Painless, progressive weakness – Could this be Motor Neurone Disease?

1. Does the patient have one or more of these symptoms?

Bulbar features

- Dysarthria
  - Slurred or quiet speech often when tired
- Dysphagia
  - Liquids and/or solids
  - Excessive saliva
  - Choking sensation especially when lying flat
- Tongue fasciculations

Limb features

- Focal weakness
- Falls/trips – from foot drop
- Loss of dexterity
- Muscle wasting
- Muscle twitching/fasciculations
- Cramps
- No sensory features

Respiratory features

- Shortness of breath on exertion
- Excessive daytime sleepiness
- Fatigue
- Early morning headache
- Orthopnoea

Cognitive features

- Behavioural change (with or without dementia)
- Emotional lability (with or without dementia)
- Fronto-temporal dementia

2. Is there progression?

Supporting factors

- Asymmetrical features
- Positive family history of MND or other neurodegenerative disease

Note that MND can present at any age.

Factors NOT supportive of MND diagnosis

- Bladder/bowel involvement
- Prominent sensory symptoms
- Double vision/ptosis
- Improving symptoms

If the answer is YES to questions 1 and 2 – query MND and refer to Neurology.

If you think it might be MND please state explicitly in the referral letter.
Common causes of delay are initial referral to ENT or Orthopaedic services.

Bulbar features

25% of patients present with bulbar symptoms

- Dysarthria
  - Quiet, hoarse or altered speech
  - Slurring of speech often when tired
- Dysphagia – more often liquids first and later solids. Initially can be sensation of catching in throat or choking when drinking quickly.
- Excessive saliva
- Choking sensation when lying flat
- Weak cough – often not noticed by the patient

Consider referral to neurologist rather than ENT if painless, progressive dysarthria is present.

Limb features

70% of patients present with limb symptoms

- Focal weakness – painless with preserved sensation
- Distal weakness
  - Falls/trips – from foot drop
  - Loss of dexterity
    e.g. problems with zips or buttons
- Muscle wasting – hands and shoulders. Typically asymmetrical.
- Muscle twitching/fasciculations
- Cramps

Respiratory features

Respiratory problems are often a late feature of MND and an unusual presenting feature. Patients present with features of neuromuscular respiratory weakness:

- Shortness of breath on exertion
- Excessive daytime sleepiness
- Fatigue
- Early morning headache. Patients often describe a 'muzziness' in the morning, being slow to get going or as if hung over.
- Un-refreshing sleep
- Orthopnoea
- Frequent unexplained chest infections
- Weak cough and sniff
- Nocturnal restlessness and/or sweating

Consider MND if investigations for breathlessness do not support a pulmonary or cardiac cause.

Cognitive features

Frank dementia at presentation is rare. Cognitive dysfunction is increasingly recognised, as evidenced by:

- Behavioural change such as apathy or lack of motivation
- Difficulty with complex tasks
- Lack of concentration
- Emotional lability (with or without dementia)

Ask specifically about a family history of these features.

MND Australia resources for health professionals:

www.mndaustralia.org.au/healthprofessionals

To contact the MND association in your State or Territory call: 1800 777 175

mnda motor neurone disease association

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RACGP Accepted clinical resource

The Motor Neurone Disease diagnostic tool has been officially recognised as an Accepted Clinical Resource by the Royal Australian College of General Practitioners.

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