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## CLINICAL INFORMATION

# Tropical spastic paraparesis – anesthetic approach



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### Abstract

**Introduction:** HTLV-1 infection is endemic in Japan, Caribbean, Africa, and South America. It is transmitted from mother to child, sexual contact, blood transfusions, or sharing needles. Tropical spastic paraparesis (TSP) is a chronic degenerative neurological disease associated with this infection. It results from a spinal cord symmetrical degeneration at the thoracic level and is characterized by progressive motor weakness in the lower limbs, hyperreflexia, sensitivity changes, urinary incontinence, and bladder dysfunction.

**Clinical case:** Female, 53 years old, HTLV-1 infection and TSP. She had decreased strength in the lower limbs and hyperreflexia, paretic gait, spasticity, and neurogenic bladder symptoms, with recurrent urinary infections. She was scheduled for cystectomy. The patient was monitored according to standard ASA. Due to severe coagulopathy and the possibility of neurological worsening, epidural catheter was not placed. The induction of general anesthesia was performed with midazolam and fentanyl, followed by etomidate and cisatracurium. She was intubated with a tube size seven and maintained with desflurane and oxygen. Anesthesia was uneventful; the surgery lasted 1 hour and 50 min. There were no complications in the immediate postoperative period, during hospitalization, nor deterioration of the neurological examination. The patient was discharged 20 days later.

**Discussion/Conclusion:** There are reports of decreased electromyographic response and neurological deterioration associated with propofol in these patients, etomidate was used. The hepatic metabolism of rocuronium posed a risk, we chose to use cisatracurium. It was concluded that the anesthesia chosen did not affect the course of the disease.

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**PALAVRAS-CHAVE**

Human  
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 tropical

**Paraparesia espástica tropical – abordagem anestésica****Resumo**

**Introdução:** A infecção por HTLV-1 é endêmica no Japão, nas Caraíbas, na África e na América do Sul. A transmissão ocorre de mãe para filho, por contatos sexuais, transfusões de sangue ou partilha de agulhas. A essa infecção está associada uma doença neurológica degenerativa crônica, a paraparesia espástica tropical (TSP). Essa resulta de uma degeneração simétrica da espinal medula em nível torácico. Caracteriza-se por diminuição progressiva da força nos membros inferiores, hiperreflexia, alterações de sensibilidade, incontinência urinária e disfunção vesical. **Caso clínico:** Mulher de 53 anos, infecção por HTLV-1 e TSP. Apresentava diminuição da força nos membros inferiores e hiperreflexia, tinha uma marcha parética, espasticidade e sintomas de bexiga neurogênica com infecções urinárias de repetição. Foi proposta para cistectomia. Foi monitorada de acordo com o padrão da ASA. Devido à coagulopatia grave e à possibilidade de agravamento neurológico, não se colocou cateter epidural. A indução da anestesia geral foi feita com midazolam e fentanil seguidos de etomidato e cisatracúrio. Foi entubada com um tubo sete e mantida com desflurano e oxigênio. A anestesia decorreu sem intercorrências, a cirurgia terminou em uma hora e 50 minutos. Não houve quaisquer complicações no pós-operatório imediato, durante a internação, nem deterioração do exame neurológico. A doente teve alta 20 dias depois.

**Discussão/Conclusão:** Há relatos de diminuição da resposta eletromiográfica e deterioração neurológica associadas ao propofol nesses doentes, razão para uso de etomidato. A metabolização hepática do rocurônio representava um risco e se optou pelo cisatracúrio. Conclui-se que o plano anestésico escolhido não teve qualquer interferência no curso da doença. © 2015 Sociedade Brasileira de Anestesiologia. Publicado por Elsevier Editora Ltda. Este é um artigo Open Access sob uma licença CC BY-NC-ND (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

**Introduction**

The human T-lymphotropic virus 1 (HTLV-1) infection is endemic in some countries, notably in Japan, Caribbean Countries, and parts of Africa and South America. The prevalence increases with age and is higher in females.<sup>1</sup> It is transmitted from mother to child, by sexual contact, blood transfusions, or sharing contaminated needles. Two diseases are associated with this infection: adult T-cell leukemia/lymphoma (ATL) and tropical spastic paraparesis (TSP) – a chronic degenerative neurological disorder. TSP results from symmetrical degeneration of the spinal cord lateral columns at the thoracic level.<sup>2</sup> It is characterized by progressive motor weakness in the lower limbs and hyperreflexia, associated with autonomic dysfunction with urinary incontinence and bladder dysfunction. Changes in gait due to decreased strength in the lower limbs are the main symptom of disease. Autonomic dysfunction symptoms may precede, be concomitant, or be manifested later in disease progression. Noteworthy, the symptoms of neurogenic bladder, which results in high mortality rates, especially feeling of incomplete emptying, pollakiuria, urinary urgency, recurrent urinary tract infections, gallstones,

and even severe cases of chronic pyelonephritis or kidney failure.<sup>3</sup>

Unlike the multiple sclerosis (MS), the symptoms develop gradually, without periods of crisis and remission. Furthermore, there is no involvement of the cranial nerves and cognitive function. This syndrome develops in less than 1% of those infected by HTLV-1.<sup>1</sup>

Current knowledge of the anesthetic management of these patients results from clinical isolated rare cases. The aim of this paper is to present a case of TSP and expose its anesthetic approach.

