

ANESTHESIA FOR A PATIENT WITH WILSON'S DISEASE

- A Case Report -

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Introduction

Wilson's disease (WD), or hepatolenticular degeneration, is an autosomal recessive disorder characterized by a reduction in the synthesis of the copper transporter protein ceruloplasmin. The outcome of this genetic defect is the accumulation of copper in body tissues and consequent hepatic and neurological impairment.

Case Report

A 53 year old woman presented to the operating room for treatment of a rotator cuff tear of the right shoulder. The symptoms exhibited by the patient were dysarthria, tremors of the upper extremities and a strong pain in the right shoulder. She had been diagnosed with WD some 25 years earlier during pregnancy, and was being treated with the copper chelating agent D-penicillamine (750 mg/day).

Preoperative tests were normal. Under standard monitoring and venoclysis, the patient was sedated using midazolam (6.0 mg). The interscalene block was performed on the right side according to the

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Winnie technique using ropivacaine (35 mL, 0.5%) with the aid of a peripheral nerve stimulator (Stimuplex®; B. Braun Medical Inc., Allentown, PA, USA). Latency was 12 minutes and there were no complications during surgery. The interscalene block regressed completely after 16 h and the patient was discharged from the hospital the following day.

Discussion

The clinical manifestations of WD vary amongst patients, and may be present as neurological (69%), hepatic (15%), psychiatric (2%) and osteomuscular (2%) symptoms¹. Osteomuscular problems are generally diagnosed during the second decade of life and are typically less severe than the other symptoms, as appears to be the case in the patient to be presented.

Only three reports concerning the administration of anesthesia (general, subarachnoid and epidural) in WD patients have been published so far, and there appears to be no consensus regarding which of these techniques is the most appropriate for such patients²⁻⁴. General anesthetics are disadvantageous in that they may aggravate the already impaired hepatic function and may not be properly metabolized. Hypnotic and sedative drugs interfere significantly with the central nervous system and may, therefore, exacerbate neurological and psychiatric problems in the post operative period. Finally, WD patients may be more sensitive to neuromuscular relaxants than normal patients by virtue of reduced muscle function resulting either from the disease itself or from the use of D-penicillamine.

Some researchers have observed that visual and auditory evoked potentials are altered in WD patients, suggesting damage of the encephalic structures and cerebral trunk. However, the peripheral nerve conduction and the somatic-sensory evoked potentials are normal¹⁴. Such evidence suggests that the regional administration of local anesthetics to WD patients may be safe, since the peripheral nerve transmission is not altered.

In summary a case of a WD patient is presented exhibiting neurological and osteomuscular symptoms, who was submitted to a successful shoulder surgery under an interscalene brachial plexus block.

References

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