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Sudden dysphagia in an elderly, quadriparetic patient

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Abstract

Clinical case. - We report the case of a 92-year-old quadriparetic patient who suddenly presented a major swallowing disorder following trauma to the neck. A diagnosis of myasthenia gravis was suggested by single-fibre EMG of the *extensor digitorum communis* muscle. In view of the continued absence of dysphagia after 6 months of pyridostigmine treatment, this diagnosis was considered as definitive.

Discussion. - We review the various characteristics of myasthenia gravis and ways of investigating dysphagia.

Conclusion. - It is important to be aware of and investigate all the direct and indirect signs of dysphagia and establish the exact aetiology in order to provide the best possible treatment.

Keywords: Dysphagia; Myasthenia gravis; Pharynx; The elderly *Mots clés :* Dysphagie ; Myasthénie ; Pharynx ; Patient âgé

1. Case report

Mr G.M. (92 years of age) was living alone and was fully independent. His personal medical history included only minor injuries (fracture of the humerus and a sprained left ankle) and mild hypertension (controlled by dietary and lifestyle measures and 2.5 mg of indapamide per day). His general practitioner had also prescribed an aspirin a day as a stroke preventive measure.

However, after tripping on a step, he fell forward and landed on his face. Mr G.M. remained conscious but presented incomplete tetraparesis at the C6 level and which was rated as a "C" on the American Spinal Injury Association (ASIA) spinal cord damage scale (incomplete spinal cord lesion - motor function is preserved below the neurological level, and more than half of key muscles below the neurological level have a muscle grade less than 3 out of 5).

Magnetic resonance imaging (MRI) of the cervical spine evidenced:

- a spinal cord contusion behind the C6 vertebral body;
- a left posterolateral disc herniation involving compression of the C6 C7 left roots.

The suspected cervical instability in C5/C6 and C6/C7 was treated conservatively by instructing the patient to wear a neck brace for 3 months.

After 1 month of slow but steady recovery, Mr G.M. suffered from recurrent bouts of

bronchopneumonia and was treated with intravenous amoxicillin and clavulinic acid.

Furthermore, he did not present diplopia and was able to close his eyes completely and chew correctly (as assessed by a speech therapist).

An ears, nose and throat (ENT) examination evidenced primary and secondary choking on food. A videofluoroscopic swallow study revealed defective epiglottal tilting with major tracheal aspiration. Consequently, the patient was fitted with a nasogastric probe.

Several investigations were performed in order to identify the causes of the choking: a comprehensive set of blood tests (including assays for thyroid hormones, fasting glycaemia, antibodies against bacteria [*Borrelia*, *Streptoccoccus*], viruses [cytomegalovirus, herpes, para-influenza, Epstein-Barr virus] and parasites [toxoplasma] and anti-acetylcholine receptor and anti-muscle-specific tyrosine kinase [MuSK] antibodies), electroneuromyography (EMG, for detecting either myopathy, neuropathy or anterior horn cell disease) and a brain MRI scan. All these examinations and tests were negative. No thymoma was found.

Given that electrophysiological decrement analysis at 3 and 50 c/s in the *abductor digiti mini*, *anconeus*, *trapezius* and *nasalis* was negative and that the electrodetection results for the tongue, *sternocleidomastoidus* and *deltoidus* were normal, single-fibre EMG examination of the *extensor digitorum communis* was performed; it revealed mild deterioration of neuromuscular transmission (increased jitter), with two blocks in 14 recordings (14%), a mean difference between consecutive discharges (MCD) of 78 µs and individual MCD values of between 22 and 156 µs. Our patient presented initial C6 damage which was capable of producing abnormalities in the single-fibre EMG analysis [10]. It was thus necessary to integrate these single-fibre EMG results into the clinical context and the findings from other investigations; it should also be remembered that the MCD tends to increase with age [9,10].

A diagnosis of myasthenia gravis was thus considered and so a neostigmine test was performed in the presence of the speech therapist. The clinical result was good for swallowing solid and liquid food and was confirmed by videofluoroscopy; 20 min after the injection of neostigmine, epiglottis motility had improved and there was no tracheal aspiration. A treatment with pyridostigmine (60 mg, three times a day) was initiated and enabled a progressive return to normal eating.

A rehabilitation programme was started in order to improve the mechanisms for protecting and closing the respiratory pathways during swallowing and enhance the voluntary control of the three steps of the swallowing [5].

A control ENT examination did not evidence any choking and confirmed complete emptying of the hypopharynx and better epiglottal motility.

