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CASE REPORT

Subtotal arytenoidectomy for the treatment of laryngeal stridor in multiple system atrophy: phonatory and swallowing results☆

Aritenoidectomia subtotal para o tratamento de estridor laríngeo na atrofia de múltiplos sistemas: resultados na fonação e deglutição

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Introduction

Multiple system atrophy (MSA), according to second consensus on MSA, is a neurological disorder characterized by a combination of autonomic failure and parkinsonism, or cerebellar ataxia, or both.[1] Among MSA manifestations, diurnal and nocturnal inspiratory stridor associated with sleep apnoea may help in clinical diagnosis, and its most accepted explanation is vocal folds abductor dysfunction. A possible complication of this situation is sudden nocturnal death. As described in the literature, the use of C-PAP and tracheotomy are the most common treatment proposed for the therapy of the laryngeal disfunction.[2] Alternatively laterofixation of the vocal fold according to Ejnell[3] or laser arytenoidectomy have been proposed. Here we present the case of nocturnal inspiratory stridor in a MSA patient treated with CO_2 laser subtotal aritenoidectomy, with particular attention on phonatory and swallowing outcome.

Case report

We report the case of a 60-year-old man with a 5-year history of rigid-akinetic syndrome, unbalance, mild orthostatic hypotension and Rem behaviour disorder. Since 2 years he presented cornage, marked weariness and dyspnea during night and daytime at rest. Pneumological and cardiological investigations were made without any clear diagnosis; cognitive tests did not show any impairment. The diagnosis of possible MSA-P was therefore made. During follow-up no more clinical feature was added, in particular the patient presented mild and slow worsening of motor and non-motor symptoms. He remained independent in activity daily living (ADL) and in activity instrumental daily living (IADL). The most life-threatening feature was the two-year history of snoring and sleep apnea with referred nocturnal stridor and occasional stridor during wakefulness. In February 2013, an endoscopic evaluation of larynx showed a reduced vocal fold abduction during inspiration with reduction of the breathing space. Vocal folds

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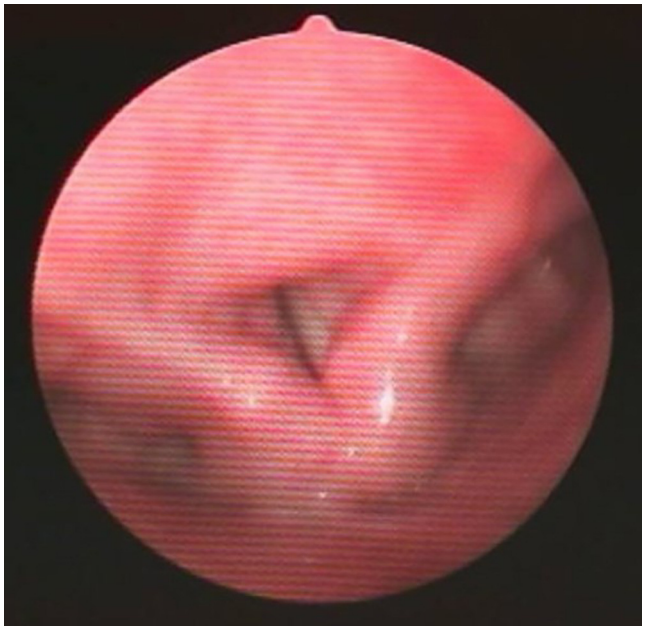

Figure 1 The endoscopy executed during sleep evidences the paradoxical movement of adduction during inspiration.

adduction was normal with no alteration of voice. A laryngeal electromyography was performed both on thyroarytenoid and cricoarytenoidal muscles: no denervation activity, but an alteration of maximum recruitment during phonation and deep breathing was observed. Polysomnography highlighted an OSA syndrome of mild grade, an apnea-hypopnoea index (AHI) of 9.2 with lowest SpO2 at 88%. Swallow evaluated with anamnesis and with endoscopic and radiological dynamic study of swallowing was normal. A Propofol-induced Sleep Endoscopy (PDSE) was performed using a low dose of propofol (0.01 mg/kg) followed by a titration of propofol (3 mg/kg/hr). The endoscopy revealed a paradoxical movement of adduction during inspiration (Fig. 1) with a marked inspiratory stridor, while expiratory abduction movement of the vocal folds was conserved.

