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LITERATURE REVIEW

Functional recovery after surgical approach of jugulotympanic glomus tumors

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ABSTRACT

Paragangliomas are tumors of the neuroendocrine system, which arise from the neural crest and chemoreceptor cells. They can be found in the adrenal gland, in the aortic arch, in the jugular bulb or at the level of the cochlear promoter. Glomus tumors grow slowly, are well vascularized, invasive and locally aggressive. Because of their anatomic relation with the jugular bulb, the internal carotid artery, the facial nerve and the cranial nerves IX, X, XI, XII, the patients may develop important complications. Therapeutic options for head and neck paragangliomas are surgical treatment with endovascular embolization, conventional radiotherapy, stereotactic radiosurgery or a combination of these therapeutic methods.

In case of paragangliomas, it is essential to take into consideration the size and localization of the tumor, in order to establish the proper therapeutic approach, either conservative, radiotherapy or surgical, due to the risk of postoperative nerve palsies, details that we try to cover in this review.

In order to emphasise the importance of correct diagnosis, proper treatment, postoperative complications and postoperative functional recovery in the case of glomus tumors, we present the case of a 35-year-old female who was diagnosed and surgically treated for a right jugulotympanic paraganglioma. The postoperative cranial nerve injuries involved VII, IX and X nerves and the patient followed multiple protocols of functional recovery for the affected nerves. After specific therapy, the patient partially recovered the function of the right facial nerve; deglutition and voice were significantly improved.

KEYWORDS: paraganglioma, jugulotympanic, jugulo-carotid, facial palsy, swallowing disorders.

INTRODUCTION

Paragangliomas are tumors of the neuroendocrine system, which arise from the neural crest and chemoreceptor cells. They can be found in the medulla of the adrenal gland, in the aortic arch in the chemoreceptor system or in the bifurcation of the carotid artery, in the temporal bone at the level of the jugular bulb or at the level of the cochlear promoter along the Jacobson nerve[1].

Depending on localization, head and neck paragangliomas are classified in temporal bone paragangliomas, jugulotympanic paragangliomas, neck paragangliomas (vagal and carotid body paragangliomas).

The glomus tympanic tumor appears within the inferior tympanic canaliculi or mastoid canaliculi, while the jugular paraganglioma originates from the paraganglia of the adventitia of the jugular bulb. Paragangliomas of the middle ear or the tympanic glomus grow on the Jacobson nerve or the Arnold nerve. Histologically, they appear as a group of cells surrounded by vascular structures (the Zellballen model). At the level of the middle ear, the tympanic glomus has the capacity to extend through areas with low resistance, even producing bone erosions. The tympanic glomus is very rarely at risk of malignant transformation.

Glomus jugulare is localized in the region of the jugular foramen involving the jugular bulb, internal carotid artery or the lower cranial nerves. Usually, jugular paragangliomas originate from the chemoreceptors that lie on the auricular branch of Arnold's nerve (CN

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X) or on the tympanic plexus of Jacobson's nerve (CN IX). The jugular foramen has various sizes, in almost 70% of cases being larger on the right than on the left, and it is incompletely divided by a septum into pars nervosa and pars vascularis[2]. The incidence of glomus jugulare tumors is 1:1.300.000 of the population and they are more common in women (female:male ratio = 3-6:1) between the ages of 40 and 70. They can be found alone or in a family context, in neurofibromatosis type I, von Hippel Lindau disease or MEN II syndrome (multiple endocrine neoplasia type II)[3,4].

1-3% of paragangliomas are functional with secretory activity, mainly norepinephrine. Even though temporal paragangliomas are usually benign, sometimes they induce autonomic dysfunction secondary to catecholamine release[5].

Glomus tumors grow slowly, are well vascularized, invasive and locally aggressive. Because of their anatomic relation with the jugular bulb, the internal carotid artery, the facial nerve and the lower cranial nerves IX, X, XI, XII, the patients may develop important complications, involving phonation and deglutition. Although invasive, they rarely become malignant (less than 10% of cases). They can metastasize to the ribs, vertebrae or spleen[1].

Paragangliomas extend from their origin through areas with low resistance. Ischemic necrosis may determine cochleovestibular damages. Jugulotympanic tumors cause cranial nerve palsies in 35% of cases[6,7].

DIAGNOSIS OF JUGULOTYMPANIC GLOMUS TUMORS

Clinical presentation

In patients with head and neck paragangliomas, symptoms are insidious at the onset. Patients present usually with pulsatile tinnitus, aural fullness, conductive hearing loss (60-80%)[8], which appear as a result of the tumor development into the mesotympanum; sensorineural hearing loss on the same side with the glomus tumor shows the labyrinthine invasion. Patients present to the ENT specialist usually with 2-3 years delay after the beginning of the symptoms, due to the slow growth of the tumor[9].

As a result of lower cranial nerves damage, patients may present with dysphagia, dysphonia, loss of airway protection, shoulder or tongue weakness. Cranial nerve palsies may be silent in nearly 10% of patients. 30-45% of cases have cranial nerve IX and X palsies, almost 30% present XI and XII cranial nerve palsies and in 10–39% of cases the facial nerve is affected[10,11]. When patients present with multiple lower cranial nerve palsies, a jugular foramen pathology should be considered. Facial paralysis is frequently a late sign in evolution, suggesting a poor prognosis.

Sometimes patients may experience purulent or bloody otorrhea due to erosion of the tympanic membrane (14-24% of cases)[10,11].

Rarely, patients may present with vertigo by extension to the bone labyrinth, damage to the vestibulo-cochlear nerve, or extension of the brainstem[12-14].

Physical examination

A pulsatile middle ear mass is pathognomonic of a temporal bone paraganglioma. On otoscopic examination, the jugulotympanic glomus tumor appears as a reddish tumor formation highlighted by the transparency of the tympanic membrane (Figure 1), the so-called "rising sun" sign. Another characteristic of the glomus tumor is the Brown's sign or pulsation sign, given by the whitening of the retrotympanic pulsating reddish tumor mass, secondary to the emission of a positive pressure (pneumatic otoscopy).

