Dysphagia as the sole manifestation of myasthenia gravis

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CASE DESCRIPTION

- Man, 92 years
- Incomplete traumatic quadriplegia
- RMI : wide spinal cord contusion at the sixth cervical segment
- Treatment : rigid neckbrace during 3 months

EVOLUTION

- Fair during one month, normal diet, no decreased swallowing ability or no tracheal aspiration reported
- Many episodes of pneumonia → treated with antibiotherapy
- Oropharyngeal examination : primary and secondary tracheal aspiration
- Nasogastric tube

EXPLORATION

- CT-scan of the brain, CT-scan and RMI of the brain stem : neg
- CT-scan of the thorax : neg

- Blood exam :- bacterial, viral and parasitic serology : neg
  - acetylcholine receptors antibodies : neg

- ENMG : - no motor unit disorders (tongue, SCM, deltoidus)
  - repetitive nerve stimulation (trapezius, anconeus, nasalis) : normal

- Videofluoroscopic swallow study : defect of epiglottis closing with important tracheal aspiration

Prélèvement: 23/11/2005 à 08:00

No. de Référence : 0511-65793 Votre référence :
Date de réception: 22/11/2005

ANALYSE		RESULTAT	UNITE	REFER.
		0511-65793 23/11/05		
BACTERIOLOGIE:SEROLOGIE ET RECHERCHE DIRECTE				
ASL:		41	U/mL	
Borrelia IgG:		<5	Eu	<14
Borrelia IgM:		<5	Index	<10
VIROLOGIE				
Virus Syncytial Resp. (IgM+A)		4	Index	<12
Influenza A (IgM + IgA)		7	Index	<11
Influenza B (IgM + IgA)		8	Index	<11
Parainfluenza 1-2-3 (IgM+A)		3	Index	<10
Adénovirus (IgM + IgA)		2	Index	<12
Herpes simplex (1 et 2) IgG:	+	>2999		<110
herpes simplex (1 et 2) IgM:		Négatif		
Varicelle - Zona IgG:	+	738	mUI/mL	<165
Varicelle - Zona IgM:		Négatif		
Cytomégalovirus IgG:	+	>250	AU/mL	<15
Cytomégalovirus IgM:		Négatif		
Paul et Bunnell:		Négatif		
EBV VCA IgG :	+	230	Index	<25
EBV VCA IgM:		Négatif		
Parvovirus B19 IgG:	+	38	U	<20
Parvovirus B19 IgM:		Négatif		
Pool Echovirus/Coxsackies (FC)		Inférieur à 10		
PARASITOLOGIE				
Recherche de Toxoplasme				
Toxoplasme IgG:		<1	UI/mL	<2
Toxoplasme IgM:		Négatif		

-Protocole COMPLET- Avec l'expression de nos sentiments confraternels

Validé par : Dr J.M. Minon,Dr. J.M. Senterre

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Dr.JM.SENTERRE - Mme.N.GILLAIN - Dr.M.BOLAND - Dr.N.BROUWERS - Dr.O.KETELSLEGERS - Dr.C.NEVE

DIAGNOSIS

Single-fiber EMG : neuromuscular transmission defect

ID #: ANONYMOUS

HOPITAL DE LA CITADELLE
SERVICE UNIVERSITAIRE DE NEUROLOGIE
NEUROPHYSIOLOGIE CLINIQUE
Boulevard du XIIe de Ligne, 1
4000 LIEGE
Tél: 04/225.61.11 Rendez-vous: 04/225.60.70

DATE D'EXAMEN:

NOM: XXX
ADRESSE:
VILLE:

Patient ID:
SEXE:
D.N.:

MEDECIN : DR KAUX

MOTIF DE L'EXAMEN:

Dysphagie et dysphonie isolées.

Single Fiber EMG:

Extn. Dig. Com Number of recordings analyzed: 14 Number of recordings with blocking: 2
Mean test values are: MCD: 78 µs. MSD: 67 µs. Percentage of recordings with blocking: 14 %
Individual MCD values ranging from 22 µs to 156 µs.