A surgical treatment of the glottis was considered because the patient refused the hypothesis of C-PAP and any future tracheotomy. Perceptual evaluation of voice according to GIRBAS scale,[4] evidenced a mild alteration of voice (G1R0B1A0S0); the spectrographic examination carried out using CSL model 4500 B (Kay Elemetrics Corp.) by means of a narrowband filter of the prolonged vowels ''a'' and of the Italian word ''aiuole'' was classified in the second class, according to Yanagihara classification; finally, an examination of voice using the Multi-Dimensional Voice Program (MDVP) by Kay elemetrics showed alteration of frequency perturbation (Jitt – 1.564; RAP – 0.924; vF0 – 2.366), as well as amplitude perturbations (ShdB – 0.562; Shim – 6.246; vAm – 17.635) with a mild alteration of noise to harmonic ratio (NHR = 0.156) (Fig. 2). Maximum phonatory time (MPT) was 11 seconds. Self-evaluation of voice with voice handicap index (VHI)[5] was submitted to patient in order to measure physical, emotional and functional complaints of dysphonia; preoperatory score was of 5 (mild alteration). Then

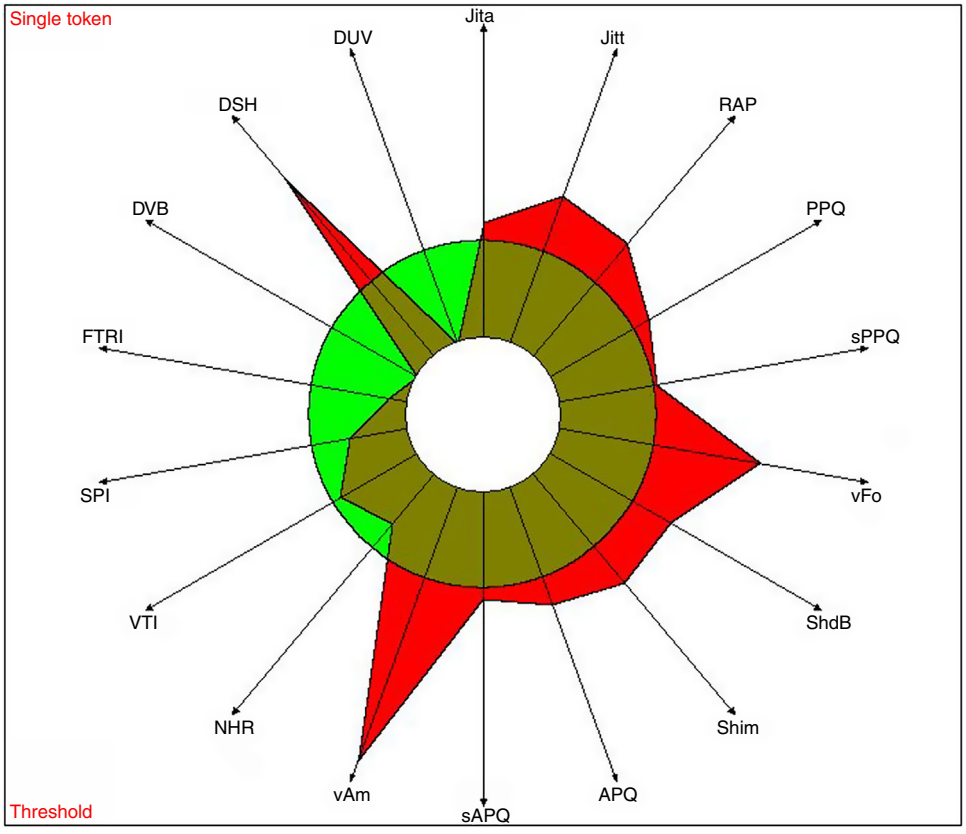

Figure 2 Multidimensional voice program (MDVP) underlines preoperatory perturbations in frequency and amplitude.

