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CASE REPORT

Efficacy of botulinum toxin type-A and swallowing treatment for oropharyngeal dysphagia recovery in a patient with lateral medullary syndrome

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ABSTRACT

BACKGROUND: Wallenberg's syndrome (WS) is known as posterior inferior cerebellar artery syndrome. Dysphagia has been reported from 51% to 94% of the patients, ranging from mild to severe.
CASE REPORT: We reported a case of a patient (male; 52 years) with WS. MRI showed an intense hypodense area in the dorsolateral part of the ponto-medullary junction. The clinical signs were severe dysphagia, fed by PEG (FOIS 1; PAS 7), sialorrhea, trismus and ataxia.
CLINICAL REHABILITATION IMPACT: Dysphagia was treated by botulinum toxin type-A (BoNT-A), which was injected into the parotid and submandibular salivary glands, temporalis and masseter muscles, cricopharyngeal muscle associated with specific swallowing exercise and food trails. The 3-months follow-up showed significant saliva reduction and improvement of swallowing to from PEG feeding to consistent oral intake of food (FOIS 3, PAS 5). The treatment with BoNT-A combined with swallowing rehabilitation was fundamental in order to restore the swallowing functions.

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Key words: Lateral medullary syndrome - Botulinum toxins, type A - Deglutition disorders - Sialorrhea.

Lateral medullary syndrome (LMS) is also known as posterior inferior cerebellar artery syndrome or Wallenberg's syndrome (WS). It results from an occlusion of posterior inferior cerebellar artery or the vertebral artery.[1] The symptoms of LMS are recognised to be several and vary from patients to patients.[1, 2] Zhang *et al.* 2008 classified LMS in five type on the basis of sensory involvement pattern, anatomical localization and radiological pattern of involvement.[2] Type 1: ipsilateral face and contralateral trunk and limbs; type 2: Ipsilateral face and contralateral face, trunk and limbs; type 3: contralateral face and body; type 4: Ipsilateral face and contralateral trunk and leg; type 5: contralateral face, arm and upper trunk.

The swallowing pattern generator is located in the dorsolateral medulla oblongata and it consists primarily of the nucleus tractus solitaries, nucleus ambiguous and the reticular formation.[3] For this reason, occurrences of dysphagia symptoms are frequent from 51% to 94% and severity is documented to range from mild to severe in LMS patients.[4]

Here, we report a case of WS-type 1- associated with

severe dysphagia, sialorrhea and trismus treated by botulinum toxin type A (BoNT-A) and by designed swallowing rehabilitation.

Case report

A 51-year-old Caucasian man diagnosed with stenosis of right vertebral artery due to arterial hypertension and consecutively lateral medullary syndrome (Figure 1). The patient was fed via PEG, positioned since the first month after stroke. Four months after the event, he was recovered at Neurorehabilitation Hospital at San Camillo Foundation IRCCS (Venice, Italy). MRI showed in flair/T2 an intense hypodense area in the dorsolateral aspect of the ponto-medullary junction of the brain stem with extension into the right inferior cerebellar peduncle (Figure 2). The motor limb examination showed presence of ataxia and weakness of the left upper limb. He could walk independently using a walker, the 10 meter walking test was 16 seconds. Functional Independence Measure (FIM) [5] Score was 92/126 and Fugl Meyer Lower [6] Limb Test was 97/112. The major problem was his balance and coordination, confirmed by Berg Balance Scale [7] Score 38/56. The tactile sensation of the upper and lower limb was normal.

