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Case report

Tropical Spastic Paraparesis and Anesthesia: Case Report and Topic Review

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ABSTRACT

Introduction: Tropical spastic paraparesis is an endemic infection in Colombia caused by the HTLV-1 retrovirus. It is characterized by a slow and progressive myelopathy that initially targets lower limbs. Complications such as eschars due to a prolonged decubitus, urinary retention to sphincter dysfunction, fractures, etc. make these patients potential surgery candidates.

Objective: To report a case and to review the physiopathology, epidemiology, clinical manifestations, treatment and basic anesthetic considerations of this disease.

Methods: Case report and topic review. The research included clinical trials, meta-analysis, practice guides, randomized controlled assays, revisions, case reports, classic articles, comparative studies, consensus conferences, magisterial classes and textbooks regarding published articles on tropicalsSpastic paraparesis/HTLV-1 (TSP/HAM) Associated myelopathy and anesthetic implications. Publications focused on etiology, physiopatology, epidemiology, clinical manifestations, treatment and anesthetic repercussions of TSP/HAM were included in this article. Research was carried out through PubMed, MdConsult, EBSCOhost, OvidSP, and Scielo, of articles in English and Spanish. The MeSH terms used were: *paraparesis, tropical spastic, anesthesia* and the DeCS terms were: *paraparesia espástica tropical, anestesia*. Titles and abstracts of articles identified in the database were studied independently.

Results: We describe the case of a male adult patient who was admitted to surgery for urethral reconnection after presenting a classic complication of tropical spastic paraparesis.

Research on the topic yielded 1829 studies. A total 20 writings met the inclusion criteria. We present implications regarding anesthesia and the disease.

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Paraparesia espástica tropical y anestesia: reporte de caso y revisión temática

Palabras clave:
Paraparesia espástica tropical
Enfermedades de la médula espinal
Anestesia
Rotavirus

RESUMEN

Introducción: La paraparesia pspástica tropical es una infección endémica en Colombia, causada por el retrovirus HTLV-1. Se caracteriza por una mielopatía de lenta instauración que compromete principalmente los miembros inferiores. Las complicaciones, como escaras por decúbito prolongado, retención urinaria por disfunción esfinteriana, fracturas, etc., hacen de estos pacientes candidatos quirúrgicos potenciales.

Objetivo: Reporte de caso y revisión temática de la fisiopatología, epidemiología, clínica, tratamiento, y de los aspectos anestésicos básicos de la enfermedad.

Metodología: Reporte de caso y revisión temática. Se incluyeron en la búsqueda ensayos clínicos, meta-análisis, guías para la práctica, ensayos controlados aleatorizados, revisiones, reportes de casos, artículos clásicos, estudios comparativos, conferencias de consenso, clases magistrales y libros de texto, de artículos publicados sobre paraparesia espástica tropical/mielopatia asociada al HTLV-1 (PET/MAH) e implicaciones anestésicas. Se incluyeron las publicaciones cuyo tema central fuese etiología, fisiopatología, epidemiologia, manifestaciones clínicas, tratamiento y repercusiones anestésicas de PET/MAH. Se realizó una búsqueda PubMed, MdConsult, EBSCOhost, OvidSP, y Scielo, de artículos en inglés y español. Se utilizaron los términos MeSH: *paraparesis, tropical spastic, anesthesia* y los términos DeCS: *paraparesia espástica tropical, anestesia*. Se estudiaron de forma independiente títulos y resúmenes de los artículos identificados en las bases de datos.

Resultados: Se describe el caso de un adulto masculino quien fue llevado a cirugia para reconexión uretral, después de presentar una de las complicaciones características de la paraparesia espástica tropical. La búsqueda arrojó 1.829 estudios. 20 escritos cumplieron con los criterios de inclusión. Se hace una presentación de implicaciones anestésicas y de la enfermedad.

