CLINICAL CASE

Urologic repercussions of Shy-Drager syndrome

V.M. Telich\textsuperscript{a}, C.C. Merayo\textsuperscript{a}, S.E. Cartagena\textsuperscript{b,\ast}, A.C. González\textsuperscript{b}, O.G. Zubieta\textsuperscript{b}, M.J. Garzón\textsuperscript{b} and A.A. Flores\textsuperscript{c}

\textsuperscript{a} Department of Urology, Hospital Ángeles Pedregal, Mexico City, Mexico
\textsuperscript{b} General Surgery, Department of Urology, Hospital Ángeles Pedregal, Mexico City, Mexico
\textsuperscript{c} Department of Urology, Instituto Mexicano del Seguro Social, Department of Urology, Hospital Ángeles Pedregal, Mexican School of Medicine, ULSA, Mexico City, Mexico

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Abstract

Background: Shy-Drager syndrome, also known as multiple system atrophy, is a neurodegenerative entity of autoimmune etiology. Clinical symptoms encompass Parkinsonism and cerebellar and autonomous symptomatology, including urinary and sexual alterations and cortical dysfunction of the pyramidal pathway. There is little drug response. Incidence is similar by sex, disease onset is in adulthood, and it is a progressive pathology with a poor prognosis.

Case 1: We present herein the case of a 39-year-old woman that presented with signs of neurocardiogenic syncope and dysautonomia 5 years prior to her present hospital admission. That evaluation protocol showed data of sinus bradycardia, treated with a pacemaker, as well as gastric intolerance and urinary retention. Due to the insidious progression of her illness, the clinical data, and auxiliary diagnostic studies, the diagnosis of Multiple System Dysautonomia, or Shy-Drager syndrome, was made. In relation to urology, the patient presented with recurrent urinary infections and urinary retention on numerous occasions. A urodynamic study was performed, but was inconclusive due to a dysautonomic event during the study. In the filling cystomanometry, storage was 700 cc, but the patient was unable to micturate. Given the above, long-term management with clean intermittent catheterization was decided upon, but it could not be achieved due to another dysautonomic event, and therefore a transurethral catheter was left in place. The patient sought an alternative, because she wanted to continue to have an active sex life and the urethral catheter caused episodes of intense dyspareunia and dysautonomia. Thus, the joint decision was made to perform cystostomy, which the patient retains today.

Aim: To report on the urologic implications and management in a patient with Shy-Drager syndrome.

Materials and methods: The case record of a patient diagnosed with Shy-Drager syndrome was employed and she was treated for urologic complications at the Hospital Ángeles Pedregal.

Results: Following medical-surgical management, the patient was released to her home 3 days after her hospital admission due to clinical improvement. Currently she is being monitored as an outpatient.
Resumen
Antecedentes: El síndrome de Shy-Drager, también conocido como atrofia sistémica múltiple, es una entidad neurodegenerativa, de etiología autoinmune. El cuadro clínico abarca sintomatología parkinsoniana, cerebelosa y autonómica, incluyendo alteraciones urinarias y sexuales, así como disfunción córtico-vertebral. La incidencia por género es similar, con edad de inicio en la adultez, su evolución es progresiva y relacionada con mal pronóstico.

Exposición del caso: Una mujer de 39 años, quien 5 años antes de su ingreso actual presentó datos de síncope y disautonomía neurocardíogenica, por lo que se le realizó protocolo de estudio, mostrando datos de bradicardia sinusal, tratada mediante marcapasos, así como intolerancia gástrica y retención urinaria. Debido a su evolución de carácter insidioso, así como a los datos clínicos y de auxiliares diagnósticos, se diagnostica la presencia de disautonomía sistémica múltiple o síndrome de Shy-Drager. En el rubro urológico presenta infecciones urinarias de repetición, así como retención urinaria en múltiples ocasiones. Se realiza estudio de urodinamia, durante el cual presenta evento de disautonomía, por lo que se deja inconcluso; en la cistomanometría de llenado almacena 700 cc, sin poder realizar micción. Debido a lo anterior, se decide tratamiento a largo plazo mediante la realización de cateterismo limpio intermitente, que no logra realizarse debido a un nuevo evento de disautonomía, por lo que se deja sonda uretral a derivación. La paciente acude en busca de alternativas, ya que desea continuar con vida sexual activa, y debido a la presencia de la sonda presenta episodios de despareunia intensa y disautonomía, por lo que se decide en conjunto la realización de cistostomía, la cual porta actualmente.