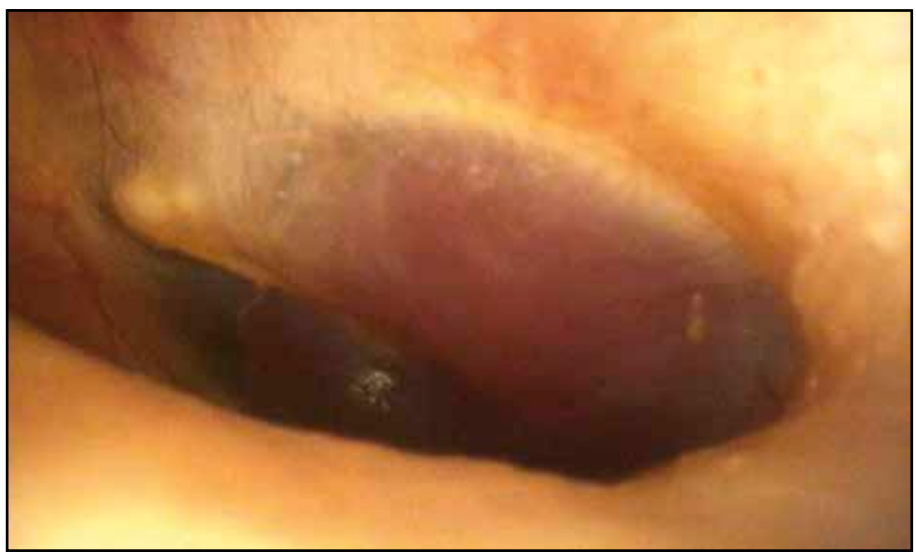

Figure 1. Reddish tumor formation highlighted by the transparency of the tympanic membrane.

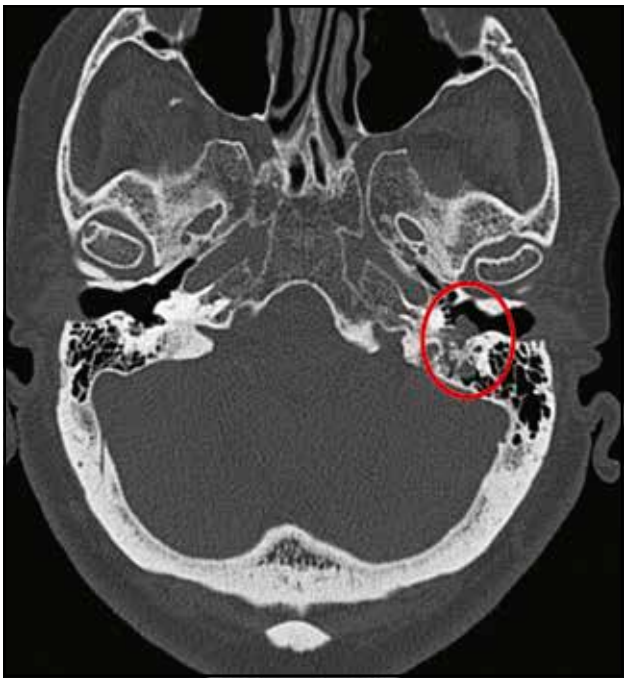

Figure 2. Cranio-facial CT scan, axial slice, left jugulotympanic paraganglioma.

Aquino's sign can be noticed by the manual compression of the ipsilateral carotid artery by decreasing the tumor pulsations and whitening it[15].

Otoscopy alone is not sufficient to evaluate the extent of the tumor, most significantly related to the degree of hypotympanic extension. Paragangliomas may be confused with inflammatory polyps, because they can extend through the tympanic membrane.

Assessment for cranial nerve dysfunction, possible signs and symptoms related to catecholamine secretion, as well as evidence of malignant transformation can be also considered. In patients with advanced disease, lower cranial nerve insufficiency may become visible.

Laboratory tests

Routine laboratory tests are not significant. In some patients with functioning tumors, preoperative and postoperative catecholamine measures may help to confirm complete resection of the tumor[1].

Diagnostic imaging

Contrast computed tomography, magnetic resonance imaging and angiography in combination are ideal for a good diagnosis and localization of the paragangliomas.

Computed tomography (CT) (Figure 2) shows the bone erosion and helps to differentiate the jugular glomus from the tympanic one. Jugular paragangliomas are seen within the jugular foramen, they are enhancing soft-tissue masses at the skull base, but skull-base artifact can mask their presence. The bony destruction of the jugular foramen and petrous apex is often a significant finding for the diagnosis. For this, it is essential to check the bony windows[1,16].

Magnetic resonance imaging (MRI) may show intense tumor enhancement and it is a useful finding for the diagnosis and to discover the degree of intracranial involvement. In jugulotympanic paragangliomas, a salt-and-pepper vascular pattern may be observed, characteristic for intrinsic tumor neovascularity (T2-weighted). Coronal sections may show relationships between the paraganglioma and the skull base, the brainstem, the deep structures of cervical soft tissue. MRI with gadolinium contrast is a complementary imaging tool that can be helpful for assessing head and neck paragangliomas, the tumors presenting pronounced enhancement of the signal. The role of magnetic resonance angiography has lim-

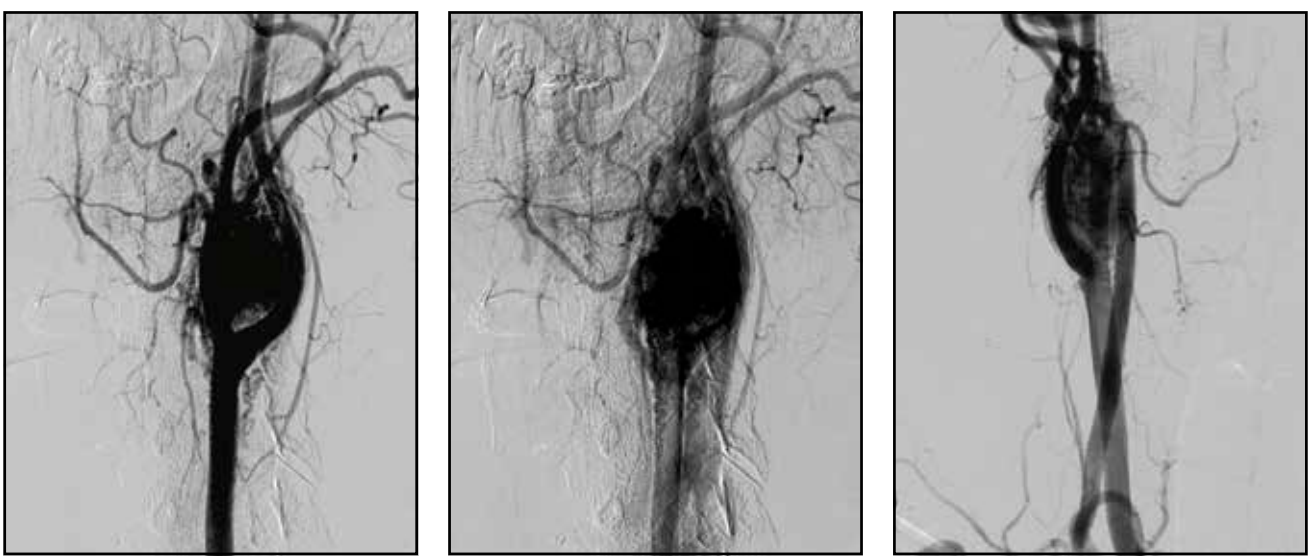

Figure 3. Angiography – aspect of a glomus tumor.

Table 1. Fisch classification of glomus tumors[19].

