

# Anesthetic management of a patient with spinocerebellar degeneration

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## Abstract

Spinocerebellar degeneration or olivopontocerebellar degeneration denotes a group of disorders of various etiologies manifesting as degenerative changes of various part of the central nervous system. We describe the anesthetic management of a patient with severe olivopontocerebellar degeneration posted for vaginal hysterectomy. A combined spinal epidural technique was performed at the level of L2-L3. The anesthetic implications of the various aspects of spinocerebellar degeneration are discussed.

**Key words:** Ataxic disorders and anesthesia, combined spinal epidural, olivopontocerebellar degeneration, spinocerebellar degeneration

## Introduction

Spinocerebellar degeneration or olivopontocerebellar degeneration denotes a group of disorders of varied etiologies manifesting as degenerative changes of various part of the central nervous system. The cerebellum, cerebral cortex, inferior olives, basal ganglia, substantia nigra and the spinal cord are involved. The predominant areas to be affected are the cerebellum and its afferent and efferent pathways including spinocerebellar pathways and fronto-ponto-cerebellar pathways. In severe cases, the degeneration is also seen in the cerebral cortex, basal ganglia and substantia nigra. We describe the anesthetic management of a patient with severe olivopontocerebellar degeneration posted for vaginal hysterectomy.

## Case Report

A 45-year-old woman with history of progressive ataxia, dysarthria and intentional tremors presented with complete

uterine prolapse. She had difficulty getting up from squatting position, difficulty in swallowing and had episodes of coughing on rapid oral fluid intake. Speech difficulties started about three years back. She complained of progressive increase in difficulty in walking and speaking in the last three years. Her family history was unremarkable. On examination her higher functions revealed scanning speech and emotional instability. She exhibited signs of recent memory loss but her long-term memory was intact. Muscle tone was markedly increased in all four limbs and spastic paraparesis was present. Motor power was Grade 4/5 in upper limb and 3/5 in lower limbs. Plantar reflex was extensor and deep tendon reflexes were exaggerated. Cerebellar signs were positive including intentional tremors, ataxia and past pointing. Her pupils were dilated and reacted sluggishly to light, denoting autonomic neuropathy. Other signs of autonomic imbalance like postural hypotension, absence of heart rate variability were absent. Sensory system was intact. Cardiovascular and respiratory systems were within normal limits.

Magnetic resonance imaging (MRI) of the brain showed atrophy of cerebellar hemispheres, vermis, pons and cerebellar peduncles. Small inferior olives and small pons were noted. Prominence and widening of sulci and basilar cisterns were noted with partial atrophy of the frontal, temporal and cerebral lobes. Prominence of transverse and sylvian fissures was also noted. No focal mass lesion or hyperintensity was noted. The above findings were indicative of olivopontocerebellar degeneration. The patient was not on any medications, and the treating physician opined that her clinical condition was stable and did not require any intervention till the surgery.

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Chest X-ray showed increased bronchovascular markings. Electrocardiogram (ECG) and routine blood investigations were normal.

Considering the possibility of erratic response to neuromuscular blockade and risk of aspiration and delayed recovery of airway reflexes, a combined epidural-spinal technique was planned. The procedure was explained to the patient and consent taken. Midazolam 1 mg intravenous was administered as premedication. Baseline pulse rate was 80/min and blood pressure was 130/80 mmHg. Routine monitoring with 5-lead ECG, NIBP and pulse oximetry was started and patient preloaded with 500 ml of lactated Ringer's solution. Combined spinal-epidural anesthesia was administered via L2-L3 interspace, 2.5 ml of 0.5% bupivacaine heavy was administered intrathecally and 18G epidural catheter was passed and fixed at 9 cm (space located at 5 cm). Maximum sensory level achieved was T8 as judged by absence of pinprick sensation.

Her blood pressure fell to 90/60 mmHg from 130/80 mmHg after 15 min of the procedure. She responded to 6 mg of intravenous ephedrine, and did not require any other vasopressors. Epidural top-up of 5 mL of 0.5% bupivacaine was given after two-segment regression after a negative test dose. The surgical procedure lasted for 3 h and was uneventful. At the end of the procedure, 5 mL of 0.0125% bupivacaine was given epidurally for postoperative analgesia. The postoperative period was uneventful. Her motor recovery was complete within approximately one hour of the surgery with return of her original spasticity and power. There was no progression of her neurological symptoms in one month of follow-up.

## Discussion

Spinocerebellar atrophy or olivopontocerebellar degeneration belongs to a group of ataxic disorders characterized by diverse neurological symptoms depending on the anatomical site of lesions. The common signs include ataxia, progressing to static and kinetic motion disorders and dysarthria gradually rendering the speech unintelligible. Later in the course of the disease the patient can develop other motion abnormalities, signs of Parkinsonism, ocular disturbances, autonomic neuropathy and mental deterioration. Bowel and bladder disturbances also occur.

The etiology of the ataxic disorders can be classified according to the onset and progress of the disease.<sup>[1]</sup> Symmetric ataxias can be classified as Acute (hours or days), Subacute (weeks or months) or Chronic (months to years) [Table 1].