Objetivo: Reportar la presencia de implicaciones urológicas en una paciente con síndrome de Shy-Drager, así como su manejo.

Material y métodos: Se empleó el expediente de una paciente con diagnóstico documentado de síndrome de Shy-Drager, quien fue tratada por complicaciones urológicas en el Hospital Ángeles Pedregal.

Resultados: Después del tratamiento médico-quirúrgico, la paciente es egresada a su domicilio 3 días después de su internamiento, debido a su mejoría clínica. Actualmente, es vigilada de forma ambulatoria.

Discusión: El síndrome de Shy-Drager fue descrito por primera vez en 1960 por Milton Shy y Glenn Drager. Se ha descrito clásicamente como la presencia de hipotensión ortostática, anhidrosis y disfunción cerebelosa y/o parkinsoniana de magnitud variable. Las alteraciones en el vaciamiento vesical y la disfunción eréctil son las afecciones urológicas predominantes. En el caso actualmente presentado, el rubro cardiovascular había sido tratado previamente y en este momento se realiza el tratamiento de las complicaciones urológicas, de manera satisfactoria. Las alteraciones en el vaciamiento vesical y la disfunción eréctil son las afecciones urológicas predominantes. En el caso actualmente presentado, el rubro cardiovascular había sido tratado previamente y en este momento se realiza el tratamiento de las complicaciones urológicas, de manera satisfactoria.

Conclusiones: Se ha observado la presencia de síntomas urológicos en el 60% de los pacientes que padecen esta enfermedad. Desafortunadamente, no existe terapéutica efectiva, por lo que el objetivo es controlar la sintomatología, principalmente relacionada con disautonomía térmica y cardiovascular. La calidad de vida de estos pacientes es pobre, y su pronóstico, muy sombrío. El manejo de las complicaciones es relativamente complicado y con resultados poco satisfactorios. Estos pacientes generalmente derivan en una incompetencia esfinteriana, por lo que la meta frecuentemente consiste en facilitar el almacenamiento urinario.
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Introduction

Shy-Drager syndrome, also known as multiple systemic dysautonomia, is an autoimmune neurodegenerative disease. Symptoms include Parkinsonism and cerebellar and autonomous symptomatology, as well as urinary and sexual alteration, and cortical dysfunction of the pyramidal pathway. Drug response is low. Incidence by sex is similar, age of onset is adulthood, and disease progression is progressive and related to poor prognosis.

The neurologic lesions characteristic of the disease are consistent with disseminated and diffuse gliosis, which explains the early and severe urinary symptomatology. The main affected areas are the cerebellum, substantia nigra, globus pallidus, caudate nucleus, putamen, inferior olives, mediolateral tract of the spinal cord, and Onuf's nucleus.

Case report

A 39-year-old woman presented with data of neurocardiogenic syncope 5 years before her present hospital admission, the study protocol for which revealed signs of sinus bradycardia, treated through pacemaker placement, and gastric intolerance and urinary retention. Due to the insidious progression of her illness and the clinical data and auxiliary diagnostic studies, the patient was diagnosed with multiple systemic dysautonomia, or Shy-Drager syndrome. Her urologic manifestations were recurrent urinary infections and urinary retention on numerous occasions. A urodynamic study was inconclusive because while it was being done the patient had a dysautonomic event. Storage was 700 cc in the filling cystomanometric study, but the patient was not able to micturate. Clean intermittent catheterization was therefore decided upon for long-term management, but it was not achieved due to a new dysautonomic event, and so a transurethral catheter was placed. The patient came to us in search of alternatives, given that she wished to continue having an active sex life, and the catheter caused her to have intense dyspareunia and dysautonomia. Thus, the joint decision was made to perform cystostomy and the patient continues to be managed with it today.