Despite the absence of anti-acetylcholine and anti-MuSK receptor antibodies, the diagnosis of autoimmune, seronegative myasthenia gravis restricted to the pharyngeal muscles was retained, in view of the abolition of swallowing disorders and the absence of further lung infections 6 months after starting the pyridostigmine treatment. Two years on, our patient has not presented any further episodes of bronchopneumonia and thus belongs to grade 1 in Osserman and Genkins [16] classification (see below), even though there are no ophthalmological problems (only one muscle group is affected).

Although we do not believe that there is a direct relationship between the patient's spinal

trauma and the observation of myasthenia gravis, the symptomatology of this latter condition was more easily evidenced because of the perturbations linked to the tetraparesia; for example:

- decreased thoracic muscle strength makes the cough reflex less efficient;
- the need to lie down or recline because of orthostatic hypotension (prompted by the spinal trauma) increases the risk of choking on food.

2. Discussion

2.1. Myasthenia gravis [12,24]

Myasthenia gravis is a neuromuscular junction disease characterized by weakness and fatigue of the skeletal muscles. It has aprevalence of 1/10 000. In terms of age, incidence of the condition is highest in women between 20 and 30 and in men between 50 and 60. However, there is a second peak in geriatric patients (between 70 and 80 years of age) because of the demographic increase in this age range [1].

The lower incidence in old patients complicates the diagnosis, especially when the symptoms fluctuate. Weakness increases with repeated efforts and may be improved with rest [1]. In general, the face muscles (including the palpebral and oculomotor muscles) display alterations early in the course of the disease. The voice can be nasal if the velum is weak [7]. In 85% of cases, the muscle weakness extends to the limb muscles. Deep tendon reflexes are still present [4].

Dysphagia occurs in 15 to 40% of myasthenia gravis sufferers and is the predominant symptom in 6% of cases [5,14,25]. However, it is seldom the only clinical sign. The literature only describes a few sporadic cases [15,27]. This dysphagia is characterized by weakness of the tongue and the pharynx muscles (pharingeal constrictors and the pharingopa-latine and stylopharingeal muscle) and decreased amplitude of the oesophageal peristaltic contractions, leading to choking [5].

Even though all the major symptoms prompting a diagnosis of myasthenia gravis are easily detectable in young people, the same cannot be said for elderly patients. This is why (at least in part) the diagnosis is rare in this population. In fact, the clinical symptoms are masked by age-related weakness and atrophy of the facial muscles, crumpled skin, associated pathologies (cardiovascular disease and respiratory problems) and the lesser severity of the symptoms. Ocular symptoms are more difficult to detect because of degenerative visual problems (cataracts, macular degeneration) [1].

Osserman and Genkins' clinical classification [16] is still employed today. It includes five grades for the adult form of myasthenia gravis:

- grade I (ocular form): only one group of muscles is altered (usually, the ocular muscles, causing ptosis and diplopia); this is a mild, non-lethal form;
- grade II (generalized form): progressive onset with frequent ocular symptoms progressing to a generalized alteration of the skeletal and bulbar muscles. The respiratory system is intact. This form is mild and seldom lethal;
- grade III (acute fulminant form): rapid alteration of the skeletal and bulbar muscles, with early respiratory alterations; this form is severe and highly lethal;
- grade IV (severe late form): exacerbation of the symptoms of grades I and II after

approximately 2 years of disease progression. The symptoms are similar to those of grade III;
• grade V (with muscle atrophy): similar to grades II, III and IV but with additional disuse atrophy.

There are, of course, paediatric neonatal and juvenile forms of myasthenia gravis [16] but they will not be considered here.

The fundamental abnormality in myasthenia gravis is a neuromuscular transmission disorder linked to low availability of acetylcholine receptors. In 80 to 90% of cases (essentially in the generalized forms and more seldom in the local forms), the disease is caused by auto-antibodies against the acetylcholine receptor. The antibodies prevent binding of the acetylcholine released by the nerve endings at the postsynaptic muscular membrane and, consequently, reduced efficiency of neuromuscular transmission. Around 30% of the patients lacking anti-acetylcholine receptor antibodies have anti-MuSK antibodies [12,24].

Myasthenia gravis can be induced or worsened by a number of drugs acting pre- and/or postsynaptically via inhibition of the conductance of the muscle membranes or via an immunological effect. D-penicillamine is frequently associated with this symptomatology but several classes of drugs can be concerned [2,17,18]:

• antibiotics such as the aminoglycosides (streptomycin, gentamycin, tobramycin, neomycin), the carbapenemes (imipenem) and clindamycin and its derivatives (lincomy-cin);
• antiarythmics (quinidine, procainamide, disopyramide);
• antihypertensives (captopril, propranonol, verapamil);
• antimalaria drugs (quinine, chloroquine, hydrochloroquine);
• antiepileptics (phenytoin);
• antiparkinsonian agents (amantadine);
• antidepressants (imipramine, amitryptiline);
• antipsychotics (droperidol, haloperidol);
• anesthetics (ketamine, procaine);
• immunomodulators (interferon alpha);
• magnesium (hypermagnesaemia).