Pr A. MAERTENS de NOORDHOUT
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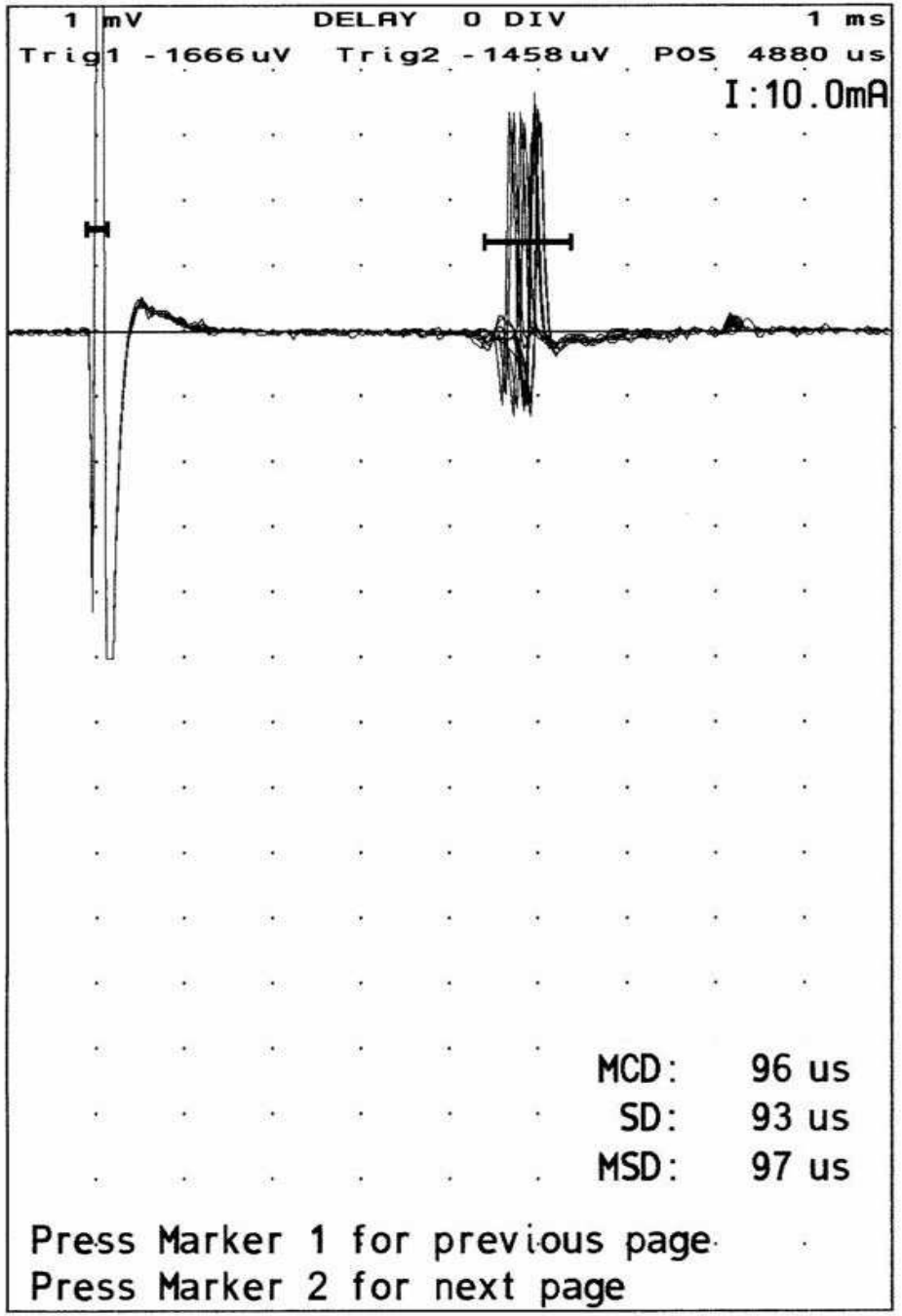
1-64610-96-760