Discussion

Shy-Drager syndrome was first described in 1960 by Milton Shy and Glenn Drager. Its classical description is the presence of orthostatic hypotension, anhidrosis, and cerebellar dysfunction and/or Parkinsonism of varying magnitude. The predominant urologic alterations are related to bladder voiding and erectile dysfunction.

Classification:

• Parkinsonism: rigidity, bradykinesia, tremor
• Cerebellar dysfunction: loss of balance or stability and a tendency to fall down
• Mixed: symptomatology of both Parkinsonism and cerebellar dysfunction


Physical examination can reveal other data, such as atrophy of the iris, paralysis of the intra and extraocular musculature, the presence of altered myotatic reflexes, and muscle mass loss.

Urologic symptoms are observed in 60% of the patients that present with said pathology; 73% of them have urinary incontinence and 19% urinary urgency. Sixty-six percent of the patients present with post-void residual volume of 100-450 cc.

Exacerbated detrusor muscle activity is a frequent finding in these patients, as is pelvic floor dysfunction, which is related to alteration at distinct levels of the autonomous innervation. As the disease progresses, difficulty for initiating and maintaining bladder voiding is frequent, which can be related to pontine and sacral dysfunction.

Cystourethrography and urodynamic studies usually reveal the presence of an open internal bladder sphincter, related to denervation data of the striated sphincter determined through electromyography.

Analysis

Unfortunately, there is no effective therapy and so the aim is to control the symptomatology mainly related to thermal and cardiovascular dysautonomia. The quality of life of these patients is poor and the prognosis is bleak. The management of complications is relatively complex, and results are not very satisfying. These patients generally end up presenting with sphincteric incompetence and therefore the goal is to facilitate urinary storage.

Survival after the triggering of symptoms is 7-10 years. Patient survival of 15 years or more after symptom onset is exceptional. The main cause of death in these patients is pneumonia, but arrhythmias also account for an important portion of deaths. The triggering factor is still unknown and thus there are no known measures of prevention.

Ethical responsibilities

Protection of persons and animals. The authors declare that no experiments were performed on humans or animals for this study.

Data confidentiality. The authors declare that no patient data appear in this article.

Right to privacy and informed consent. The authors declare that no patient data appear in this article.
Table 1  Diagnostic criteria for Shy-Drager syndrome according to the American Autonomic Society/American Academy of Neurology

<table>
<thead>
<tr>
<th>Domain</th>
<th>Criterion</th>
<th>Symptom</th>
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<tbody>
<tr>
<td>Autonomic and urogenital</td>
<td>Orthostatic decrease &gt; 30 mmHg of systolic pressure or &gt; 15 mmHg of diastolic pressure</td>
<td>Orthostatic hypotension</td>
</tr>
<tr>
<td>Parkinsonism</td>
<td>Bradykinesia + rigidity or postural instability or tremor</td>
<td>Bradykinesia R rigidity</td>
</tr>
<tr>
<td>Cerebellar dysfunction</td>
<td>Gait ataxia + ataxic dysarthria or ataxia of an extremity or prolonged evoked nystagmus</td>
<td>Gait ataxia Ataxic dysarthria</td>
</tr>
<tr>
<td>Corticospinal tract dysfunction</td>
<td>Undefined</td>
<td>Extensor plantar response with hyperreflexia</td>
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Conflict of interest

The authors declare that there is no conflict of interest.

References