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Stiff Man Syndrome and Anaesthetic Considerations: Successful Management Using Combined Spinal Epidural Anaesthesia

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Stiff Man Syndrome SPS is a rare and disabling disorder, characterized by muscle rigidity with superimposed painful spasms involving axial and limb musculature.[1] The stiff man syndrome is characterised by paraspinal and abdominal rigidity with an exaggerated lumbar lordosis and superimposed spasms precipitated by movement, emotional upset, peripheral stimulation or auditory startle.[2] Most patients belonging to classical SPS have antibodies against glutamic acid decarboxylase (GAD), which is the enzyme essential for GABA synthesis. Loss of GABAergic input from higher centers and spinal interneurons causes hyperactivity of the motor neuron system and subsequent progressive muscle rigidity.[3] Treatment is mostly based on benzodiazepines, baclofen and immunosuppresants.

CASE REPORT

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A 55 year old lady suffering from SPS was scheduled to undergo bilateral amputation through her knee joints to decrease pain and to assist in getting a prosthesis, which would help in her mobility. She had a history of SPS which was progressing over the years making her bed ridden for the last 4 years. She had severe muscle rigidity affecting her axial and limb musculature. Her lower limbs, especially feet, were severely affected with rigidity and contractures causing severe, continuous pain. She also suffered from hypertension, diabetes, hypothyroidism. Her medications included baclofen, clonazepam, thyroxine, insulin, metoprolol, hydromorphone (for pain relief) and pregabalin. As she was bedridden her exercise tolerance could not be assessed. Her SPS was confirmed by GAD levels and was optimized preoperatively. On the day of surgery, all her daily medications were given orally. The patient was explained about the nature of surgery and anaesthetic plan. Severe rigidity in her axial muscles did not allow her to sit. Before positioning premedication in the form of midazolam 1 mg was given IV. In left lateral position, a lumbar epidural catheter was inserted under aseptic precautions at L4-5. A spinal anaesthetic was administered at L3-4 using 1.8 ml of 0.5% bupivacaine (heavy). The patient did not show any signs of induced spasms during the neuraxial procedure. Unfortunately the spinal anaesthesia did not obtain the desired level; but adequate level (upto T10-T12) was achieved with supplementation of epidural anaesthesia. Throughout the procedure, conscious sedation was

administered using a total of 50 mcg of fentanyl and 2.5 mg of midazolam. Patient was monitored with continuous ECG, non invasive blood pressure, and oxygen saturation during the surgery. Epidural analgesia was used for postoperative pain relief for 3 days, after which the patient was successfully discharged home, without complications.

DISCUSSION

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SPS was first described by Moersch and Woltman.[2,4] The cause of the stiff-person syndrome is unknown but an autoimmune pathogenesis is suspected because of the presence in the cerebrospinal fluid (CSF) of antibodies against GAD, the rate-limiting enzyme for the synthesis of GABA, the association of the disease with other autoimmune disorders, the presence of various autoantibodies and a strong immunogenetic association. Guilleminault et al[5] have suggested a functional imbalance between descending aminergic, possibly reticulospinal projections to the cord, facilitating flexor reflex pathways, and inhibitory GABAergic systems. Two types of drugs have been used in SPS: drugs that enhance GABA activity and immunosuppressing drugs. Diazepam, which increases the frequency of opening of the GABA-A receptor and leads to hyperpolarization, is the initial treatment of choice at daily doses up to 200 mg. Intrathecal or oral baclofen may improve the physical symptoms along with steroids and immunosuppresants.[4] There are only a few case reports in literature regarding anaesthetic management of SPS patients. Earlier case reports observed increased hypotonia with general anaesthesia. Johnson reported postoperative muscle weakness requiring mechanical ventilation despite appropriate reversal of muscle blockade.[6] Bouw et al suggested that, not only muscle relaxants, but volatile anaesthetic agents can also potentiate hypotonia by their action on GABA-B receptors in patients taking baclofen preoperatively.[4]However, Ledowski and Russel also describe the successful use of total intravenous anaesthesia (TIVA), in a patient of SPS.[7] With its known benefits of superior analgesia, Regional anaesthesia (RA) is advocated as the safest mode of anaesthesia. However RA can be potentially difficult and can induce spasms because of the pain during needle entry. Care must be taken by proper preoperative explanation to alleviate fear and anxiety and adequate premedication. Elkassabany et al demonstrated successful use of paravertebral block in a patient of SPS undergoing hernia surgery.[8] It is significant to note that loud auditory stimulation can induce spasms; conscious sedation sufficient to keep the patient calm must be employed, as in this case, to prevent any SPS symptoms. In our case spinal anaesthesia failed to achieve adequate levels, perhaps because of reduced volume. Since any high spinal anaesthetic can potentially cause respiratory difficulty, especially with increased chest rigidity, we opted for a lesser volume. Continuous epidural analgesia provided effective pain relief postoperatively.

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