Myasthenia gravis can be associated with other autoimmune diseases (in 5% of the cases), such as diabetis, thyroiditis, rhumatoid arthritis, lupus erythematous and sickle cell anaemia [1,4,17].

Treatment of myasthenia gravis depends on each patient's situation and may involve pyridostigmine, corticosteroids, thymectomy, immunosuppression, intravenous immunoglobulins and/or plasmapheresis.

3. Causes and investigation of dysphagia in the elderly patient

Dysphagia is mainly a public health problem in the elderly, most frequently when they are confined to bed because of health problems (after a femoral neck fracture, for example) [19,28,29].

Seventy-five percent of cases of dysphagia appear in the oropharingeal phase of swallowing and result from a neurological problem such as a stroke, dementia or Parkinson's disease. The dysphagia can be either transitory (such as in strokes) or can increase with disease progression, such as in dementia or Parkinson disease's [8,29].

Only 25% of cases of dysphagia occur in the oesophageal phase of swallowing and often result from obstruction by an oesophageal tumour, candidiasis, oesophageal reflux, a Zenker diverticulum or the effect of a drug [8,29].

When a patient presents dysphagia (coughing when eating or recurrent bronchopneumonia), a standardized three-step examination [29] must be performed:

- 1: examine the patient, his/her state of consciousness, posture, voluntary coughing, voice quality and salivation control;
- 2: ask the patient to drink a dessert spoonful of water;
- 3: if the patient can drink the spoonful of water without choking, ask him/her to next drink a small glass of water.

If this simple test is positive, the patient has to be further investigated.

The most sensitive and specific test for dysphagia is the videofluoroscopy, which enables the three phases of swallowing to be studied and reveals whether the pharyngeal and oesophageal muscles function normally [6,29].

Fibroscopic examination (pharingolaryngoscopy) enables visualization of the dysphagia and, in some cases, determination of its cause [6,29].

If no aetiology is found, other functional tests and investigations have to be performed:

- blood tests, to exclude an infectious aetiology (bacterial, viral or parasitic) or iron deficiency. Antibodies against acetylcholine receptors (specific if positive) and secondary anti-MuSK antibodies have to be screened for;
- computer tomography and MRI examination of the brain stem and encephalon, to exclude any lesions at this level;
- gastroscopy, to detect acquired lesions of the oesophagus and stomach;
- EMG (see below).

If, as in our case, functional investigations suggest myasthenia gravis, a swallowing test 20 min after neostigmine injection should be considered. False positive tests are possible (in Guillain Barré syndrome or amyotrophic lateral sclerosis). The diagnosis should be considered after integration of all the results generated by the various, complementary functional investigations [6].

4. EMG examinations in myasthenia gravis

Conventional EMG enables the physician to rule out diagnoses such as anterior horn cell disease, myopathy and chronic inflammatory demyelinating polyneuropathy.

A repetitive stimulation test on a nerve innervating a symptomatic muscle is advisable when anticholinesterase treatment has been stopped 12h previously, (with a skin temperature of 35 °C and a stimulation rate of between 2 and 5 Hz) at rest and perhaps also after effort. The test is positive if one sees a decrease in the muscle's action potential [3,12,21].

In the event of a negative test or in local forms, single-fibre EMG should be performed [11,13,20,22,23,26]. According to the literature, the sensitivity of this examination varies between 60 and 99%, depending on the type of myasthenia and the muscle examined [13,20]. Its specificity is, however, decreased by the presence of abnormalities also potentially found

in neuropathies or radiculopathies. If the symptoms are focal but not ocular, the analysis can start with the *extensor digitorum communis* [3]. Neuromuscular blocks and/or increased MCD values (otherwise located between 10 and 50 µs) make it possible to diagnose myasthenia gravis [21]. The MCD value tends to increase with age and certain abnormalities can be evidenced with single-fibre EMG - particularly after spinal damage, as in our case [10].

5. Conclusion

In the elderly patient, it is important to be aware of all signs of dysphagia - either direct (choking or nasal regurgitation) and/or indirect (coughing when eating, recurrent lung infections, denutrition). Videofluoroscopic and pharyngolaryngoscopic examinations can sometimes evidence these signs. However, other functional investigations (blood tests and radiological and electrophysiological examinations) may be necessary to determine the exact cause and to optimize patient care.

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