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ORIGINAL ARTICLE

Results of surgical treatment for juvenile myasthenia gravis☆

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Thymectomy;
Radical thymectomy;
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Abstract
Introduction: Radical or extended thymectomy is an effective treatment for myasthenia gravis in the adult population. There are few reports to demonstrate the effectiveness of this treatment in patients with juvenile myasthenia gravis.
Objective: The main objective of this study was to show that extended transsternal thymectomy is a valid option for treating this disease in paediatric patients.
Results: Twenty-three patients with juvenile myasthenia gravis underwent this surgical treatment in the period between April 2003 and April 2014; mean age was 12.13 years and the sample was predominantly female. The main indication for surgery, in 22 patients, was the generalised form of the disease (Osserman stage II) together with no response to 6 months of medical treatment. The histological diagnosis was thymic hyperplasia in 22 patients and thymoma in one patient. There were no deaths and no major complications in the postoperative period. After a mean follow-up period of 58.87 months, 22 patients are taking no medication or need less medication to manage myasthenic symptoms.
Conclusions: Extended (radical) transsternal thymectomy is a safe and effective surgical treatment for juvenile myasthenia gravis.

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PALABRAS CLAVE
Mediastino;
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Timectomía radical;
Timo

Resultados del tratamiento quirúrgico en la miastenia gravis juvenil

Resumen
Introducción: La timectomía radical ampliada para el tratamiento de la miastenia gravis es una opción efectiva en la población adulta. No ocurre lo mismo en el caso de la miastenia gravis juvenil, ya que existen pocos reportes que demuestren su efectividad.
Objetivo: El principal objetivo de esta investigación fue el de demostrar que la timectomía transesternal radical ampliada es una alternativa validada para el tratamiento de esta enfermedad en este grupo de pacientes.
Resultados: Con esta técnica fueron intervenidos 23 pacientes con miastenia gravis juvenil en el periodo comprendido entre abril del 2003 y abril del 2014. La edad media fue de 12,13 años y hubo un predominio en el sexo femenino. La principal indicación quirúrgica fue, en 22 pacientes, la forma generalizada de la enfermedad (estadio II de Osserman) sin respuesta al tratamiento médico luego de 6 meses. El diagnóstico histológico fue de hiperplasia tímica en 22 pacientes y timoma linfocítico tipo I en un paciente. No hubo fallecidos y no se presentaron complicaciones mayores en el periodo postoperatorio. Con un seguimiento medio de 58,87 meses, 22 pacientes se encuentran sin tratamiento o necesitando menor cantidad de medicamentos para el control de los síntomas miasténicos.
Conclusiones: La timectomía transesternal ampliada es una opción segura y efectiva para el tratamiento quirúrgico de la miastenia gravis juvenil.

Introduction

Juvenile myasthenia gravis (JMG) is an autoimmune disorder of unknown aetiology. In JMG, serum antibodies alter neuromuscular transmission when they bind to acetylcholine receptors located in the muscular membrane at the motor end plate. This results in premature muscle fatigue progressing to paralysis during muscle contraction. The incidence of myasthenia gravis (MG) during the first 18 years of life is 4 cases per 100 000 population. The most frequent forms are juvenile MG (18%), transient neonatal MG (1.5%), and congenital MG. JMG manifests with generalised myasthenia (47%), ocular myasthenia (43%), and myasthenic crises (10%). After 1 to 3 years, generalised myasthenia accounts for 80% of all manifestations, whereas ocular myasthenia drops to 20% and myasthenic crises, which only occur in the context of generalised mysathenia, decrease to 5%.[1–5] The thymus has been suggested as a possible site of origin given that 75% of patients older than 20 have thymic abnormalities, 85% display thymic hyperplasia with active germinal centres, and 15% have thymomas. In addition, thymectomy improves patients' outcomes in most cases. Acetylcholine receptors in thymic myoid cells may act as autoantigens and trigger an immunological reaction at the level of the thymus. Thymic $CD4^+$ T-cells stimulate serum B-cells, which in turn will start producing antibodies against acetylcholine receptors.[6,7]

JMG is pathophysiologically heterogeneous. In most cases, it is associated with presence of acetylcholine receptor antibodies. However, it has also been reported to be associated with anti-MuSK antibodies and some cases are considered 'seronegative' since no antibodies can be detected using currently available techniques. Therefore, diagnosis of the disease is not based solely on antibody detection. The patient's clinical profile and such neurophysiological studies as repetitive nerve stimulation and especially jitter analysis play an important role in diagnosing the disease. Pharmacological tests are also useful. Treatment for JMG is symptomatic and aetiological. In patients with generalised involvement and incomplete response to treatment, a wide range of therapeutic measures are applied to halt the autoimmune response, including thymectomy, immunosuppresant agents, plasmapheresis, and immunoglobulins.[6,8–14] Despite the controversy surrounding treatment for MG, surgery has been shown to be superior to medical treatment alone, even in cases of JMG.[12,13,15–22] The main purpose of our study was to confirm the validity of radical or extended transsternal thymectomy for the treatment of JMG.

Patients and methods

Study characteristics

We conducted a prospective non-experimental study including 23 patients diagnosed with JMG who underwent extended thymectomy between April 2003 and April 2014. The study was conducted in the cardiac surgery department at Cardiocentro Ernesto Guevara and the Neurology Department at Hospital Pediátrico Provincial, in Santa Clara, Cuba.

Procedure

We used the criteria by Cheng et al.[21] for diagnosing JMG, indicating extended thymectomy, and assessing patients' outcomes; the clinical criteria by Osserman and Genkins[14]