AMYOTROPIC LATERAL SCLEROSIS, FROM DIAGNOSIS UNTIL DEATH: HOW DOES PALLIATIVE CARE FIT INTO THE JOURNEY OF MR. JOHNS

Heron Warren, CRNP, ACHPN
Erie VA Medical Center

OBJECTIVES

• Attendees will be able to:
  • Verbalize a basic understanding of amyotrophic lateral sclerosis.
  • Verbalize an understanding of the progression of amyotrophic lateral sclerosis along the chronic disease trajectory.
  • Verbalize an understanding of palliative care.
  • Identify specific changes in someone with amyotrophic lateral sclerosis that would meet criteria for the person to be appropriate for hospice care.

CASE STUDY

• Mr. Johns is a 72 year male with a past medical history of:
  • Hypertension
  • Coronary artery disease, status post stent x1
  • Prostate cancer, status post radiation treatment
  • Gastroesophageal reflux disease (GERD)

Social history:
  • Married for 44 years
  • Three adult children
  • Retired mechanic
  • Smoked cigars for 10 years, stopped five years ago
  • Likes an occasional beer on the weekends.
CASE STUDY
Mr. Johns arrives at your office complaining of:
• Symptoms over the last year
  • Weakness affecting control of head and neck
  • Forgetfulness
  • Jaw locking
  • Awkwardness, primarily affecting the R arm
  • Has had falls
  • Family has noticed some issues with speech
  • His wife reports he has lost weight
  • Additional information: Has not seen a medical provider for any of his concerns

EXAM AND TESTING
• Physical exam
  • Vital signs within normal limits.
  • Neck flexor and extensor weakness
  • Mild asymmetrical weakness of proximal upper extremities
  • Brisk muscle stretch reflexes
  • Tongue Fasciculations
  • Testing:
    • EMG showed widespread acute & chronic motor axon loss in upper extremities, tongue & thoracic para-scapal muscles.
    • CT head shows no acute process. Small lacunar infarct in the L caudate head.
  • Differential diagnosis and testing are indicative of a motor neuron disease

DIFFERENTIAL DIAGNOSIS
• Spectrum of motor neuron disease:
  • Progressive muscular atrophy
  • Primary lateral sclerosis
  • Progressive bulbar sclerosis
  • Flail arm syndrome
  • Flail leg syndrome
  • ALS plus syndrome
  • Amyotrophic lateral sclerosis (ALS)
AN EXAMPLE OF EMG TESTING IN ALS

SPECTRUM OF MOTOR NEURON DISEASE
- Amyotrophic lateral sclerosis (ALS) is the most common form of motor neuron disease; the pathology includes upper and lower motor neurons.
- Based on testing and physical exam results, the diagnosis of ALS is made.

WHAT HAPPENS TO THE NERVE CELLS
**ALS AN INEVITABLE DECLINE**

**Overview of ALS**
- Causes muscle weakness, disability, and eventually death
- Another name for this is Lou Gehrig’s disease
- Presently incurable
- Management is sporadic
- ALS has an annual occurrence of 1-3 cases per 100,000
- Prior to age 65 or 70, ALS is found to be higher in the male population than female
- Around 90% or more of cases are sporadic
- 70% of those affected die within 3 years of onset

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**WHAT IS ALS**
- ALS is a progressive degenerative condition which results in the loss of physical function due to the degeneration of upper and lower neurons
- Onset classified as being either bulbar, respiratory, or spinal
- Patients can present with cognitive and/or behavioral impairment
- Presentation
  - Asymmetric limb weakness (most common, around 80%)
  - Upper extremity with hand weakness
  - The “split hand syndrome”
  - Lower extremity weakness (presents with foot drop)
  - Bulbar involvement with dysarthria or dysphagia (20% at presentation)
ALS SPECIFICS

- How does it affect the upper motor neurons (UMN)?
  - Slowness of movement
  - Akimobility & stiffness
  - Hyperreflexia
  - Spasticity resulting from the degeneration of the frontal motor neurons
    - Indicated by abnormal reflexes and clonus