Tympanomastoid paragangliomas	**Class A** Tumors limited at the middle ear.	**A1**	On otoscopy, the margins of the tumor are well visible.
		A2	On otoscopic examination, the margins of the tumor are not visible. Tumors can spread to the posterior mesotympanum or anterior to the Eustachian tube.
	Class B Tumors circumscribed at the tympanomastoid cavity without bony destruction in the compartments of the temporal bone.	**B1**	Tumors localised at the middle ear spreading to the hypotympanum.
		B2	Tumors localised at the middle ear with spread to the mastoid and hypotympanum.
		B3	Tumors circumscribed at the tympanomastoid cavity, but with erosion of the carotid canal.
Tympanomastoid paragangliomas	**Class C** Tumors spreading outside the tympanomastoid cavity, damaging the apical section of the temporal bone and infralabyrinthine compartment, affecting the carotid canal.	**C1**	Tumors restricted to the vertical portion of the carotid canal.
		C2	Tumors invading the vertical portion of the carotid canal.
		C3	Tumors with invasion of the horizontal portion of the carotid canal.
		C4	Tumors reaching the anterior foramen lacerum.
	Class D Tumors spreading intracranially.	**De1**	Tumors up to 2 cm dural displacement.
		De2	Tumors with more than 2 cm dural displacement.
		Di1	Tumors up to 2 cm intradural extension.
		Di2	Tumors with more than 2 cm intradural extension.
		Di3	Tumors with inoperable intradural extension.
	Class V Tumors affecting the vertebral artery.	**Ve**	Tumors affecting the extradural vertebral artery.
		Vi	Tumors affecting the intradural vertebral artery.

Table 2. Glasscock - Jackson classification of glomus tumors[6].

Glomus tympanicum	**Type I**	Small tumor localised in the promoter
	Type II	Tumor that completely occupies the middle ear
	Type III	Tumor that occupies the middle ear and spreads to the mastoid
	Type IV	Tumor that occupies the middle ear, spreads to the mastoid or in the external auditory canal through the tympanic membrane; it may spread anteriorly to the carotid artery.
Jugular paraganglioma	**Type I**	Small tumor localised at the jugular bulb, middle ear and the mastoid
	Type II	Tumor that spreads in/ under the external auditory canal; it may invade the endocranium
	Type III	Tumor with extension in the petrous apex; it may have endocranial invasion
	Type IV	Tumor that exceeds the petrous apex, spreading into the infratemporal fossa or the clivus; it may have endocranial extension

ited importance in diagnosis, but it remains useful in preoperative planning for tumor resection. Obtaining superselective angiography and performing embolization of the arterial supply can decrease the risk of intraoperative blood loss. Radionuclide imaging can be used to target the biochemical pathways of catecholamine synthesis[17].

Angiography helps to demonstrate the feeding artery (Figure 3). Jugular tumors involve higher external carotid branch vessels; the ascending pharyngeal, tympanic and occipital arteries dominate the arterial blood supply. Also, arteriovenous fistulae may be discovered. Rarely, the internal carotid and vertebral arteries may supply feeders to the tumors. Usually, when these tumors are assessed, it should be paid attention to all potential feeding arteries. Angiography is important in the diagnosis and in the management of temporal paragangliomas. This technique proves the vascular feed of the tumor, the degree of internal carotid artery involvement, the contralateral cerebral blood flow, the venous drainage, help in preoperative embolization in case of surgery[18].

Currently, in the literature, there are two classification systems accepted for the glomus tumors: Fisch classification[19] and Glasscock Jackson classification[6].

TREATMENT OF JUGULOTYMPANIC PARAGANGLIOMAS AND FUNCTIONAL RECOVERY OF SECONDARY DEFICITS

Therapeutic options for head and neck paragangliomas are as follows: surgical treatment with endovascular embolization, conventional radiotherapy, stereotactic radiosurgery or a combination of these therapeutic methods.

Surgical treatment has been for a long time the first choice in the therapeutic management of the head and neck paragangliomas. Observing the multiple postoperative functional deficits due to cranial nerve damages, more conservative therapeutic methods have been attempted in recent years, such as cyber knife radiosurgery or linear accelerator stereotactic radiosurgery (LINAC) and gamma knife stereotactic radiosurgery, minimally invasive methods, that have been shown to have increased accuracy, with a decrease in postprocedural cranial nerve damages[5,20].

Stereotactic radiosurgery with linear accelerator with a higher energy that penetrates deeper, thus reaching a higher dose at the level of the paraganglioma and a lower dose at the bone level, showed to have a tumor control of up to 99%[21].

Gamma knife/cyber knife radiosurgery proved to have good local control, with the protection of the surrounding tissues. Post-procedural cases were not identified with aggravation of neurological deficits or the appearance of new lesions, but there were improvements or stabilizations of cranial nerve injuries. The results of cyber knife radiotherapy were like those after gamma knife radiotherapy. Clinically, patients showed improvements or were stable, while the complications were transient or mild[22].

The surgical approach can be transcanal for small lesions located in the tympanum. For more extensive lesions, the transmastoid approach or hypotympanotomy is recommended. When the tumor erodes the bony part of the carotid artery canal, the approach through the infratemporal fossa is necessary[23].

Small tympanic glomus (type I from Glasscock classification, type A from Fisch classification) can be resected by minimally invasive transcanal endoscopic approach[24]. Larger tympanic glomerular tumors are resected by microscopic approach, requiring 2 hands for hemostasis[25].

Because glomus tumors are well vascularized, angiography and sometimes preoperative embolization are performed to reduce size and minimize the risk of bleeding. Embolization has a risk of heart attack and cranial nerves paralysis may occur due to anastomoses between the internal and external carotid artery.

Biopsy should be avoided for patients with jugular paraganglioma due to potential bleeding.

Despite the various therapeutic options, currently there is no consensus for the treatment of jugulotympanic paragangliomas. Due to the neurologic disorders that can occur after surgery, an alternative therapeutic approach should be taken into consideration, especially in the case of young patients and small tumors. For example, external radiation therapy may diminish the tinnitus in patients with jugulotympanic paragangliomas. Cummings et al. performed a study on 45 patients with temporal bone paragangliomas who underwent external radiotherapy and they noticed partial improvement of tinnitus in 21% of patients and absence of tinnitus in 79% of patients. Also, sensorineural hearing loss was improved in 30% of patients[26]. Fractional radiotherapy has been used in residual or recurrent tumors in elderly patients with good local results, but with damage to the surrounding tissues. After conventional radiotherapy, it is important to note that complications can occur in up to 20% of cases, such as mucositis, temporal osteonecrosis, mastoiditis, parenchymal radionecrosis, alopecia.

Gamma knife radiosurgery is safe and has good long-term outcome, with lower cranial nerve lesions. It should be as a first line treatment option in patients with jugulotympanic paragangliomas[27].