Introduction

In the 32 years since its discovery,[1] little has been written on the HTLV-1 virus and its relation to procedures and drugs used in anesthesiology. Although most of the literature on TSP/HAM is Japanese, Colombia has enough motives not to remain indifferent. Even compared to Brazil and Peru, our country has the highest prevalence of this pathology among the thirteen South American countries. Most patients reach the appearance of complications and progressive dependence as a result of the natural course of TSP/HAM, ambiguity of the current established management protocols and little effectiveness of available medication. The aim of this article is to report a case of TSP/HAM and carry out a revision of physiopathology, epidemiology, manifestations, treatment and mostly, of the basic anesthetic aspects of this disease. This knowledge will hopefully provide safe anesthesia to these patients.

Methods

Study type: case report and topic review. The research included clinical trials, meta-analysis, practice guides, randomized controlled assays, revisions, case reports, classic articles, comparative studies, consensus conferences, magisterial classes and textbooks regarding published articles on tropical spastic paraparesis/HTLV-1 associated myelopathy (TSP/HAM) and anesthetic implications. Publications focused on etiology, physiopatology, epidemiology, clinical manifestations, treatment and anesthetic implications of TSP/HAM.

Intervention type: publications focused on etiology, physiopatology, epidemiology, clinical manifestations, treatment and anesthetic repercussions of TSP/HAM were included in this article.

Bibliographic research strategy: the online research was carried out through PubMed, MdConsult, EBSCOhost, OvidSP, and Scielo, of articles in English and Spanish.

Términos clave: se realizó la búsqueda con las palabras clave obtenidas del MeSH: *paraparesis, tropical spastic, anesthesia*. También se realizó con las palabras clave obtenidas del DeCS: *paraparesia espástica tropical, anestesia*.

Revision methods: titles and abstracts of the articles selected from the database were studied.

Data collection and analysis: the studies were assessed individually.

Results

Research on the topic yielded 1829 studies. A total 20 writings met the inclusion criteria. We present implications regarding anesthesia and the disease.

Case report

Male adult patient, 49 years of age, height: 5.9 ft (175 cm), weight: 80 kg, permanent inhabitant of the Colombian Caribbean (he denies temporal residence, even journeys into other parts of the country). The patient claims he began noticing weakness in his lower limbs fifteen years ago. Even though this weakness of the legs and thighs was initially overcome with the use of walking aids, such as canes, after seven years the symptoms evolved into paraplegia. In 2009 he was hospitalized due to a superinfection of a sacral eschar (as a result of a prolonged decubitus position). Together with muscle weakness, the patient began having difficulties when starting urination, which was managed with urethral catheterization. The repeated use of the probe led to narrowing of the urethra, so an advanced permanent catheterization was carried out by cystotomy. The patient was finally programmed to undergo urethroplastic procedure (urethral reconnection) via perineum. Other medical record data included: appendectomy 20 years before. Laboratory findings: base glucose: 96 mg/dl, BUN: 13 mg/dl, creatinine: 1 mg/dl, Hb: 13 mg/dl, hematocrit: 37%, platelets: 259 000/µl, leukocytes: 7700/µl, neutrophils: 58%, PT: 7,8 (9,45), INR: 0,82, PTT: 27 (28,7), total cholesterol: 195 mg/dl, TGD: 263 mg/dl, HDL: 35 mg/dl, LDL: 108 mg/dl. A stress ECG with dobutamine was performed and reported a 65% left ventricle ejection fraction and negative coronary insufficiency.

On the day of the procedure, the patient had a fine nutritional status and noticeable lower limb muscle atrophy. The vital signs were: blood pressure: 140/90 mmHg, heart rate: 78 bpm, respiration rate: 14 rpm, and temperature: 36,5 °C. The patient was admitted into the operating room and placed under monitoring (heart rate, blood pressure, pulse oximetry, capnography, cardioscopy), he was previously oxygenated at 100%, and began anesthesia with 80 mg IV lidocaine, 200 Mgr IV fentanyl, 160 mg of IV propofol and 10 mg IV cisatracurium. Intubation was carried out with a 8 mm internal diameter. Maintenance anesthesia was managed with 2-3% sevofluorane and 1 l/min O_2. Total anesthesia duration: 2h45min. Once the airway reflexes and spontaneous ventilation pattern is regained, the tube is removed and the patient is taken to the post-anesthesia recovery unit. No complications were reported.