**Case report**

Female patient, 53 years old, 60 kg, 158 cm, with TSP of several years of development and diagnosed seven years before, presented with complaints of decreased strength in the lower limbs for about 15 years and delayed urinary symptoms. Initially, she was diagnosed with multiple sclerosis, but due to the atypical clinical course and the presence of HTLV-1 antibodies in blood and cerebral spinal fluid (CSF), HTLV-1 infection was confirmed. Cystectomy was proposed.

At the time of surgery, the patient was neurologically stable. She had bilateral decreased strength in the lower limbs, hyperreflexia, and no sensitivity changes; walked with third-party support; had a paretic march, with spasticity; had no changes of cranial nerves or upper limbs. She also had symptoms of neurogenic bladder with recurrent urinary, an indication to undergo the surgery in question. Her personal history included infection with hepatitis C with thrombocytopenia ( $37 \times 10^9 \cdot L^{-1}$  platelets) and severe coagulation disorders (aPTT 38.6, TP 13.8), type 2 diabetes, and depressive syndrome. The remaining preoperative tests (analytical study, ECG, chest X-ray, and kidney function test) were normal.

## Anesthesia

Monitoring: ASA standards, BIS, and TOF.

Premedication: midazolam.

It was decided not to insert an epidural catheter due to patients' severe coagulopathy and the possibility of neurological worsening.

Induction: Fentanyl followed by etomidate and cisatracurium. Intubation with a tube size seven.

Maintenance: General anesthesia (GA) was maintained with desflurane and oxygen. Two additional bolus of cisatracurium were given according to TOF.

Induction and maintenance of anesthesia were uneventful: Due to technical difficulties, the surgical team chose not to perform the initially scheduled radical cystectomy. A cystostomy was performed, and the procedure lasted one hour and 50 min.

Recovery: The neuromuscular blockade was reversed with neostigmine and atropine and, five minutes later, the patient had 400 mL tidal volume, respiratory rate 12 bpm, and was wide awake, so she was extubated in the operating room. There were no complications in the immediate post-operative period, during hospitalization, or deterioration of the neurological examination. The patient was discharged 20 days later.

## Discussion and anesthetic considerations

Patients with tropical spastic paraparesis have numerous complications resulting from the disease progression; notably, infected scabs of prolonged recumbency, urinary retention due to sphincter dysfunction, and fractures resulting from peripheral neuropathy, which predominantly affects the lower limbs. They are, therefore, potential surgical candidates.

In recent years, and mainly the result of thematic review articles or case reports in endemic regions, we have sought to understand the implications and anesthetic needs of these patients, avoid exacerbations/crisis or post-surgical complications. Thus, the anesthetic management of these patients should take into account some special features.

Many of these patients are under daily corticosteroids,<sup>4</sup> and need to keep doses not below the usual during the perioperative period (hydrocortisone 200 mg in major surgery

and 100 mg in minor surgery) due to the risk of left ventricular dysfunction and refractory hypotension.

Classically, the neuraxial anesthetic approach is contraindicated in patients with active disease or active neurological symptoms. However, there are several reports of neuraxial anesthesia in patients with TSP, without deterioration of neurological symptoms, or accelerated disease progression.<sup>3</sup>

Some authors have reported that the induction doses of propofol ( $2-3 \text{ mg} \cdot \text{kg}^{-1}$ ) causes decreased electromyographic activity (about 20%) in patients with TSP, without, however, decreased muscle strength or neuronal conduction velocity.<sup>5</sup>

The choice of neuromuscular relaxant to use in such patients is also a subject of great controversy. The presence of a high number of muscle cholinergic receptors out of the neuromuscular plate on these patients greatly increases the sensitivity to acetylcholine. Thus, the use of depolarizing muscle relaxants, such as succinylcholine, carries a high risk of severe hyperkalemia (increase of about  $3 \text{ mmol} \cdot \text{L}^{-1}$ ) that may result in cardiac arrest. If necessary, the group of nondepolarizing muscle relaxants should be preferred, although these patients often exhibit a prolonged action. Thus, neuromuscular block monitoring is essential.

The choice of the technique and anesthetic protocol in this patient took into account the current recommendations for patients with known myelopathy. Neuraxial anesthesia was discarded due to coagulopathy and risks of exacerbation/disease progression. The selected drugs, notably inducing and relaxing drugs, minimize the risk of serious perioperative complications.

## Conclusions

Tropical spastic paraparesis is a rare neurological complication of HTLV-1 infection, is endemic in Japan, Colombia, Brazil and Caribbean countries. It is characterized by slow and progressive spasticity, hyperreflexia, muscle weakness mainly in the lower limbs, bladder disorders, and changes in sensitivity.

The management of these patients is challenging. Usually, general anesthesia is selected at the expense of neuraxial anesthetic techniques due to the possibility of disease exacerbation. The choice of the induction drug and especially the muscle relaxant (wherever possible, avoid the depolarizing relaxants) is essential to prevent major perioperative complications that prolong hospitalization and result in the progression of disease activity. Careful monitoring of the neuromuscular block is essential and indisputable.

## Conflicts of interest

The authors declare no conflicts of interest.

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