After surgical treatment of jugular and tympanic paragangliomas, patients may experience pharyngolaryngeal disorders secondary to total vagal paralysis, such as incompetence of the velopharyngeal sphinc-

ter, paralysis of the vocal cords, impaired laryngeal ascension, glottic insufficiency, anterior subluxation of the arytenoid, loss of laryngeal sensitivity, paralysis of the pharyngeal constrictors, lack of coordination of the upper esophageal sphincter.

For the neurologic deficits, specific rehabilitation should be promptly initiated. Among the principles of swallowing therapy, we mention the learning of compensatory methods to protect the airways and ensure the transit of the bolus, compensatory mechanisms for pathophysiological changes (impairment of the movement of the tongue and pharyngeal muscles movement, reduction of ascension and laryngeal closure, reduced opening of the upper esophageal sphincter). For recovery from severe dysphagia, the following surgical options have been described in patients who have not had swallowing therapy: palatoplasty, arytenoidopexy with medialization and vocal cord lift, vocal cord medialization, vocal cord augmentation, upper esophageal sphincter myotomy, pharyngoplasty laryngeal suspension.

A study conducted by Cheesman et al. investigated the postoperative deglutition outcomes in patients with jugulotympanic paragangliomas[28]. The methods of medialization of the vocal cords had unsatisfactory results and the practice of arytenoidopexy was chosen. Laryngeal ascension and upper esophageal myotomy have been performed in some patients. In the upper esophageal myotomy, the incision along the entire length of the upper esophageal sphincter was chosen in favour of the cricopharyngeal myotomy, which, although more frequently described, is more limited. Indications for surgical approach for swallowing recovery were residues in the oral cavity or valecules, secondary to tongue weakness by unilateral damage to the hypoglossal nerve with contralateral compensation, decreased pharyngeal clearance, impossibility or reduction of the tongue contact with the pharyngeal posterior wall, the need for multiple swallowing. The weakness of the pharyngeal constrictor with the reduction of the pharyngeal shortening during swallowing was compensated in most cases by swallowing therapy, but one case remained with significant deficiency and surgical treatment was performed. The optimal time for the surgical approach in swallowing recovery is about 6 months, to see the effects of swallowing therapy and neurological compensation. After 3 months of continuous swallowing therapy, signs of functional recovery should be observed[28].

Oestreicher-Kedem et al.[29] described the surgical principles of swallowing and speech recovery after surgical treatment of jugular foramen tumors. The study included 21 patients, 14 of whom had glomus jugular tumors, 5 schwannomas and 2 meningiomas. 8 patients showed damage to the cranial nerves VII, IX, XII. Postoperatively, there were 18 patients with cranial nerve palsies VII, IX, XII. 3 patients had new postoperative nerve lesions in addition to the preoperative ones. Iatrogenic vagus nerve palsy resolved spontaneously in 2 patients (2 weeks, respectively 2 years postoperatively). Among the patients with paralysis of the glossopharyngeal nerve, 3 patients recovered their nervous function, appearing at 2 weeks, 1 year and 2 years after surgery. There was no evidence of functional recovery in patients with preoperative paralysis of the nerves VII, IX, XII. In 8 patients, surgery was performed, in which laryngoplasty was performed by collagen injection, thyroplasty, arytenoid adduction, cricopharyngeal myotomy. Patients who required surgery for functional recovery had large tumors[29].

R. Laskawi and S. Rohrbach highlighted some conservative and surgical methods for recovering the motor function of cranial nerves VII, IX, XI[30]. Functional recovery of the facial muscles of the paralyzed mimicry can be achieved through physical exercises such as electromyographic feedback training in the early postoperative period after hypoglossal-facial anastomosis.

Surgical methods for the functional recovery of paralyzed facial muscles may be direct anastomosis of the central and peripheral ends, in case of iatrogeny, performed within one month from the injury. Neural grafts can be used to fill defects in a peripheral nerve. The sural nerve is the most used for large deficiencies and the auricular nerve for short portions. The hypoglossal facial nerve graft has been used frequently in recent years.

Conservative methods of recovery in accessory spinal nerve palsy are physiotherapy, with facilitation of physiological movements of the cervical spine and perception of the spatial orientation of the head. As a surgical method of functional recovery is the termino-terminal anastomosis of the accessory nerve or the graft interposition from the atrial or sural nerve.

Recovery of the motor function of the injured hypoglossal nerve can be achieved through vocal therapy, visual feedback or neuromuscular electrostimulation[30].

D Li et al.[31], in a recent study of 51 patients, present the long-term effects of surgical treatment of jugular foramen paragangliomas (total resection, subtotal resection, partial resection). Postoperatively, patients had the following functional complications: 25.5% of patients with new facial paralysis, 19.6% with dysphagia, 15.7% with dysphonia, 5.9% with hearing loss, accessory nerve dysfunction in 5.9% of cases and impairment language mobility in 13.7% of cases. Compared to the preoperative condition, swallowing improved in 13.7% of patients and worsened in 3.9%. Grade I / II facial paralysis was present in 84.3% of cases and grade III / IV in 15.7% of cases. Facial nerve palsy with acute onset, immediately postoperatively was observed in 29.4% of cases, facial nerve function improved compared to the preopera-

tive status in 3.9% of cases. Long-term facial nerve function improved in 31.4% of patients, did not change in 64.7% and worsened in 5.9%[31,32].

Duzlu et al.[33], in a study of 34 patients, 18 with tympanomastoid paraganglioma and 6 patients with tympanic-jugular paragangliomas, presented the effects of surgical treatment of these diseases. Preoperatively, 8.8% had facial paralysis, 63.7% hearing loss, 2.9% dysphagia. Postoperatively, 31% had facial paralysis. In the long term, most patients with facial paralysis recovered reaching grade II and III House Brackmann (HB). One year postoperatively, patients with grade IV facial paralysis recovered from grade II HB facial paralysis. Postoperatively, 3 patients remained with cophosis and one patient with X and XII nerve palsy[33].

González-Orús Álvarez-Morujo et al.[34] performed a retrospective study on 126 patients with jugulotympanic paragangliomas, 45% of them presenting lesions of the cranial nerves IX, X, XI, XII. Considering the function of the facial nerve, a comparison was made regarding the functional effects after nerve transposition (28 patients) and without nerve transposition (9 patients). Of those who did not undergo transposition of the facial nerve, 44% had facial impairment grade I HB, 22% grade II, 33% grade III. In 64% of patients who required facial nerve transposition, they had facial paralysis grade III HB or more.

Surgical interventions for facial aesthetic recovery, for the recovery of swallowing and phonation, were required in 56 patients. 41 patients required surgery to recover swallowing and phonation – medial thyroplasty (15 patients), intracordal injection of autologous fat (16 patients), injection of botulinum toxin into the cricopharyngeal muscle (7 patients), myotomy of the cricopharyngeal muscle (3 patients), hyaluronic acid in the soft palate (1 patient). 5 patients required surgery for facial cosmetic recovery by injection of hyaluronic acid on the eyelid and 10 patients benefited from the implantation of a weight on the upper eyelid[34].

