to be a safe and effective alternative to GA in appropriate surgical procedures.

Conflict of interest: None declared by the authors

Author contribution: All of the authors took part in the management of this patient and in manuscript preparation.

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Prof. Naveed Masood

⇒ Traumatic shock. Anaesthesiologist 's perspective, insight and management
Prof. Fazal H. Khan

⇒ Pain, sedation and delirium guidelines
Prof. Salman Waris

⇒ Obstetric patient coming in for non obstetric surgery. Safety guidelines
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⇒ Acute and Chronic pain. Basics of management
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