

A case of bilateral upper extremity weakness

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Case Presentation

- An 82-year-old right handed man presented to UAMS for evaluation of bilateral upper limb weakness
- Symptoms started two years ago with left hand weakness characterized by trouble gripping things and holding objects
- Weakness slowly progressed to the extent he is having difficulty writing, having difficulty with holding a “spoon” and lifting heavy objects
- One year later, similar symptoms started in right hand
- Wife noticed “muscle twitching” in multiple areas of the body
- No focal weakness in legs reported
- Denies dysphagia, dysarthria, diplopia, ptosis
- No sensory symptoms reported
- ROS: pertinent for unintentional weight loss of 20 pounds over the course of two years.
- Denies history of SOB.



Physical Examination

Gen: NAD, appears stated age, good personal hygiene

HEENT: NC/AT, no tongue fasciculations, MMM, oropharynx clear

Chest: **No gynecomastia**

Mental Status: alert & oriented x4, repetition intact, speech is spontaneous and fluent, comprehension and naming intact, object recognition normal, recent and remote memory intact

Cranial Nerves: 2-12 grossly intact

No facial fasciculation

Motor: Decreased tone in both hands, **atrophy of intrinsic hand muscles bilaterally, Fasciculations in chest and both arms noted**, no rigidity or spasticity



Physical Examination(Cont...)

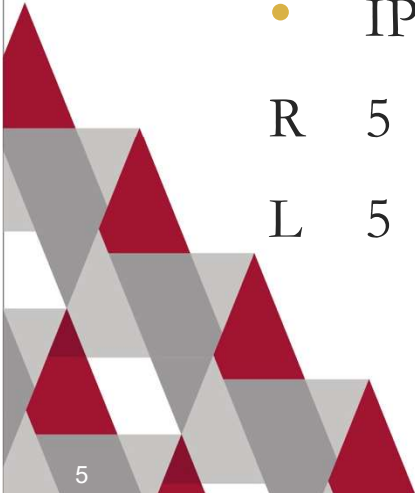
Strength :	Right	Left
• Deltoid	5/5	5/5
• Biceps	5/5	5/5
• Triceps	4/5	4/5
• Wrist ext	4+/5	5/5
• Wrist flex	4/5	4+/5
• Finger abd	3/5	3/5
• Finger flex	4/5	4/-5
• F. extension:	3/5	3/5
• Hip flex	5/5	5/5
• Knee ext	5/5	5/5
• Knee flex	5/5	5/5
• Dorsiflex	5/5	5/5
• Plantarflex	5/5	5/5
• Sensation: intact b/l to light touch, pinprick. Normal vibration/proprioception		
• Reflexes: BR Bic Tri Pat Ach Plantar Clonus		
• Right	0 0 0 1+ 1+ flexion	absent
• Left	0 0 0 1+ 1+ flexion	absent
• Cerebellar/gait: normal		



Physical Examination(Cont...)

Strength Testing (MRC out of 0-5)

- | | Del | SS | IS | BC | TC | WE | WF | FE | FF | APB | FDI | ADM |
|---|-----|----|----|----|----|----|----|----|----|-----|-----|-----|
| R | 5 | 5 | 5 | 5 | 5 | 4+ | 4 | 3 | 4 | 3 | 1 | 1 |
| L | 5 | 5 | 5 | 5 | 5 | 5 | 4+ | 3 | 4- | 3 | 0 | 0 |
- | | IP | Quads | Hip Ad | Gmed | HS | TA | G | EHL | TF |
|---|----|-------|--------|------|----|----|---|-----|----|
| R | 5 | 5 | 5 | 5 | 5 | 5 | 5 | 5 | 5 |
| L | 5 | 5 | 5 | 5 | 5 | 5 | 5 | 5 | 5 |



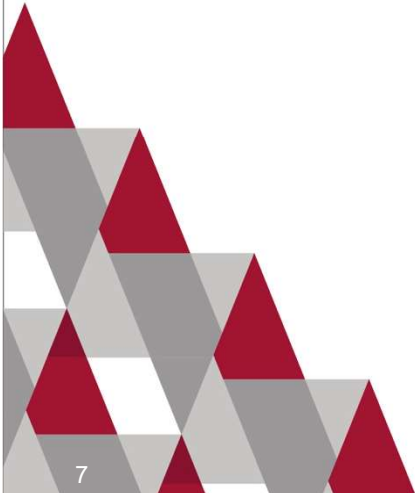
Diagnostic testing

- Labs including C - reactive protein, Erythrocyte Sedimentation Rate, Creatinine Kinase and Thyroid stimulating hormone were normal
- Magnetic resonance imaging (MRI) of the cervical spine showed multilevel degenerative disc disease without spinal cord abnormalities
- EMG needle exam at outside facility showed active and chronic denervation in all the muscles tested in both arms. Has chronic denervation changes(Polyphasics) in cervical and thoracic paraspinals.
- Fibrillations/Positive sharp waves/Polyphasics noted in bilateral deltoid, triceps, extensor digitorum communis, flexor digitorum profundus (digits 2,3) and first dorsal interossei muscles.