UPPER MOTOR NEURON SYMPTOMS

- Slowness of movement, lack of coordination, and stiffness are the result of loss of UMN's
- How are the arms and hands affected
  - Poor dexterity
  - Difficulty in performing ADL's
- How are the legs affected
  - Spastic gait
  - Poor balance
  - May include random leg flexor spasms and ankle clonus

UPPER MOTOR NEURON SYMPTOMS

- Most common bulbar UMN symptoms
  - Dysarthria
    - Produces a strained vocal tone
  - Dysphagia
    - The swallowing muscles become slow and uncoordinated
      - May lead to choking and coughing
  - Pseudobulbar affect
    - Manifests early on or can develop throughout the disease
    - Inappropriate laughing, crying, or yawning
UPPER MOTOR NEURON SYMPTOMS

- Most common bulbar UMN symptoms
  - UMN bulbar dysfunction can also result in laryngospasm
    - Aspirating liquids, food particles or saliva
    - Described as a squeezing feeling in their throat
    - Often accompanied by difficulty speaking (audible stridor)
    - Impaired inspiration
  - Increased nauseate tone
  - Difficulty opening the mouth
  - Trismus
  - Involuntary clenching of jaw
  - Biting the sides of cheeks and tongue
  - Balance problems and stiffness
  - Slurring speech

WHAT IS ALS CONTINUED

- How does it affect lower motor neurons (LMN)
  - Weakness
  - Atrophy or amyotrophy
  - Fasciculations
    - Resulting from the degeneration of LMNs in the brainstem and spinal cord producing muscle denervation.
  - Cognitive symptoms
  - Autonomic symptoms
  - Parkinsonism and supranuclear gaze palsy

LOWER MOTOR NEURON SYMPTOMS

- Loss of LMNs has many implications
  - Weakness
  - Atrophy
  - Fasciculations
  - Muscle cramps
LOWER MOTOR NEURON SYMPTOMS

- Affects of weakness
  - Hands
    - Difficulty operating small objects such as coins, buttons, zippers
    - Difficulty using pens and pencils.
  - In the proximal arm there is difficulty elevating the arm to the level of the chin or above their head.
  - Creates issues with dressing, grooming, bathing and eating.
  - Ankles and feet
    - Results in a slapping gait, tripping and falling.
  - In the proximal leg there is now difficulty climbing stairs, getting off the floor and rising from chairs.
  - Balance may now adversely be affected.

LOWER MOTOR NEURON SYMPTOMS

- Weakness cont.
  - Upper face
    - The eye may be unable to completely close
  - Lower face
    - Poor to no seal
    - Drooling
    - In patients that already have difficulty swallowing, sialorrhea
  - Masseter
    - Difficulty opening mouth
    - Difficulty moving the jaw from side to side
  - Extraocular motor neurons aren't usually affected until very late in the disease process.

LOWER MOTOR NEURON SYMPTOMS

- Weakness cont.
  - Trunk and spine
    - Unable to hold the head up
    - Difficulty maintaining erect posture
  - Abdominal protuberance
  - Diaphragm
    - Progressive dyspnea
    - Dyspnea at rest and with talking
  - Reduced vocal volume
  - Orthopnea
  - Sleep-disordered breathing
LOWER MOTOR NEURON SYMPTOMS

- Dysarthria and dysphagia result from LMN damage as well
  - Dysarthria
    - Can result from weakness of the tongue, lips or palate
    - Speech is slurred
    - May now have a nasal quality
    - Vocal cord weakness can result in hoarseness
  - Dysphagia
    - Results from tongue weakness
    - Disruption of the oral/consensual phase
    - Can lead to swallowing
    - Results from pharyngeal or esophageal weakness
    - Disruption of the pharyngeal phase of swallowing
    - May result in choking, nausea or vomiting
    - Aspirations may occur
    - Could be a combination of both previous factors

CASE STUDY

- Mr. Johns is seen in your office six weeks later to review findings and diagnosis
- Mr. Johns has had a recent fall and now is using a cane.
- His voice tone quality has decreased.
- He reports trembling in his right hand and has difficulty with some care needs.
- His wife voices concerns as she feels he is getting worse and wants to know "what are you going to do?"