Thematic review results

Tropical spastic paraparesis is a disease coused by the HTLV-1 retrovirus. It is also known as HTLV-1 associated myelopathy (HAM). The virus is associated with T cell lymphoma/leukemia;[1,2] in Japan and in the Caribbean gulf it is also linked to TSP/HAM.[3,4] HTLV-2 has been associated with polyneuropathies, myopathies and syndromes resembling TSP/HAM, but the evidence is not sufficient to establish a correlation between infection and disease. While Guessain found a high prevalence of anti-HTLV-1 antibodies in Martinica, Rodgers-Johnson & Gajdusek did the same in Jamaica and Colombia.[5] In other areas such as equatorial Africa, the Seychelles Islands, Central and South America and southern Japan the HTLV-1 is highly endemic.[1,5,6] In the Caribbean, the highest prevalence of the HTLV-1/2 virus in serum among the general population is in Dominica (38,6%); while in Colombia the prevalence is estimated between 0,73 and 4,3%, concentrated among indigenous tribes of Chibcha dialect and Africa-descendant people of the city of Tumaco.[6,7] In South America, only Brazil and Peru match the serum prevalence of HTLV-1/2 of Colombia.[7]

Nonetheless, infection of the HTLV-1 virus does not necessarily mean there will be TSP/HAM. In Kyushu, Japan, the prevalence of TSP/HAM has been estimated in as much as 8.6/100 000 among the general population and 68.3/100 000 for serum HTLV-1 positive individuals.[4] According to Kaplan, the incidence of TSP/HAM among infected individuals is around 3.1/100 000 people/year. According to this data; if we assume a person is infected with HTLV-1 at birth, the cumulative probability of actually developing TSP/HAM at the age of 75 is 0,25%.[1,4,8,9]

This is a disease of the upper motor neuron characterized by paraplegia or spastic paraparesis (97,8%),[3,4] bladder alterations (93,8%),[3,4] lower limb muscle weakness (88,2%),[3,4] altered sensitivity especially in thorax (56,2%),[3,4] and cerebellar ataxia (5-20%).[3,4,10] Kuriyama et al[11] reported alterations in sympathetic myocardial reflex activity are due to spinal cord compromise in thoracic vertebrae. The patient may also present back pain or symptoms such as burning, tingling or stinging sensations in lower limbs. Neuropathic pain is common in advanced stages of the disease.[6] The WHO has recommendations for identifying TSP/HAM based on clinical and laboratory criteria (www.who.int/). HTLV-1 is also associated to other diseases such as Sjögren syndrome, thiroyditis, artropathy, polimyositis, uveitis, and infection susceptibility such as tuberculosis, leprosy, strongyloidiasis and scabiosis.[1,4,6,12] The evolution of the disease is accountable for a great part of our patient's clinical course. Fortunately, up until the time of assessment, the patient showed no evidence of any of the previously mentioned infectious or autoimmune diseases.

The HTLV-1 was the first retrovirus that was ever discovered, as well as the first to be linked to illness in humans.[1] Transmission of the virus may occur during unprotected coitus,[1,4,13] hemocomponent transfusion, shared needle use, pregnancy and birth, and lactation.[1,4,6,13,14] The incubation period has been classically considered to last for years and even decades,[1,4] and 50% of cases associated to blood transfusion become ill 3,3 years after the transfusion.[4] However, a significant decrease has occurred in blood transfusion related cases since the implementation of donor blood laboratory testing.[1,4] Multiple cases in families have also been reported.[2,4]

The mechanism of the virus in lymphocyte proliferation (leukemia/lymphoma) in some cases and chronic inflammation (TSP/HAM) in others remains unknown.[5] The immunologic reaction beween Th1 type infected T CD4 lymphocytes and T CD8 lymphocytes would explain many of the disorders of TSP/HAM.[5] Pathologic findings show degeneration in the lateral columns of the spinal cord in its medium and lower parts,[15] as well as mononuclear infiltration

(mainly T lymphocytes[15]) and a marked axonal and myelin destruction[15] and astrocytic gliosis.[3] Atypical flower-shaped lymphocytes (resembling those found in lymphoma/leukemia) account for 1% of peripheral blood lymphocytes in half of patients with TSP/HAM.[4] The presence of HTLV-1 antibodies in blood and cerebrospinal fluid is confirmed with ELISA and Western blot,[1,4] while lymphocyte proviral DNA is detected through the PCR technique.[1] Determination of neopterin in cerebrospinal fluid is recommended, since it is a product of immune cell activation and is useful for differential diagnosis of chronic myelopathies in the same patient. MRI findings show periventricular and subcortical white matter alterations, together with high intensity signals and highlighting of posterior and lateral spinal cord columns.