STIM SFEMG REANA # 2



DIAGNOSIS

Single-fiber EMG : discreet neuromuscular transmission defect

↓

Swallow test after neostigmine injection : satisfying for solids and liquids

↓

Videofluoroscopic swallow study 20min after neostigmine injection : epiglottis mobility improvement no more tracheal aspiration

↓

DIAGNOSIS : MYASTHENIA settled in the pharynx

TREATMENT

- Medicinal : Mestinon® 60mg 3x/day
- Taking out of the nasogastric tube and progressive coming back to a normal diet
- Logopedic rehabilitation :
  - Strenghtening of the other protection mechanisms of the breathing passages
  - Swallowing instructions to strenghten the early closing of the breathing passages

EVOLUTION (>6 months)

- Normal diet
- No recurrence of secondary pulmonary infection
- Complete oropharyngeal examination: normal

DYSPHAGIA IN MYASTHENIA

- 15-40% in myasthenia gravis
- 6% as chief complaint
- Very uncommon as the only manifestation
- Literature review : few cases
- Characteristics :
  - Tongue and pharyngeal muscle fatigue (→ with repeated swallowing)
  - Decreased of the amplitude of peristaltic contractions along the entire oesophagus

EXPLORATION OF DYSPHAGIA

1. ENT examination : swallow test, laryngoscopy, …

2. Blood examination:
   - Bacterial, viral and parasitic serology
   - Acetylcholine receptors antibodies sensitivity:
     - 55-64% in ocular form
     - 80-89% in generalised form
   - Musk antibodies

3. Videofluoroscopic swallow study

EXPLORATION OF DYSPHAGIA

4. Oesophageal manometric study
5. CT-scan, RMI : brain, brain stem, thorax
6. Gastroscopy

EXPLORATION OF DYSPHAGIA

7. ENMG
   - Conventional
   - Repetitive nerve stimulation
   - Single fiber EMG

   SENSITIVITY
   - Extensor digitorum communis
     - 99% generalised form
     - 97% ocular form
   - Several muscles
     - 89% generalised form
     - 60% ocular form

EXPLORATION OF DYSPHAGIA

8. If myasthenia is suggested
   → endophonium test (neostigmine, pyridostigmine)

SENSITIVITY : 86% ocular form
95% generalised form

SPECIFICITY :  false positive results (Guillain-Barré, ALS)

CONCLUSION

- Always be careful of signs wich could make think of swallowing troubles
  - Cough while eating
  - Repetitive pulmonary infection (silent aspiration)
- Always explore when there is a doubt

DIFFERENTIAL DIAGNOSIS

1. Obvious cause
2. Non obvious cause
3. Uncommon

1. OBVIOUS CAUSE

- Neurologic disorders : cerebrovascular accident
- Traumatic oesophagus
- Infectious origin (steptococcus B, heamophilus, staphylococcus, syphillis, EBV, herpes, mycosis)
- Obstructive disorders : object inhalation
- Sequelae of burn or radiotherapy

2. NON OBVIOUS

- Cancer (oropharyngeal, oesophagus)
- Neurologic or motor unit disorders : myasthenia, multiple sclerosis, amyotrophic lateral sclerosis, muscular dystrophy, polymyositis, Guillain-Barré
- Inflammation of oesophagus, ulcer
- Zenker’s diverticulum, spasm of upper oesophagus

3. UNCOMMON

- Mobility problems : achalasia, scleroderma
- Anaemia, Plummer-Wilson syndrome
- Nearness disease : mediastinal tumour, costal anomaly…
- Mild form of tumour of oesophagus
- Oropharyngal dryness : neurologic drug, Gougerot-Sjögren syndrome…

Thank you for your attention.