Clinical swallowing evaluation showed presence of severe dysphagia (Functional Oral Intake Scale=1-SS, no oral intake) [8] and severe sialorrhea, saliva pools were removed with tissues every 10-20 minutes during day time and every 1-2 hours during night time. This impacted on his sleep-awake cycle as well as quality of life. Water Swallowing Test (3 mL) confirmed the presence of severe dysphagia associated a wet voice and episodes of choking and coughing. Food trials were not carried out because of the open mouth impairment. Cranial nerve exanimation confirmed dysfunctions of V°, VII°, IX°, X° cranial nerves ipsilateral to the lesion. Trigeminal nerve deficit (V°) resulted in hypercontraction masseter and temporalis muscles causing a severe trismus (interincisal opening of 20 mm) and impaired tactical sensation of right side of the face and 2/3 anterior of the tongue. Facial nerve (VII°) evaluation showed a peripheral right facial paresis and Horner syndrome in the right eye. Absence of GAG reflex confirmed the glossopharyngeal (IX°) dysfunction. He had a hoarseness and wet voice and a deficit of reflex cough test due to vagal nerve impairment (X°). Tongue protrusion, elevation and lateralization were possible, although the examination was compromised by deficit of mouth opening.

The instrumental swallowing evaluations were carried out through videofluoroscopy (VFS) and fibroendoscopic (FEES) ex-

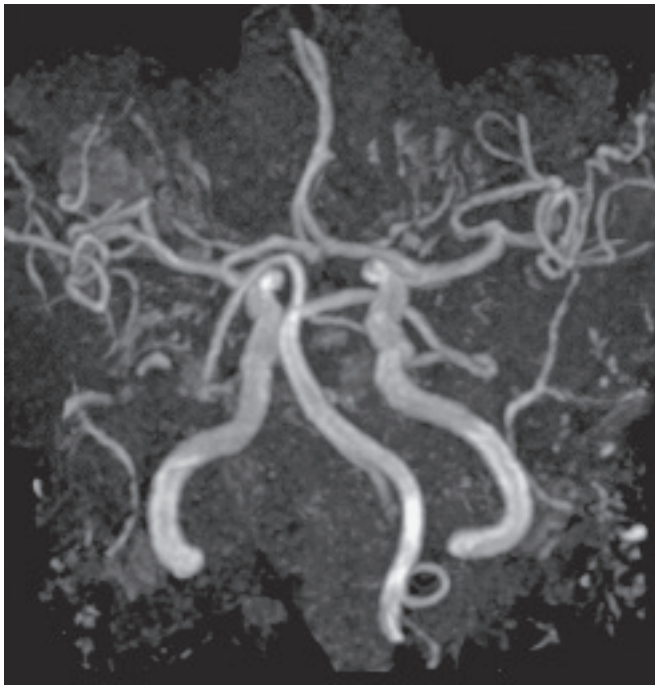

Figure 1.—The magnetic resonance angiography shows occlusion of the right vertebral artery.