Anesthetic implications

The pre-anesthetic assessment must consider the patient could currently be under immunomodulatory therapy with prednisone or anti-viral therapy with α interferon, β1a interferon, or daclizumab (IL-2 receptor blocker).[5,16] The classic Udelsman et al[17] report published in 1986 demonstrated that patients that take glucocorticoids must remain under treatment through the entire peri-operative period. It was shown that patients undergoing steroidal treatment will regain pituitary-adrenal axis functionality 9-12 months after treatment is concluded. These patients cannot take doses lower than those of their treatment plan due to a high risk of low cardiac ejection fraction volumes and refractory hypotension.[18] Roizen et al gave 200 mg/day IV phosphate hydrocortisone to a 70 kg adult for major surgery, and 100 mg/day IV phosphate to the same 70 kg for a minor procedure. The dose is decreased 25% every day until the patient tolerates oral administration and is able to resume the usual dose.[18]

It is often considered that the presence of a neurologic illness is a counterindication for neuroaxial anesthesia.[19] However, Sugimoto et al[14] reported epidural block with ropivacaine in a patient undergoing lung bullectomy and found no residual neurologic damage or acceleration during the procedure. In addition, Yuasa et al[20] found no neurologic deterioration or complications of the intra-operative course in a patient enduring subarachnoid block for a cystolithotomy procedure.

The type of surgery performed on our patient could have been approached with a conductive anesthetic technique; given that the case reports do not mention any complication in patients with TSP/HAM. Nevertheless, we chose a general technique, following more conservative recommendations of avoiding neuroaxial punctures in patients with myelopathies. Neurologic manifestations in these patients can be mistakenly thought of as a defective technique on our behalf rather than worsening or recurrence of the base condition.

Nitahara et al[8] reported propofol causes a 20% reduction of electromyographic response in a female patient with TSP/HAM, a rare event not reported in normal controls. Muscle strength and neuron conduction speed were not reduced. This is a crucial finding when analyzing diagnostic procedures in operating rooms for patients with TSP/HAM.[8]

The following issue is the proper type of muscle relaxant. Cholinergic muscle receptors outside the neuromuscular junction are especially numerous in denervated patients. Jointly with the myoneural plate receptors, these patients are particularly sensitive to acetylcholine. Consequently, the omnipresent risk of inducing hypokalemia after the use of muscle relaxants such as suxamethonium must always be considered with rigorous and utmost care in a patient with TSP/HAM. Potassium elevation can reach levels high enough to cause cardiac arrest. While serum potassium elevation in a regular patient does not generally exceed 0,5 mmol/L, it can reach 3 mmol/L in a patient with a neurologic illness associated with motor deficit.[19]

Non depolarizing muscle relaxants are used with extreme caution because of the potentially prolonged effect that may occur.[3] Muscle fasciculations immediately after pancuronium have been reported in a 6 year old patient with demyelinating disease.[19] Our choice of using cisatracurium was intended to avoid hyperkalemic peaks and their respective complications as much as possible.

Conclusions

TSP/HAM is a slow and progressive myelopathy caused by the HTLV-1 retrovirus. Among the complications of this illness are urinary retention due to sphincter malfunction, eschars due to prolonged time periods in the same position, urolithiasis and fractures.

The anesthesiologist must carefully review the medical record and assess the patient's nutritional status, hemodynamic and airway conditions. He/she must carry out the transition between regular treatment and the perio-operative treatment.

Patients with TSP/HAM can present severe complications when treated with depolarizing muscle relaxants and their use should be avoided if posible.

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Conflict of interests

None declared.

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