OUR EXPERIENCE

In order to emphasise the importance of correct diagnosis, proper treatment and postoperative complications and postoperative functional recovery in case of glomus tumors, we present the case of a 35-year-old female who was diagnosed and surgically treated for a right tympanojugular paraganglioma. She presented to the ENT specialist for right hypoacusis, fullness sensation and pulsatile tinnitus in the right year, symptoms which occurred approximately one year ahead.

The clinical ENT and otoendoscopic examination revealed a reddish-blue, pulsatile mass behind the right tympanic membrane, which was intact, with no clinical manifestations of associated facial nerve palsy. The pure-tone audiometry showed a mild hearing conductive hearing loss in the right ear (Figure 4).

The cervical and cerebral MRI, native and contrast-enhanced, revealed an extra-axial mass developed in the right temporal bone, with maximum dimensions of 28/35/33 mm, centered on the right jugular foramen, where it occupies entirely the pars vascularis, with its widening and modelling of the adjacent bone by pressure atrophy; the formation has minimal exobasal development in the carotid space, superiorly evolving towards the middle ear cavity, which it partially and endocranially fills with minimal occupation of the internal auditory canal; it invades the sigmoid sinus, imprints the cerebellum without perilesional edema, causes the partial collapse of the IV ventricle; the formation is intensely vascularized with a spongy appearance with arterial afferents from the external carotid artery – branches from the occipital and posterior auricular arteries, but also from the internal carotid artery (Figure 5).

The patient also underwent angiography, by right femoral artery approach, which identified a voluminous jugulotympanic tumoral mass with intradural component on the posterior part of the right petrous pyramid, with vascularization from the posterior branch of the middle meningeal artery and ascending pharyngeal artery. All the vascular afferences were embolized using polyvinyl alcohol particles (PVA). A small part of the tumor was visible, vascularized by terminal branches of the right antero-inferior cerebral artery, which could not be embolized, due to the increased risk of ischemic lesions.

The symptoms, the clinical and imaging findings suggested a diagnosis of right jugulotympanic paraganglioma and it was decided that the best approach in this case would be surgical therapy, due to the tumor size and localization.

The patient underwent surgery in another ENT clinic from abroad, with removal of the extradural part and the one inside the internal auditory canal of the tumor. According to the medical documents, during the same intervention, the reconstruction of the right facial nerve by tarsorrhaphy was performed. The surgical cavity was obliterated with abdominal fat. Postoperatively, the patient showed a right facial nerve palsy. During hospitalization, the eye was protected with artificial tears, special eyeglasses and eyelid weight. Postoperative evolution of the patient was favourable, without complications. The sutures from the temporo-cervical wound and abdomen were removed 15 days postoperatively.

The histopathological exam of the surgical specimen revealed the aspect of a paraganglioma, con-

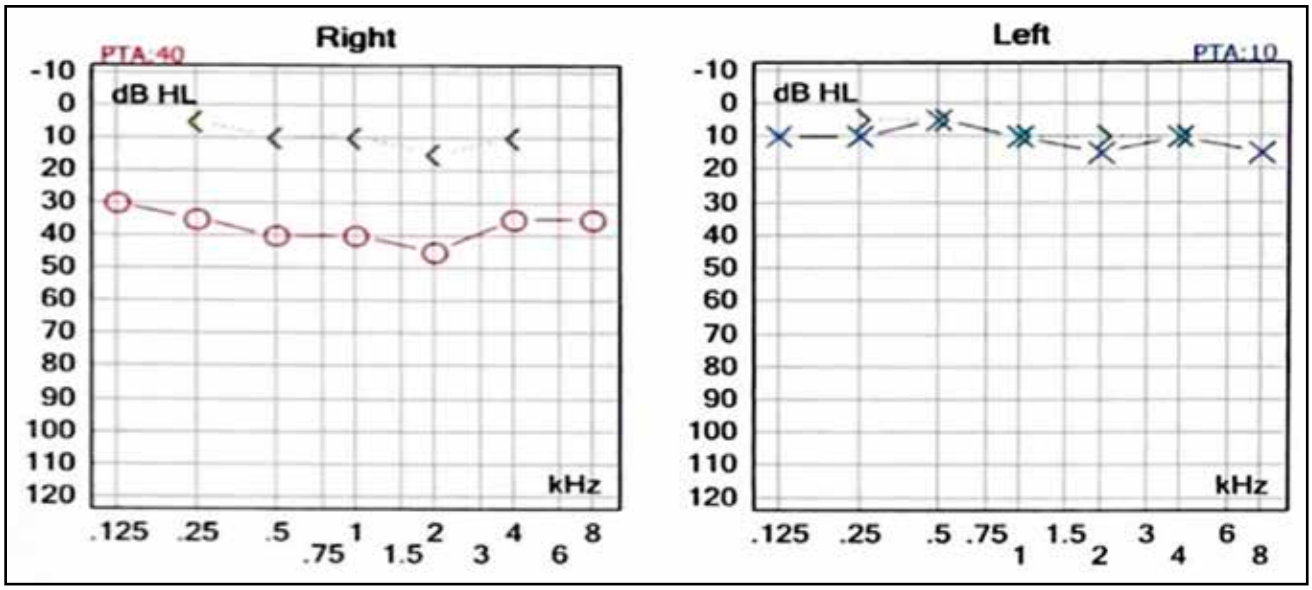

Figure 4. Pure-tone audiometry – mild conductive hearing loss right ear, normal hearing left ear.