More than 30 types of ataxias are identified with various mutations. The predominant being spinocerebellar ataxias

Type 1 through Type 22, Friedreich's ataxia, Kearns-Sayre syndrome, MELAs (mitochondrial encephalopathy, lactic acidosis and stroke syndrome), Leigh's disease and ataxia telangiectasia. Most inherited ataxias follow an autosomal dominant pattern. Autosomal recessive, sporadic mutations and mitochondrial mutations account for the rest. The clinical features of most of the ataxias overlap, and genotypic evaluation is the basis of classification and predicting the course of the disease. It is important to note that certain ataxias are associated with other disorders relevant to the anesthesiologist like seizures, cardiomyopathy, various heart blocks, diabetes mellitus, scoliosis, and immune disorders like IgA and IgG deficiency. There is no effective treatment for this group of disorders other than supportive measures.<sup>[2]</sup>

Patients with spinocerebellar degeneration have been successfully managed under regional and general anesthesia.<sup>[11,14,15]</sup> Anesthetic considerations in a patient of olivopontocerebellar degeneration are multiple [Table 2]. Tremors and rigidity may cause difficulties in patient positioning. Although difficult to perform, central neuraxial blockade helps in proper positioning of patients with spasticity. The patient had intentional tremors but was comfortable at

**Table 1: Classification of ataxic disorders**

Acute	Subacute	Chronic
Alcohol intoxication	Malnutrition	Inherited
Drugs (barbiturates, phenytoin, lithium, fluouracil, paclitaxel)	(B1, B12 vitamins) Hyponatremia	Sporadic mutation
Infections (post varicella, toxoplasmosis, Epstein Barr Virus)	Malignancies (small cell carcinoma lung, Hodgkin's lymphoma, neuroblastoma, carcinoma breast, carcinoma ovary)	Syphilis
		Hypothyroidism
		Multiple sclerosis (not symmetrical)

**Table 2: A comparison of regional and general anesthesia for spinocerebellar disorders**

Anesthesia	Pros	Cons
Regional Anesthesia (spinal, epidural)	Profound Analgesia Spasticity relieved, muscle relaxation Predictable pharmacodynamics Avoid airway and ventilation-related problems Postoperative analgesia	Technically difficult Autonomic imbalance ?Progression of neurological deficits Difficulties in positioning Impaired communication
General Anesthesia	Patient comfort Stable hemodynamics Neurological assessment easy Positioning easier (Muscle relaxation may be suboptimal though)	Unpredictable responses to muscle relaxants Risk of aspiration (delayed recovery of gag reflex) Ventilatory depression

rest and able to lie down in supine position without discomfort. Emotional instability and dysarthria may cause communication problems.

Autonomic neuropathy and involvement of spinocerebellar tracts pose a threat of unpredictable responses to central neuraxial blockade. Autonomic neuropathy has been shown to cause hypotension during positive pressure ventilation.<sup>[3]</sup> Our patient had history of difficulty in deglutition, choking and regurgitation on fluid intake which indicated bulbar muscle dysfunction. Bulbar muscle dysfunction predisposes to aspiration and delayed recovery of gag reflexes.

Scoliosis and muscular dystrophies, though not present in this case, should be ruled out. Exaggerated response to barbiturates, and both depolarizing and nondepolarizing muscle relaxants may exist in patients with spinocerebellar atrophy, increasing the potential for prolonged neuromuscular paralysis and mechanical ventilation.<sup>[4]</sup> Circulatory collapse after administering succinylcholine in a patient with diffuse motor neuron disorder has been reported.<sup>[5]</sup> Conversely, resistance to atracurium and increased number of acetylcholine receptors has been demonstrated in patients with multiple sclerosis, who also have spastic paraparesis.<sup>[6]</sup> Peripheral nerves may also be involved in the form of axonopathy, neuronopathy, reduction of number of neurons in anterior horns and dorsal root ganglions.<sup>[7]</sup> This might cause postoperative neuropathies which may be attributed to the regional technique.

Unlike diseases like multiple sclerosis which exhibit a relapse pattern, spinocerebellar degeneration does not show a relapsing pattern.<sup>[8]</sup> There is no evidence of exacerbation of the pathological process in spinocerebellar degeneration after neuraxial anesthesia as judged from the recent case reports.<sup>[9,11]</sup>

Case reports of successful anesthetic management of patients with olivopontocerebellar degeneration in both general and regional anesthesia exists. Spinal anesthesia has been successfully administered in a patient of hereditary spastic paraplegia.<sup>[9]</sup> Administration of epidural labor analgesia to a patient of olivopontocerebellar degeneration has been reported.<sup>[10]</sup> Similarly, labor analgesia with epidural bupivacaine has been reported in spinocerebellar atrophy.<sup>[11]</sup> Schmitt *et al.*, reported use of rocuronium in two children with Friedreich's ataxia posted for scoliosis correction with no appreciable increase in onset and recovery timings.<sup>[12]</sup> Various atrioventricular blocks, attributed to myocardial degeneration and hypertonicity of the vagus nerve, have been reported after general anesthesia.<sup>[13]</sup> Kazunori and Zuigan reported two cases of spinocerebellar degeneration managed under regional anesthesia and propofol infusion uneventfully.<sup>[14]</sup>

Epidural Anaesthesia combined with general anesthesia has been administered for abdominal aortic aneurysm repair in a patient with spinocerebellar degeneration.<sup>[15]</sup> In spite of the concerns for aspiration, Laryngeal mask airway has been used uneventfully with propofol and nitrous oxide combination.<sup>[16]</sup> Administering regional anesthesia in a patient with autonomic dysfunction is controversial. If intravascular volume is maintained exaggerated hypotension after spinal or epidural-induced further autonomic blockade is unlikely in patients with preexisting autonomic neuropathy.<sup>[3,17]</sup> Denervation hypersensitivity to directly acting vasopressor amines may be present.<sup>[18]</sup> Our patient did not exhibit refractory hypotension after regional blockade (sensory level T8) as she did not exhibit other signs of autonomic instability. A combined spinal-epidural technique was preferred to provide adequate muscle relaxation and effective postoperative analgesia. Opioids were avoided due to the potential for delayed respiratory depression.

In conclusion, we successfully administered central neuraxial blockade to a patient with olivopontocerebellar degeneration. Regional anesthesia is a safe and acceptable choice for patients with spinocerebellar degeneration.

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