PLAN OF ATTACK

- Refer to neurology (who confirms diagnosis)
- Meet with the multidisciplinary ALS team who review-
  - Timely use of gastrostomy tube
  - Respiratory assist, non-invasive ventilation, tracheotomy and mechanical ventilation
  - Therapies-OT, ST, PT
  - Social work services and counseling
  - Diet and nutrition
  - Medications: Riluzole, vitamins and supplements. Other medications will depend on symptoms and may include analgesics, anti-spasmodics, sleep aids, anti-cholinergics, stimulants, & laxatives.
TREATMENT DECISIONS

Mr. Johns has agreed to:
- PT, OT, ST
- He chooses Riluzole and B complex vitamin supplement
- They meet with the dietician and start drinking Boost daily

He and his wife meet with social work who assisted Mr. Johns in completing an advanced directive. He decides to be a DNR. He does not currently see a need to address the feeding tube concern or issues related to potential respiratory compromise.

His wife is pleased that social work will be able to assist with some concerns and help provide support. Social work has provided information on support groups.

CASE STUDY

- It has been 3 months since his diagnosis, he has completed a course of PT, OT & ST
- He has come to your office for a follow-up
- He remains on Riluzole
- New complaints:
  - He has noted episodes of coughing during eating; on one occasion his wife had to perform the Heimlich maneuver.
  - He has not been sleeping well.
  - At times he feels it is hard to get a full breath in and noted trouble breathing when laying flat in bed.
  - He had fallen and was seen in the local ER; outside of a hip contusion he had no other acute injuries.

CLINICAL SYMPTOMS AND SIGNS

- The primary clinical symptoms and signs result from the loss of motor neurons
- Differences in cranial, cervical, thoracic, or lumbosacral sites, speed that it spreads, pattern, and the gradient of upper and lower motor neuron dysfunction result in a disorder that is extremely variable among those affected by this disease.
- 20% of patients will have onset in the bulbar segment, which most often represents with either dysphagia or dysarthria.
- Patterns of ALS onset which may occur less often:
  - Respiratory muscle weakness 1-3%
  - Widespread weakness in the limbs and bulbar muscles 1-9%
  - Head drop or truncal extension weakness
  - Weight loss
    - Cramps, muscle atrophy, and fasciculation
CLINICAL SYMPTOMS AND SIGNS CONT.

- Initial presentation
  - May occur in any body segment
  - May manifest as upper or lower motor neuron symptoms or signs
  - The most common presentation of ALS, about 80%, comprises of asymmetric limb weakness.
  - Upper extremity onset is usually hand weakness that may begin in the shoulder girdle muscles.
  - Split hand syndrome
    - Frequent pattern of atrophy and weakness that "largely involves the median- and ulnar-innervated lateral hand intrinsic muscles with relative sparing of the medial muscles".
  - Lower extremity onset of ALS usually begins with weakness with foot drop while the close pelvic girdle onset is less common.

COGNITIVE SYMPTOMS

- There is a well-known link between ALS and frontotemporal executive dysfunction that may precede or trail the onset of upper and/or lower motor neuron dysfunction.
  - Frequency of the dysfunction varies from 28%-100%
  - Cognitive impairment was found in 35-51% of patients
  - Generally most patients do not have obvious dementia
  - Patients that met the criteria for frontotemporal dementia was only 15%

AUTONOMIC SYMPTOMS

- Symptoms may occur as the disease progresses
- Constipation occurs regularly
  - Multifactorial
  - Delayed colonic mobility
  - Dehydration due to dysphagia for thin liquids
- Early satiety
  - Breathing-wasting from delayed gastric emptying
  - Frequent urination without incontinence is common
  - Patients have also made complaints of excessive sweating
SENSORY SYMPTOMS
• Occurs in 20-30% of patients
• Tingling parasthesia
• Rarely have complete sensory loss
• Electrophysiologic studies demonstrated reduction of amplitudes on sensory nerve transmission