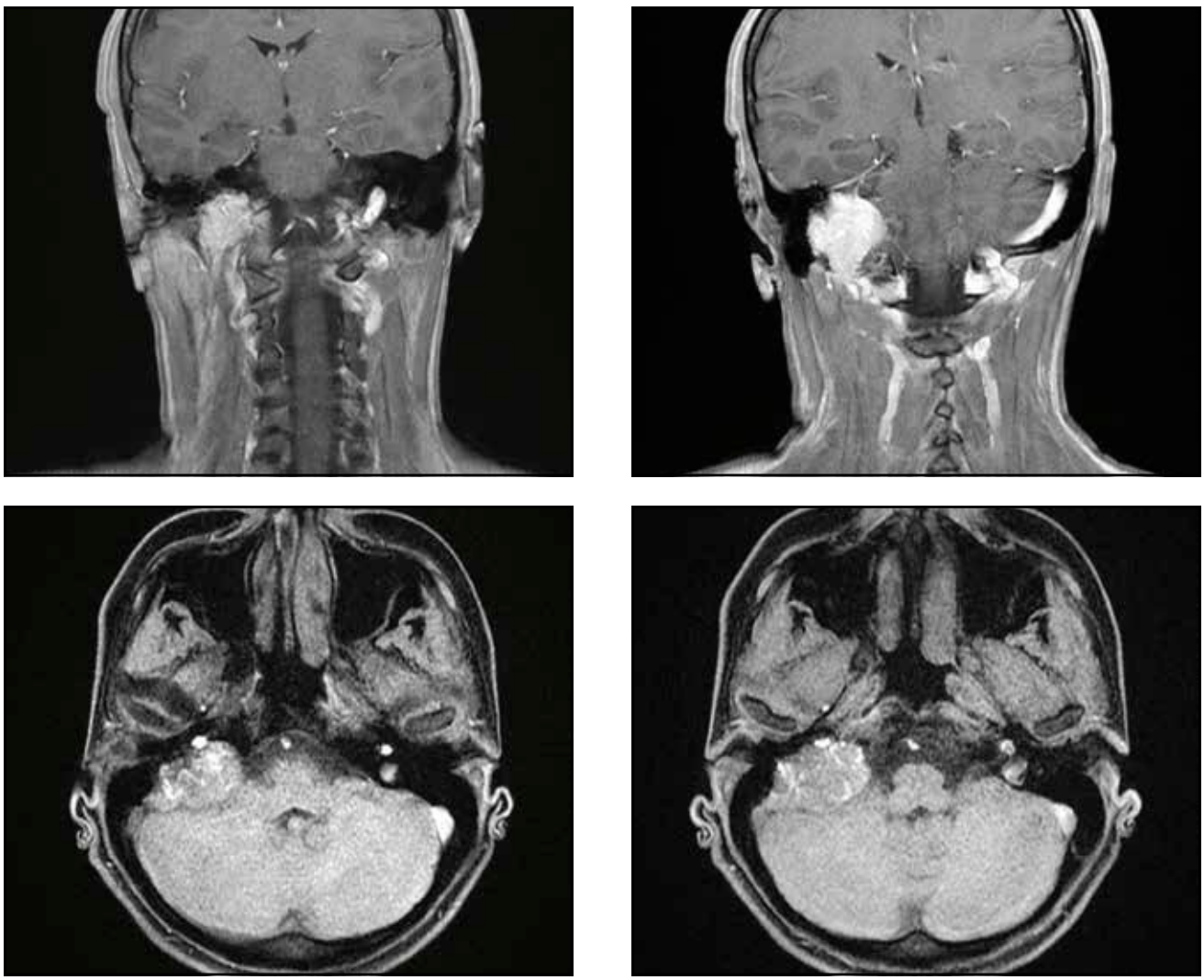

Figure 5. Cervical and cerebral MRI, native and contrast-enhanced, coronal and axial slices – extra-axial intensely vascularized mass in the right temporal rock, centered on the jugular foramen, extended towards the middle ear cavity, invading the sigmoid sinus.

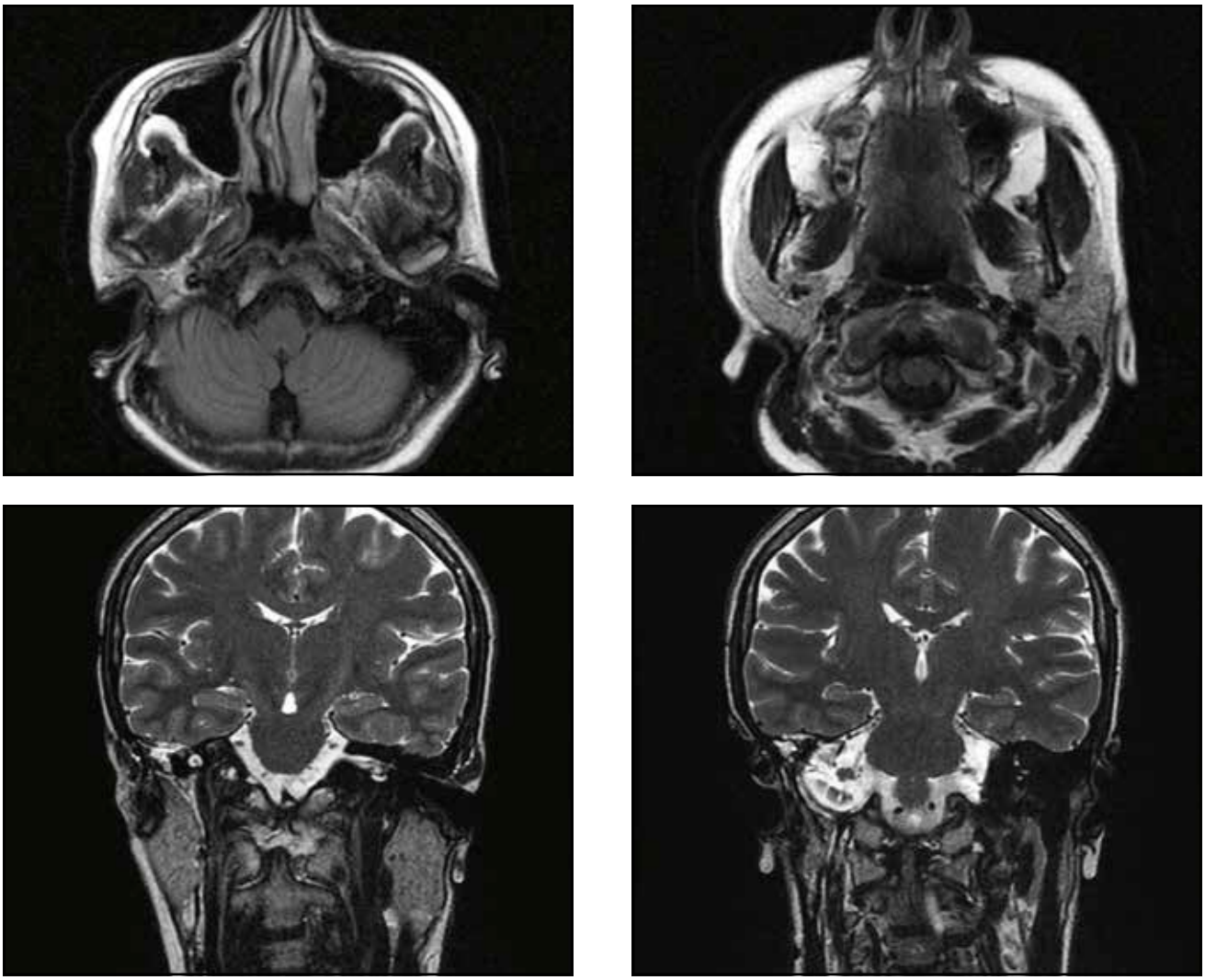

Figure 6. Head and neck MRI, 2 months postoperatively, axial and coronal slices – autologous adipose graft in the temporal bone, without intense contrast setting at this level; slight dural, linear contrast, at the ponto-cerebellar angle, possibly postoperatively, without any signs of recurrence.

firmed by immunohistochemistry using Synaptophysin, Chromogranin A, Protein S100, Vimentin and proliferation marker Ki67.

