A Retrospective Case Series of and Proposed Framework for Palliative Care Discussions in Patients with Multiple Systems Atrophy

A.M. Dayal, M.E. Jenkins, M.S. Jog, K. Kimpinski, P. MacDonald, T.E. Gofton
London Health Sciences Centre, Department of Clinical Neurological Sciences
Western University, London, Ontario, Canada

Background

- Multiple Systems Atrophy (MSA) is a rare neurodegenerative disease of the central and autonomic nervous system, with a mean survival of 7-10 years.1
- It has a prevalence of 5/100,0002.
- There is no cure for MSA and it is purely symptomatic.
- Stridor is a major complication that develops in 30% of cases.2
- Case reports indicate that untreated stridor is a marker of short survival and may lead to obstructive sleep apnea, acute laryngeal obstruction and death.
- Tracheostomy is the optimal treatment for stridor although required in a minority of cases.3
- Respiratory failure arising from stridor can present acutely, and may require a tracheostomy as a life-supportive measure.

Objectives

- To propose a framework for advance care planning and palliative care discussions in MSA.
- To evaluate advanced care planning and current practices in palliative care in MSA.
- To propose a framework for advance care planning and palliative care discussions in MSA.

Methods

- A retrospective, non-concurrent cohort study exploring the symptom burden of patients living with MSA and the palliative care discussions that take place during the disease trajectory.
- Inclusion criteria:
  - Adult patients with a clinical diagnosis of MSA, meeting the criteria for a clinical diagnosis of probable MSA (Gilman et al. 2008).
- Known to inpatient or outpatient neurology services at London Health Sciences Centre (LHSC) between January 2004 and January 2014.
- Exclusion criteria:
  - Patients with Parkinsonian syndromes of uncertain etiology or with a diagnosis of an alternate Parkinson’s plus syndrome.
- Data collection:
  - Patients were identified from the clinical practices of all movement disorder and neurology centres.
  - Electronic medical records and paper charts for all visits to LHSC were reviewed.
- Data Analysis:
  - Data on common clinical symptoms were collected.
  - Content of palliative care discussions documented in the patient’s medical record were recorded:
    - diagnosis, symptom management, prognosis, goals of care, comfort measures and decision making regarding tracheostomy.
    - cases with no evidence of documented discussions in the medical record “unknown” was used.
- Sources of bias:
  - In order to reduce bias, only data documented in the patients’ charts were included in the analysis.
- Statistical analysis:
  - Due to the small number of patients included and the descriptive nature of the data, detailed statistical analysis was not performed.

Results

- Table 1 Characteristics of patients with Multiple Systems Atrophy seen in outpatient neurology clinics or admitted under inpatient services at London Health Sciences Centre from January 2004 to January 2014

<table>
<thead>
<tr>
<th>Total MSA (%)</th>
<th>Those with Stridor</th>
</tr>
</thead>
<tbody>
<tr>
<td>All Patients</td>
<td>22 (100)</td>
</tr>
<tr>
<td>Female</td>
<td>12 (56%)</td>
</tr>
<tr>
<td>Male</td>
<td>10 (45%)</td>
</tr>
<tr>
<td>Age at diagnosis</td>
<td>63.1 ± 8.7</td>
</tr>
<tr>
<td>Age</td>
<td>65.5 ± 7.9</td>
</tr>
<tr>
<td>Duration of disease (in years)</td>
<td>6.8 ± 3.6</td>
</tr>
<tr>
<td>Decreased</td>
<td>6 (27%)</td>
</tr>
</tbody>
</table>

- Table 2 End of life discussions: The focus and timing of end of life discussions

<table>
<thead>
<tr>
<th>N=22</th>
<th>Those with Stridor</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tracheostomy</td>
<td>1 (4%)</td>
<td>0</td>
</tr>
<tr>
<td>Tracheostomy</td>
<td>6 (27%)</td>
<td>2 (9%)</td>
</tr>
</tbody>
</table>

- Table 3 End of life discussions: Decreased patient demographics

<table>
<thead>
<tr>
<th>N=22</th>
<th>Those with Stridor</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total deceased</td>
<td>6 (27%)</td>
<td>2 (9%)</td>
</tr>
<tr>
<td>Time from admission to death (days)</td>
<td>28 ± 18.2</td>
<td>24 ± 30.4</td>
</tr>
<tr>
<td>Time from palliative care discussion to death (days)</td>
<td>95 ± 9.9</td>
<td></td>
</tr>
<tr>
<td>Time from DNR to death</td>
<td>2 (13%)</td>
<td>0</td>
</tr>
<tr>
<td>Total DNR</td>
<td>3 (50%)</td>
<td>2 (10%)</td>
</tr>
</tbody>
</table>

Conclusion

- Physicians effectively engaged patients in discussions regarding diagnosis, symptom management and prognosis in MSA, but they were less thorough regarding discussions for DNAR and end of life plans.
- End of life discussions were initiated very late in the disease trajectory.
- Timing was variable for those who underwent tracheostomy.
- Limited documentation outlining the content of all palliative care discussions in the medical record.
- No standard approach to advanced care planning and palliative care discussions in patients living with MSA.
- Despite these unknowns, early discussions will allow people with MSA to make treatment decisions that align with their goals of care prior to an emergency situation.
- Advanced care planning has been shown to improve outcomes with respect to the dying process.4
- A systematic approach to palliative care in MSA would enable patients and substitute decision makers to make informed health care decisions, throughout the disease trajectory.

Proposed Framework

- Early diagnosis and early discussions of goals of care, comfort measures and decision making regarding tracheostomy.
- Including a realistic assessment of patients quality of life and value of tracheostomy as supportive measure.
- Little evidence to guide physicians in their approach to care in MSA.

References


Disclosures

The authors, A.M. Dayal, M.E. Jenkins, M.S. Jog, K. Kimpinski, P. MacDonald, and T.E. Gofton, have no disclosures or conflicts of interest to declare.