Diagnosis

- Due to progressive weakness in upper extremities associated with fasciculations, areflexia in both upper extremities and with absent sensory or bulbar symptoms, we suspect patient likely has **Progressive Muscular Atrophy(PMA) vs Brachial Amyotrophic Diplegia**. PMA thought more likely as he has evidence of chronic denervation in both cervical and thoracic paraspinals.



Progressive Muscular Atrophy(PMA)

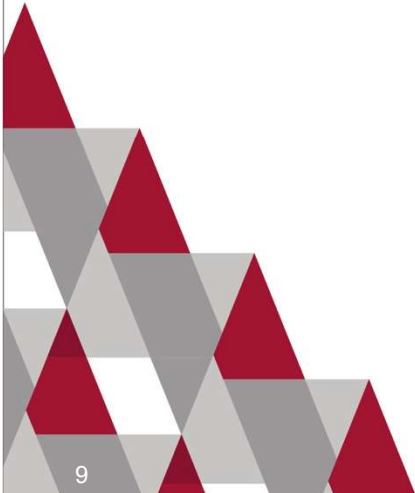
- Synonym: **Duchenne-Aran muscular atrophy**
- Rare subtype of motor neuron disease (MND) that affects the **lower motor neurons**
- Accounts for around 4-10% of all MND cases
- Usually affects people aged over 50 years but younger people can also be affected
- Males>females



Progressive Muscular Atrophy(PMA)

Causes

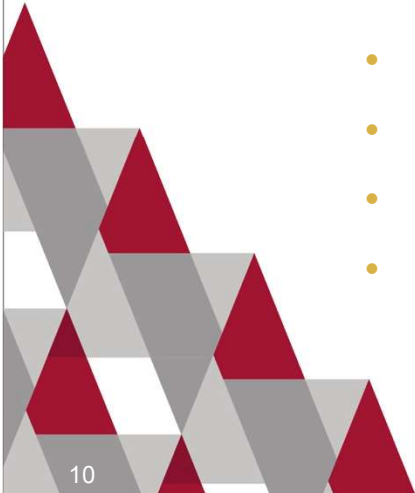
- Unknown
- May be combination of environmental and genetic factors



Progressive Muscular Atrophy(PMA)

Clinical Features

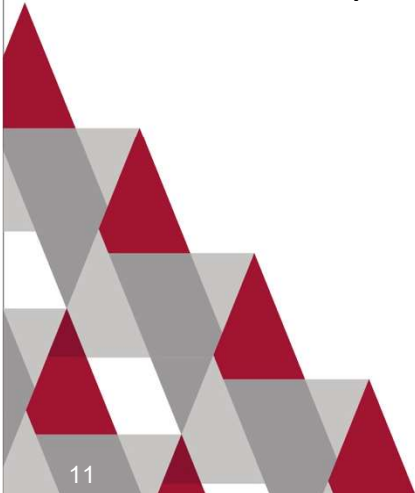
- Weakness: usually starts in hands and moves slowly to legs but can start in either hands or legs
- Clumsy hand movements
- Cramps/fasciculations
- Breathing difficulties
- Weight loss
- bulbar symptoms in late stages
- Atrophy
- Decreased reflexes
- Normal to decreased tone



Progressive Muscular Atrophy(PMA)

How is it different from other MNDs?

- Amyotrophic Lateral Sclerosis(ALS): **UMN+LMN**
- Progressive Bulbar Palsy(PBP): **LMNs in brain stem**
- Primary Lateral Sclerosis (PLS): **UMNs of arms/legs/face**
- Spinal Muscular Atrophy (SMA): **LMNs(Hereditary)**



Progressive Muscular Atrophy(PMA)

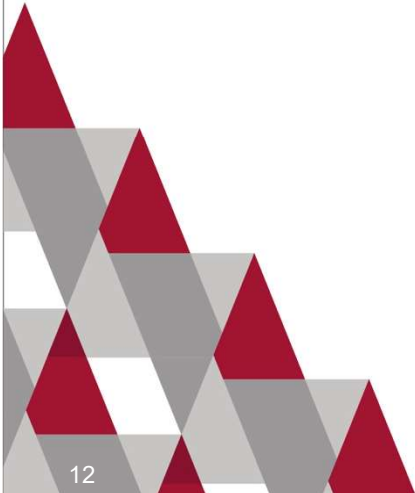
- PMA is distinguished from ALS by **absence of:**

Hyperreflexia

Spasticity

Babinski's sign

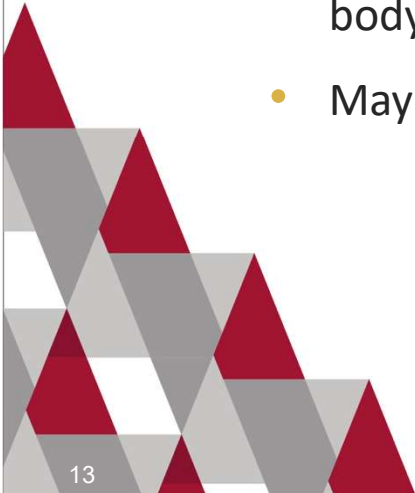
Emotional lability



Progressive Muscular Atrophy(PMA)

