

INFRACLAVICULAR BRACHIAL PLEXUS BLOCK IN WILSON'S DISEASE

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Abstract

Wilson's disease (WD) is characterized by progressive copper accumulation with hepatic and neurological impairment. Anesthesia and surgical practices may exacerbate WD and liver damage, and even cause life-threatening liver failure. Due to this existing liver damage, anesthetic management is important in WD cases in terms of drug choice, dose, and technique used. This study reports an emergency surgical procedure for trauma in a 24-year-old WD patient suffering the disease for 18 years. The operation was planned under infraclavicular brachial plexus block because of a right supracondylar/proximal humerus fracture. The selected type of anesthetic technique and agents in WD is specific. The pharmacokinetic changes in these cases are difficult to predict and require attention to drug choice and dose.

Key words: Wilson's disease, plexus block, regional, anesthesia.

Introduction

Wilson's disease (WD), also known as hepatolenticular degeneration, is a rare autosomal recessive disorder which typically occurs in older children or young adults. A reduction in the synthesis of the copper transporter protein (ceruloplasmin) leads to impairment of copper excretion into bile from lysosomes in hepatocytes, due to mutations in the ATP7B gene on chromosome 13 in patients¹. As a result, copper cannot be removed by the bile duct and accumulates in various organs and tissues, particularly the liver². It also accumulates in the liver, brain, kidney, and cornea, impairs organ function, and has toxic effects by creating free radicals^{3,4}. In 40% of patients, the first sign is liver disease, varying in type from asymptomatic with only biochemical abnormalities to acute liver failure or chronic liver disease leading to cirrhosis⁵. The most common clinical signs are neurological (dysarthria and gait disturbance) and psychiatric (personality disorders and depression) changes^{6,7}.

Due to this existing liver damage, anesthetic management is important in WD cases in terms of technique, drugs, and doses⁸. This case report will discuss the anesthesia protocol used during an emergency surgical procedure for trauma in a WD patient, and experiences in the perioperative and postoperative periods.

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Case Report

The 24-year-old male patient had suffered WD for 18 years. He was 68 kg in weight. The operation was planned following a right supracondylar/proximal humerus fracture. The patient had difficulty in walking, speech, and had tremor and involuntary muscle contractions on preoperative examination. In addition, he had complaints of double-vision and Kayser-Fleischer ring in both eyes. There was no hepatosplenomegaly. Blood biochemical analysis was normal. The patient underwent 750 mg/day penicillamine therapy for about three years, and continued a low copper diet (<1 mg/day of copper) for three years. The patient was taken to the operating room, underwent standard monitoring (non-invasive blood pressure, ECG and pulse oximetry), and given 1 mg of midazolam i.v for premedication.

For the infraclavicular brachial plexus block, the patient was placed in a supine position and the skin over the coracoid process was disinfected. Local anesthetic (0.25% 30 mL plain bupivacaine) was injected at 2.0 cm medially and 2.0 cm caudally to the center of the coracoid process. Subsequently, the needle attached to the nerve stimulator (B.Braun, Melsungen, Germany), and the syringe containing local anesthetic penetrated the skin at the same point. Once the optimal motor response in the range of 0.3-0.5 mA was achieved, 0.25% 30 mL plain bupivacaine was administered as a single injection followed by aspiration. Onset of the sensory block was assessed by cold test using an alcohol-soaked swab. The cutaneous dermatomes of the four major nerves of the upper limb were assessed and compared with the opposite side. This evaluation was repeated every 5 minutes for 20 minutes. To quantify the sensory block, the level of sensation of the alcohol-soaked swab and needle was graded as 0 (no sensation), 1 (hypoesthesia sensation), or 2 (normal sensation). We defined successful block as a lack of sensation (score of 0) in all four areas of innervations after 20 minutes, or a block that failed to provide appropriate surgical anesthesia and required supplementary anesthesia/analgesia. For evaluation of motor block, the patient was asked to make specific movements from which the physicians assessed the block of specific muscles. Thumb abduction was used to evaluate radial nerve, thumb adduction for

ulnar nerve, thumb opposition for median nerve, and elbow flexion in supination and pronation for the musculocutaneous nerve. It was assessed at 5, 10, 15, and 20 minutes after injection of local anesthetic. No sedation was added and oxygen therapy 1 L/min *via* a nasal cannula was administered. Surgery lasted 110 minutes. Afterwards the patient did not complain of any pain at the surgical or tourniquet sites. Oxygen saturation (SpO₂) remained in the range of 96% to 98%. The patient was discharged from the post-anesthesia care unit one hour postoperatively. The sensory and motor blocks lasted 20 hours.

Discussion

WD initially occurs as a liver disease in children⁸. Although clinical course varies among patients, neurological symptoms are usually the most common symptoms⁶. In early diagnosis, histological changes on liver biopsy, high and low serum ceruloplasmin level and 24-hour urine copper excretion are valuable. The goal of treatment is to prevent progression of the disease by reducing the accumulation of copper tissue and organs. For this purpose, D-penicillamine is used⁵.

Very few publications related to the anesthetic management of WD have been reported. There is no clear consensus on the safest anesthetic technique and agent^{8,9}. It has been expressed that general anesthetic agents such as hypnotics, narcotics, and muscle relaxants may aggravate neurological and psychiatric problems, increase existing hepatic damage, and affect the central nervous system in the postoperative period. It has also been expressed that patients are more sensitive to muscle-relaxing agents than normal patients due to use of d-penicillamine¹⁰. Systemic blood pressure, hepatic blood flow, and tissue perfusion are decreased due to hemodynamic changes that occur during general anesthesia. The decreased blood flow in WD with failed hepatic function may increase existing hepatic damage and adversely affect drug metabolism.

Despite all these negative anesthetic effects, the literature has also reported smooth application of general anesthesia⁸⁻¹⁰. However, this may be due to patients in the pediatric age group. Because general anesthesia may aggravate current disease, we decided to apply a regional anesthesia to our patient who has

clinical complaints and 18-year history with WD. After varicocelectomy under general anesthesia, aggravated cases of WD have been reported in the literature¹¹. Anesthesia and surgical practices may exacerbate WD and liver damage, and even cause life-threatening liver failure¹². As many agents used in general anesthesia are metabolized by the liver, the effect duration of these drugs can increase in WD. When choosing the method of anesthesia in patients with WD, the method which causes less effect to the liver must be chosen.

Electrophysiological changes in patients with WD are typically seen in the central nervous system¹³. However, some studies have found that the effect of the chelators used to treat WD caused polyneuropathy

in the peripheral nerves^{13,14}. Myelin loss and axonal degeneration in peripheral nerve biopsies have also been observed¹⁵. For this reason, the literature has shown that the peripheral nerves of patients with WD are more sensitive to local anesthetic agents. This may be so; however our patient did not require any additional analgesic in the 12 hours period after the block.

In sum, it should be considered that in a patient with WD showing hepatic or neuropsychiatric findings, general anesthesia may aggravate existing disease. If possible, regional anesthesia techniques and decreased local anesthetic dose are preferred.

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