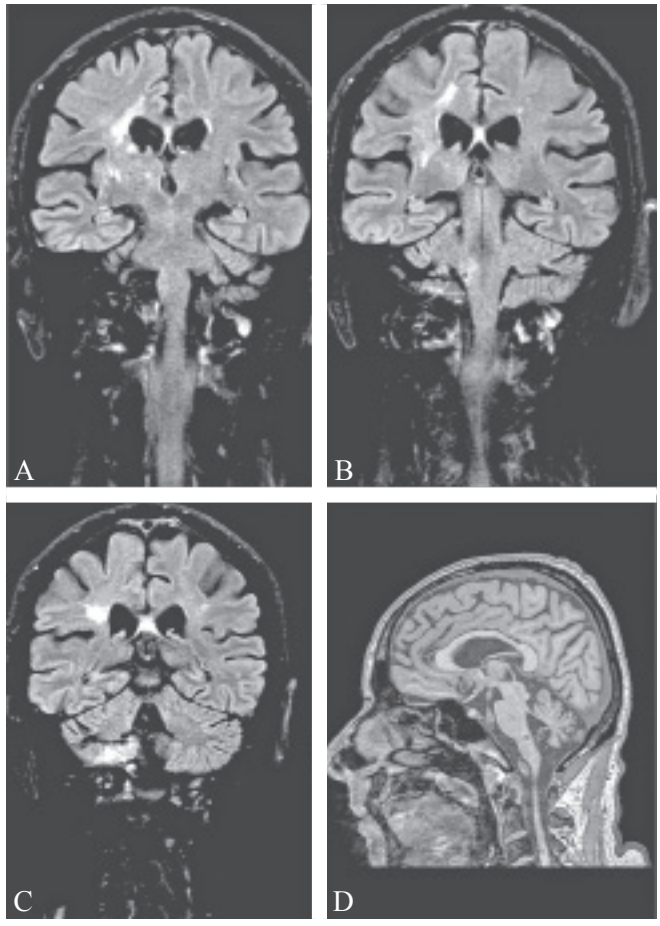

Figure 2.—Magnetic Resonances Imagine (FLAIR) at four months after the event. Coronal (A, B), axial image (C) and sagittal (D) show an ipodense area confined to a small region of the dorsolateral medulla just anterior to the floor of the fourth ventricle and rostral to the obex.

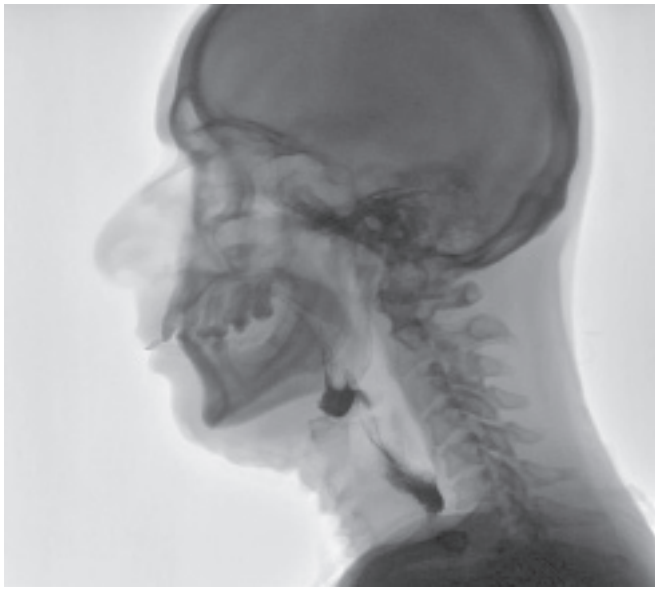

Figure 3.—Videofluoroscopy imagine which shows the residues at the level of vallaculae and piriform sinus.

ams. VFS (Figure 3) was performed using the modified barium with 5 mL semiliquid consistencies (yogurt). It showed an increased oral transit and difficulties to propel the food into oropharynx and deficit of base of tongue and posterior pharyngeal wall approximation resulting in trace oropharyngeal spillage and valleculae residues, lack of hyolaryngeal elevation, reduced pharyngeal motility and shortening and ultimately the upper esophageal sphincter failed to open, as a consequence the liquid and puree boluses pooled in the piriform sinus. Small amounts of residues were aspirated without the cough reflex (PAS: 8). FEES showed deficit right pharynx, right vocal paresis and abundant saliva pools in valleculae and piriform sinus. At the swallowing trails (3 mL of yogurt), the bolus felt into the oro-pharynx, without triggering swallowing reflex and crico-pharyngeal relaxation. There were several pharyngeal residues mostly in the right valleculae and piriform sinus and presence of food residues below the glottis (PAS: 8).

Swallowing treatment

From the onset of the event, the patient underwent a traditional swallowing treatment consisting in active exercises to strengthen the swallowing musculature and oral manipulation during the recovery in the acute hospital. No effects were reported at the follow-up assessment at 4 months; the patient was unable to swallow saliva and he was fed by PEG. Therefore, we decided to proceed with the botulinum toxin injections in order to reduce the saliva, to relax the hypercontraction of mastication muscles and to release the upper esophageal sphincter. A total dose of 100 units of botulinum toxin type A (Xeomin®, Merz Pharmaceuticals, Frankfurt/M, Germany) (2 mL dilution 0.9% saline) was percutaneously injected firstly into the bilateral parotid and submandibular salivary glands under ultrasonography-guidance (25 UI for each gland). Secondly 120 units of BoNT-A were injected to the masseter and anterior temporalis muscles under EMG control in order to monitor the muscles contractions (40 UI for each masseter, 20 UI for each anterior temporalis muscle) and ultimately 15 units were injected percutaneously under EMG control into the right side of the crico-pharyngeal using a teflon-coated needle. The needle was inserted percutaneously 1.5 cm lateral to the palpable inferior border of the cricoid cartilage. Then, it was advanced in a postero-medial direction following the contour of the cricoid cartilage. The patient was asked to vocalize in order to exclude the possibility of placement in the intrinsic laryngeal musculature and to tense their neck and to tilt their head to prevent needle placement in the strap or paraspinal muscles. Needle placement in the UES was confirmed by muscle activity as indicated by an active EMG signal at rest.