The patient underwent a control MRI two months postoperatively, which identified autologous adipose graft in the temporal bone, without intense contrast setting at this level; slight dural, linear contrast, at the ponto-cerebellar angle, possibly postoperatively, without any signs of recurrence (Figure 6).

Postoperatively, the patient remained with right ear cophosis, right facial nerve palsy, swallowing disorders, especially for liquids, and dysphonia. After the first surgical intervention, she underwent therapy in a recovery clinic for the right facial nerve – Winback therapy, accelerator for cellular regeneration, electrostimulation – 10-15 mA, for 20 minutes, pulsed monorectangular for 20 minutes, short waves.

Due to the persistence of facial asymmetry, despite rehabilitation treatment, in 2021, she underwent, in the same clinic, a right facial nerve reconstruction, by right masseteric-facial anastomosis surgery, under general anaesthesia. The stitches were removed 15 days postoperatively.

It was recommended for the patient to begin speech therapy 2 months after surgery, to strengthen the masseter muscle. The patient was admitted in our ENT Department two months after the second surgery. When she was admitted in our clinic, the ENT clinical evaluation, along with the naso-pharyngo-laryngeal fibroscopy and laryngeal videostroboscopy revealed right facial asymmetry, with lagophthalmos, ptosis of the left buccal commissure, impossibility of whistling, loss of right nasolabial fold, impossibility to smile or frown (Figure 7). According to the clinical aspect, the severity of the facial nerve was classified as House-Brackmann grade V.

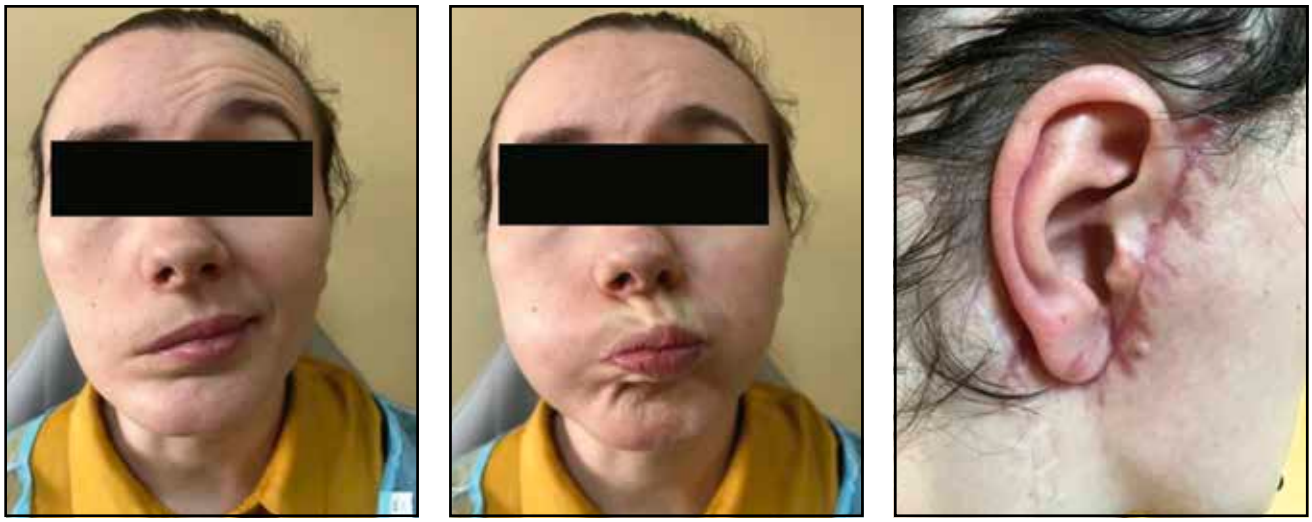

Figure 7. Clinical aspect 2 months postoperatively – facial asymmetry, with lagophthalmos, ptosis of the left buccal commissure, impossibility of whistling, loss of right nasolabial fold, impossibility to smile or frown; periauricular cicatricial tissue.

Also, there was cicatricial tissue in the right cervical region and the right external auditory canal was surgically excluded (Figure 7).

Other clinical findings were asymmetry of the right soft palate, right vocal fold palsy, with the vocal fold fixed in paramedian position, consecutive glottic insufficiency and minimal aspiration of secretions. The voice was breathy, and she used compensatory hyperfunctional manoeuvres to improve her voice quality, such as contraction of the left vocal fold and extrinsic laryngeal muscles, which later determined vocal fatigue. The patient also acquired compensatory techniques for the swallowing deficit – the chewing process took place only on the left side; the tendency to tilt the head to the left during the pharyngeal time of the swallowing process; the liquids were drunk with a straw. The postoperative cranial nerve injuries involved VII, IX and X nerves. The XI and XII nerves did not show any sign of lesion.

Due to the fact that the patient was a teacher, immediate treatment needed to be started, in order to reduce facial asymmetry, provide fast recovery of the facial muscular movement, improve swallowing and reduce the dysphonia. The patient was not able to attend her regular professional courses since she underwent the first surgery for the right paraganglioma.

She performed speech therapy, represented by breathing exercises, humming exercises and using