Diagnosis

- No specific test
- Labs like B12, TSH, CK, ESR/CRP, GM1 antibodies
- MRI Brain/Spine to rule out other pathologies
- EMG/NCS test: usually show denervation changes in most affected body parts, and in some unaffected parts
- May sometimes need LP to rule out other conditions

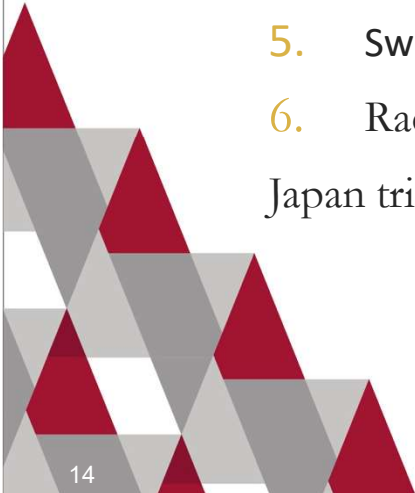


Progressive Muscular Atrophy(PMA)

Treatment

1. No cure. Treat with Riluzole as in ALS patients
2. Symptomatic and supportive treatment
3. PT/OT/Speech therapy as needed
4. PFTs
5. Swallow eval if any history of dysphagia
6. Radicava(Edaravone) is for ALS patients

Japan trial: 33 percent reduction in their rate of decline in physical function.



Progressive Muscular Atrophy(PMA)

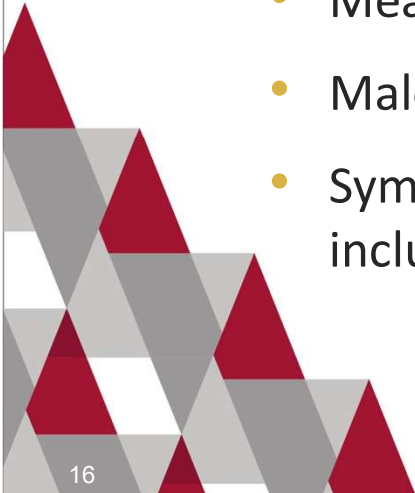
Prognosis

- 1) Better than ALS(ALS 2-5 yrs vs PMA 5-10 yrs)
- 2) An initial diagnosis of PMA could turn out to be slow progressive ALS years later after development of UMN signs

Post-mortem examinations of PMA patients frequently show pathologic evidence of UMN degeneration(Corticospinal tract abnormalities and TDP-43 positive inclusions in motor cortex, in a pattern identical to that of ALS).

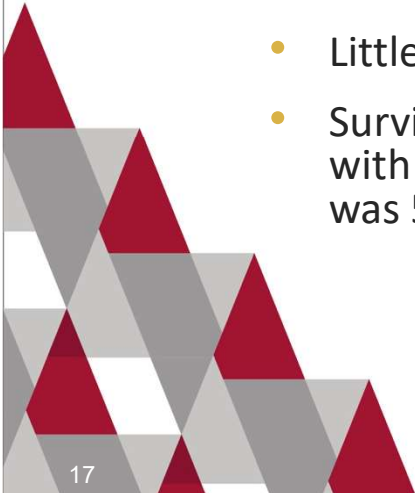
BRACHIAL AMYOTROPHIC DIPLEGIA(BAD)

- Motor neuron disease confined to the **cervical spinal cord region**.
- also been described as **Flail arm** or **Man-in-the-barrel syndrome**
- 2–11.4% of patients presenting with MND
- Mean age of onset is similar to ALS at 53.3–57.3 years
- Males>females
- Symptoms can begin asymmetrically, but usually progress to include both arms (70%)

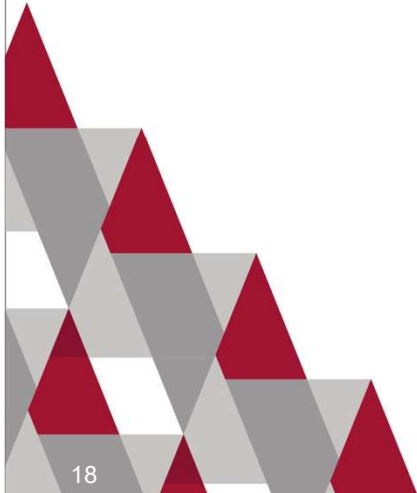


BRACHIAL AMYOTROPHIC DIPLEGIA(BAD)

- Majority of BAD patients have proximal weakness at presentation (70%).
- Specifically, the wasting and weakness of the upper limbs had to be profound, symmetric, and involve proximal muscle groups (MRC grade ≤ 3).
- In most series, patients with BAD have only lower motor neuron involvement at presentation, with decreased or absent reflexes (47–90%), and some series define this group **as a variant of progressive muscular atrophy**
- Little or no weakness of the leg or bulbar musculature.
- Survival of patients with flail arm syndrome might be better than those with other forms of ALS as the median survival in the flail arm group was 57 months



THANK YOU



References

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