After the injections, the patient underwent swallowing treatment, which consisted of active exercises and food swallowing trials for 1 hour a day for 5 days a week. The swallowing exercises comprise dry effortful swallows, shaker techniques and vocal effortful pitch glide associated with manipulation in order to foster laryngeal excursion. The food-swallowing attempts started with small quantities of ice chips in order to trigger the swallowing and increase the sensory information. The patient was asked to try to swallow down and cough afterwards to protect the airways. At one-month follow-up assessment after the BoNT-A treatment, he showed an important improvement of sialorrhea and swallowing function. The saliva amount was significantly reduced, he needed to remove saliva every five hours during day time and no during the night time, which has improved significantly his quality of life. Interincisal opening increased from 20 mm to 30 mm, allowing the use of a spoon for eating trials. Food assumptions also improved from nil-by-mouth diet (FOIS: 1), to minimal oral intake (FOIS: 2) about 100 gr of semisolid consistency (yogurt) per day. The swallowing was possible using the compensatory maneuver of effortful pitch glide and followed by voluntary cough. These maneuver were likely to facilitate the hyolaryngeal excursion and so the bolus was squeezed into the pharynx and then to the esophagus. After 3 mouths follow-up, he was able to eat an entire meal of semisolid consistency food (FOIS: 3) and no side effects were reported. The videofluoroscopy at 3 months showed delay on oral phase and incomplete tongue pharyngeal wall approximation but the several swallowing attempts. In particular, effortful pitch glide contributed to hyolaryngeal excursion and facilitated the bolus flow, with minimal pharyngeal residues after swallowing at the level of vallaculea (PAS 4).

In this single case study, the treatment with combined with the swallowing rehabilitation was fundamental for swallowing recovery. On the one hand, BoNT-A had improved the saliva control, mouth opening and facilitated the crico-pharyngeal relaxation. On the other hand, the swallowing rehabilitation exercises and food administrations promoted the hyolaryngeal elevation and consequently fostered the swallowing reflex.[9]

Discussion

To our knowledge, it has been described for the first time a case of severe dysphagia associated with trismus in a patient with WS, who was treated by BoNT-A injections in three different oral-pharyngeal areas. The diagnosis in this patient was obtained through detailed

clinical and instrumental examinations. The brainstem lesions at the level of central pattern generator for swallowing included nucleus tractus solitaries, nucleus ambigus and trigeminus cranial nerve nuclei caused severe dysphagia characterized by saliva pooling, tempo-mandibolar occlusion and crico-pharyngeal contraction. In this case, the BoNT-A was fundamental for relaxing the contraction of antagonist muscle allowing the swallowing recovery. In the literature, it is well know the effects of botulinum toxin for dysphagia.[10] However, none of the studies have described interventions in three different areas in oral-pharynx complex for the dysphagia recovery in WS. In addition, the key to success of this treatment was the combination of BoNT-A with the swallowing rehabilitation intervention, which have facilitated the food injection using specific technique and stimulations. The positive effects of the combination of BoNT-A and motor exercise are extensively documented for limbs rehabilitation, however, few studies have addressed it for swallowing recovery. Based on this evidence, we infer that the BoNT-A injections followed by a designed and intense rehabilitation contributed substantially to restoring the swallowing function.

Conclusions

In this case of Wallenberg syndrome characterized by severe dysphagia the treatment with BoNT-A injections with swallowing rehabilitation was fundamental in order to reduce the saliva amount and to start the food administrations.

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Conflicts of interest.—The authors certify that there is no conflict of interest with any financial organization regarding the material discussed in the manuscript.

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