PARKINSONISM AND SUPRANUCLEAR GAZE PALSY
• Parkinsonism may follow or precede the upper and lower neuron symptoms
  • Facial masking
  • Tremor
  • Bradykinesia
  • Postural instability
• Supranuclear gaze palsy
  • Vertical gaze palsy

PATTERNS OF PROGRESSION
• Relentlessly progressive disorder with a linear slope
• Rate of progression is variable in individuals
• Progression worsens over time without remission or exacerbations
• Relatively predictable pattern
  • Unilateral arm to contralateral arm to ipsilateral leg, then contralateral leg and then bulbar muscles
  • Unilateral leg to contralateral leg to ipsilateral arm, then contralateral arm and then bulbar muscles
  • Bulbar onset spread to one arm then to contralateral arm
CASE STUDY

- Mr. Johns continues to have decline, it has now been 6 months since his diagnosis and he is being seen in your clinic for follow-up.
  - Noted decline in ambulatory function, now occasionally using a manual wheelchair.
  - One hospitalization after a fall resulting in debility. During this hospitalization he was noted to have increased dysphagia. A swallow test showed that the swallow mechanism was slowed, with testing of nectar consistant liquids there was penetration to the level of the vocal cords with partial clear with cough. No overt aspiration noted.
  - Further discussion about placement of a feeding tube is addressed. Mr. Johns declines this intervention.
  - There was a consult placed to the palliative care team.

PALLIATIVE CARE

- What is palliative care?
  - Palliative care is specialized medical care with the focus of care for patients with a serious or chronic illness. The goal is to alleviate suffering and address pain and symptoms that can affect the individuals quality of life (QOL). This may include physical, psychological and spiritual needs.
  - Palliative care helps patients and families cope with the symptoms of the disease to improve QOL and functional status.
  - Palliative care also provides education and support in helping the patient make decisions about goals of care.

BENEFIT OF PALLIATIVE CARE

- Supportive care
- Pain and symptom management
- Interdisciplinary focused
- Resources
- Can help with the transition to hospice care
PALLIATIVE SYMPTOMS IN ALS

- Sensory symptoms
- Pain
- Spasticity
- Respiratory compromise
- Aspiration risk
- Sleep disruption
- Salivation
- Pseudobulbar affect
- Fatigue

PALLIATIVE SYMPTOMS IN ALS CONT

- Laryngospasm
- Jaw quivering/cheek biting
- Edema
- Constipation
- Urinary urgency
- Functional incontinence
- Depression and anxiety

PAIN

- Nociceptive pain can result from multiple causes
  - Reduced mobility
  - Muscle cramps
  - Muscle spasticity
  - Comorbid conditions
  - Pain with neuropathic features
    - Paresthesia
    - Allodynia
    - Hyperalgesia
  - Pain evident in 15-85% of patients

CASE STUDY

- 12 months since diagnosis: he is now primarily in a wheelchair, he still can feed himself, but has lost weight, now down 15 pounds. Due to decreased oral intake and with increased risk for aspiration he has elected to get a gastrostomy tube.

CASE STUDY

- 18 months since diagnosis: he is no longer able to use a manual wheelchair and recently received a power wheelchair. He is now spending more time in bed. He has had some increased respiratory secretions, his wife assists with oral suctioning often and he uses a cough assist device.
- His breathing is more difficult at times, he is using oxygen at night.
- He has told his wife he does not want to be on a ventilator if his breathing gets worse, he states "I don't want to be hooked up to that machine." He has agreed to non-invasive ventilation with BiPAP, which can improve survival, quality of life and help preserve respiratory function according to Connolly, Galvin & Hardiman 2015.
- His breathing is more difficult at times, he is using oxygen at night.
- He is still taking in small amounts at meal times and is on night time feedings.
- He has a lot of pain in his shoulders and some occasional pain in his legs.
- Palliative care services visit weekly.