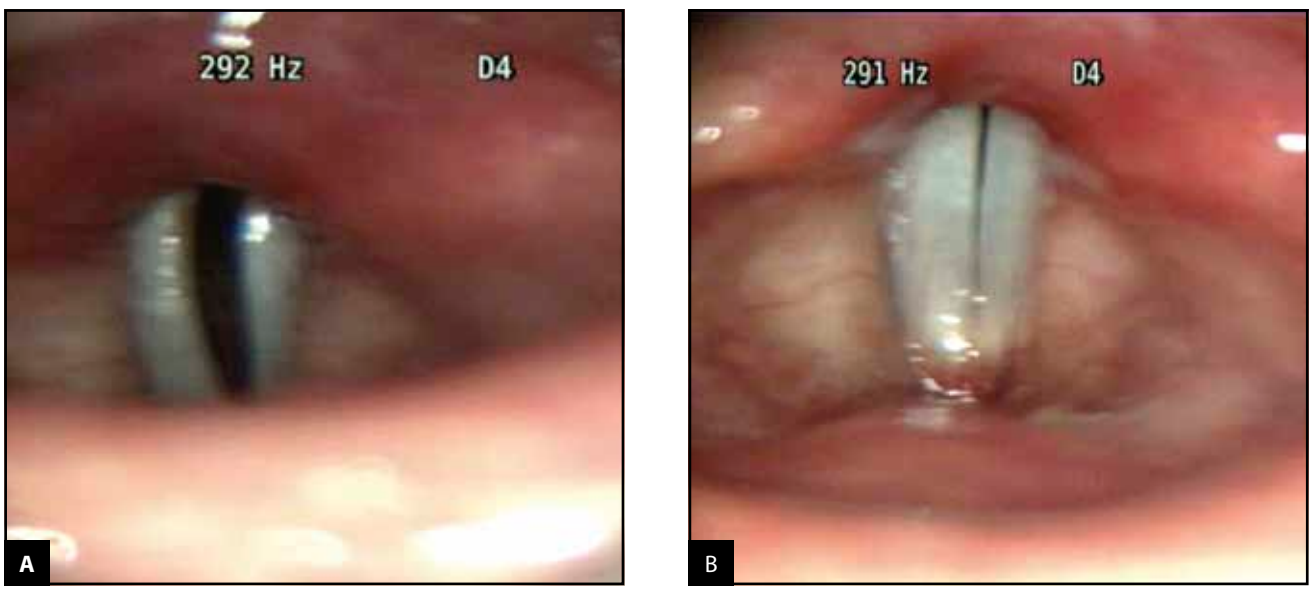

Figure 8. Laryngeal videostroboscopy **A**. Before vocal therapy – Glottal insufficiency, right vocal fold palsy; **B**. After vocal therapy – Normal glottal closure, right vocal fold palsy.

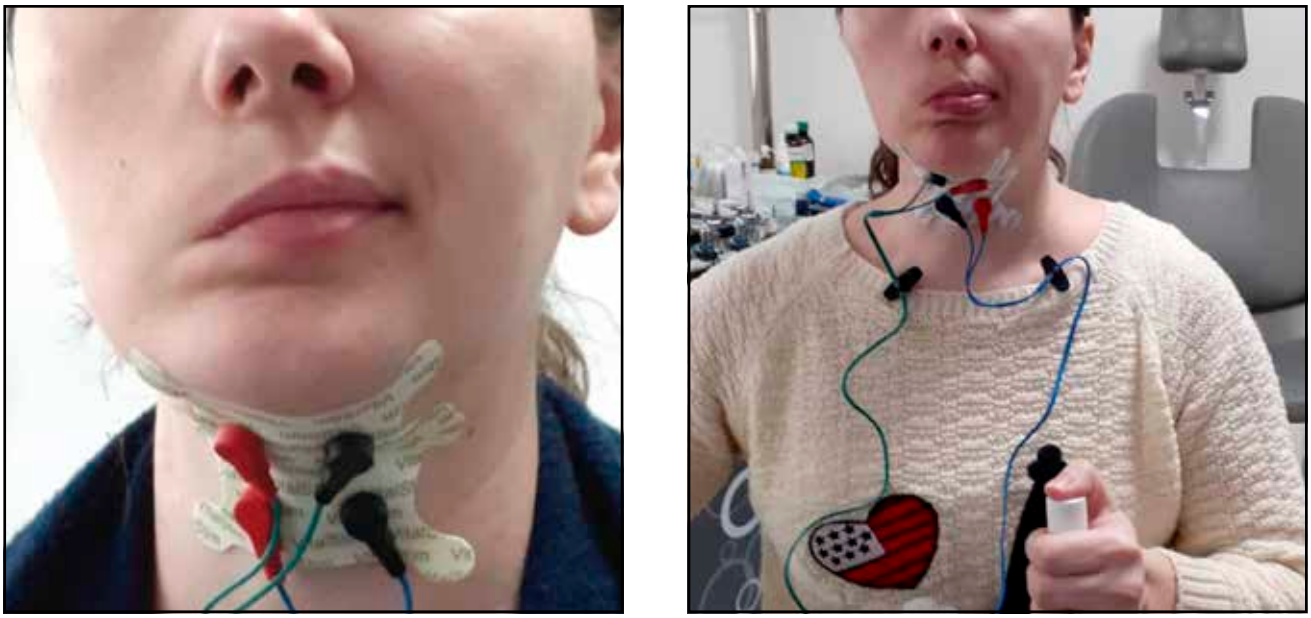

Figure 9. Electrode placement for swallowing exercises using VitalStim® Therapy System.

the Lax Vox technique, with semi-occluded vocal tract for 2 months, twice daily. Voice therapy was performed to strengthen the vocal cords, with improvement of the glottal closure, to strengthen breath control during speech, to reduce abnormal muscular contraction of the extrinsic laryngeal muscles and to prevent aspiration of secretions.

The voice was significantly improved, and the results were evaluated both subjectively, with Voice Handicap Index 10 (VHI-10) and objectively, by laryngeal videostroboscopy (Figure 8). The voice handicap index before the speech therapy was 22 and after 2 months it was 11. Although there was no clinical sign of vocal fold functional recovery, the glottic insufficiency was reduced and the patient did not present any signs of aspirations of secretions.

For the swallowing disorders, the patient performed specific exercises, using VitalStim® Therapy System (Chattanooga, USA), for 5 sessions, one session per week, 1 hour per session (Figure 9). For the therapy we used the 3b surface electrode placement in order to increase the tongue base retraction (CN XII), increase

Figure 10. Clinical status after 2 months of voice and swallowing rehabilitation, kinesiotherapy facial rehabilitation.

the pharyngeal constriction (CN X), increase the laryngeal elevation. Considering there were only 2 months since the facial nerve anastomosis, we avoided using direct electrical stimulation on the facial nerve. The neuromuscular electrical stimulation was performed with 5-7mA intensity and it was combined with home exercises. At the beginning and the end of the therapy, the patient was subjectively assessed with the Eating Assessment Tool (EAT-10), with an initial score of 26 and of 20 after the therapy sessions. After treatment, the deglutition was significantly improved.

The patient did not undergo physiotherapy for recovering the facial nerve and she was recommended to perform kinesiotherapy exercises to strengthen the facial muscles, by massaging the face and facial exercise. After two months of intensive treatment, the facial muscles activity was improved and incipient muscular contractions on the right side of her face were noticed and she started to smile. After interprofessional team strategies, facial nerve outcome was House–Brackmann grade III.

The ENT evaluation performed after two months of voice and swallowing rehabilitation, kinesiotherapy exercises, revealed: mobile soft palate, mobile tongue with fasciculation at rest; partial compensation for facial and spinal nerves; lagophthalmia persisted and wiping of the frown of the forehead. From a phonological and swallowing point of view, significant improvement was observed (Figure 10).

In this case, taking into consideration the imaging findings and the age of the patient, choosing the best therapeutic approach is complicated, due to the risk of postoperative impairments, that significantly interfere with the quality of life.

CONCLUSIONS

Lower cranial nerve deficits after surgical treatment of glomus tumors appear often and the patients need a good rehabilitative team to recover. Rarely, the patients with new postoperative palsies will recover the nerve function spontaneously. Patients with large tumors, usually need surgical rehabilitation methods and, therefore, therapeutic decision should be made carefully.

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