CLINICAL PATTERNS OF PROGRESSION

- ALS is a relentlessly progressive disorder...
- The rate of progression between each individual is different it is characterized by a gradual and progressive decline that without intervention remissions or exacerbations get worse over time.
- Symptoms initially spread with the segment of onset and is fairly predictable.
- As it spreads different body parts become immobilized until the patient eventually is confined to a wheelchair or bed.
- The end results are life-threatening and follow the path of either dysphagia or neuromuscular respiratory failure.
  - Neuromuscular respiratory failure is actually the most common cause of death with this disease.
  - Due to this about 5-10% of patients choose permanent ventilation or a tracheostomy.

CASE STUDY

- Mr. Johns has just celebrated his 74th birthday, all his children and grandchildren were able to visit. His children have noted a definite decline.
- Primarily bed bound.
- Needs assistance with all cares.
- Has stage II ulcer over his coccyx and a vascular ulcer on 2 toes on the R foot.
- Primarily only getting nutrition with feedings via his G-tube, but has not been able to tolerate the diet and has episodes of emesis. He chokes with any oral intake, but insists on having his daily coffee and refuses any thickeners.
- Voice is almost inaudible.
- Using oxygen & on BIPAP for comfort, but despite this he has hypoxia.
- He has been having more pain.
- The palliative care team is needing to visit 2-3 times a week now.

CASE STUDY

- His palliative care team reviews his advance directives and has a goals of care discussion with the patient and his family.
- Based on their evaluation and the patient's care needs they recommend a transition to hospice.
- Hospice is a program that cares for the whole person and allows a terminally ill person with a prognosis of six months or less to live comfortably. The goal is comfort and not cure according to Medicare Hospice Benefits. Most prefer to die at home.

HOSPICE CARE

- Criteria to meet for hospice care for ALS
  - Rapid progressive decline in functional status, breathing capacity, compromise in nutritional status due to dysphagia which is inadequate to sustain life.
  - Life threatening complication within the last 12 months
  - Recurrent aspiration pneumonia
  - Pyelonephritis
  - Sepsis
  - Fever recurrent after treatment with antibiotic
- Most common cause of death is progressive neuromuscular respiratory failure.
OUTCOME OF CASE STUDY

• Based on Mr. Johns known goals of care his family is electing hospice care.
• Care and comfort measures are put in place and 1 week later after Mr. Johns spent a day with his family and was more alert than he had been for days, he dies peacefully with his family at his bedside.
• Hospice will continue to follow the family for bereavement care for 12 months.

HOW DOES THIS IMPACT YOUR PRACTICE

• Being able to confidently diagnose ALS based on symptoms and test results
• Able to appropriately recommend a course of action based on where the patient is in the disease process
  • Making appropriate referrals
  • Consulting Palliative care earlier in the disease process
  • Knowing when it is appropriate to consult Hospice care

DESPITE THE TERMINALITY OF ALS SOME HOPE IS ON THE HORIZON

• The Ice bucket challenge
  • The man behind the challenge
    • Pete Frates
      • Behind the Challenge
        • Ice Bucket Challenge Inspiration
  • Saving the spoken words
    • Word Banks
FAMOUS CASE

• Stephen Hawking
  • Diagnosed at 21
  • Still rose to greatness despite his disease
  • Former Lucasian Mathematics Professor at the University of Cambridge
  • Author of the bestseller A Brief History of Time
  • Former Director of Research at the Department of Applied Mathematics and Theoretical Physics
  • Founder of the Centre for Theoretical Cosmology at Cambridge
  • Awarded the CBE (Commander of the Most Excellent Order) in 1982
  • Wheelchair bound
  • Has experienced loss of communication but adapted with different resources
  • Experiences breathing difficulties and uses a respirator at times
  • Still maintains a positive attitude which is key to living with a debilitating disease

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THANK YOU

Questions???
SOURCES

- www.lifehack.org/articles/communication/20-inspirational-quotes-stephen-hawking-everyone-should-read.html