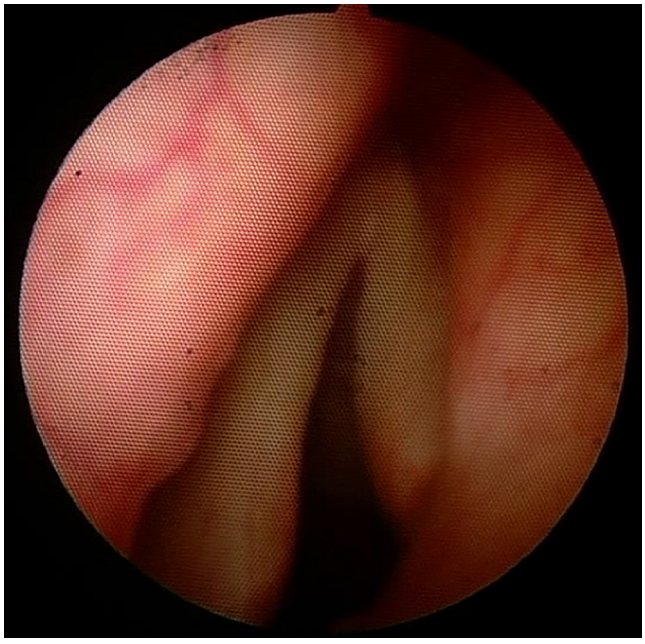

Figure 3 The control endoscopy executed at two months from surgery shows the good increase of the glottic respiratory space.

subtotal arytenoidectomy was performed using CO_2 laser at 6-10 watt in continuous mode according to Remacle technique,[6] with the resection of arytenoid body and the preservation of a small posterior shell of this cartilage. The two-months control endoscopy (Fig. 3) evidenced a good increase of the glottic respiratory space. Post-surgical perceptual evaluation of voice remained good (G1R0B2A0S0 – mild alteration), the spectrographic examination (Fig. 4) showed a mild worsening of NHR and a dyplophonia. At MDVP evaluation, all the above mentioned values were slightly worsened (Fig. 5), but no modification of voice quality was perceived by the patient and his post-surgical VHI had a score of 8 (mild alteration). MPT remained unchanged (11 seconds). Swallowing evaluated with PAS (penetration/aspiration scale) evidenced no signs of penetration/aspiration. Nocturnal stridor and dyspnea definitely improved. After two years, patient's condition is stable with no alterations in breathing and swallowing.

Discussion

Multiple system atrophy (MSA) is an adult-onset sporadic and rapidly progressive neurodegenerative disorder, characterized by autonomic failure associated with parkinsonian features and/or cerebellar ataxia and a wide variety of other clinical findings that rarely presents with predominance of respiratory disorders (respiratory failure or stridor).[1] In literature, few cases of MSA with prolonged duration of disease (more than 15 years) have been reported,[7] but it is not clear which symptom at onset, between dysautonomia and parkinsonism, correlates with slower progression.[7] The initial multisystem involvement or the short latency from one-system-disease to multiple-system-disease stage have been identified as bad predictor for disease progression and survival.[7] In our patient motor and disautonomic symptoms started simultaneously, whereas stridor appeared later. During the first 5-year follow-up, the dopaminergic treatment was not necessary, due to slight motor involvement. Stridor is defined as a harsh, strained inspiratory sound with a pitch at 260-330 Hz higher than snoring. It occurs during inspiration, reflecting an upper airway obstruction due to partial or complete vocal cord abduction impairment. Stridor, that is considered a red flag of MSA, has a prevalence of 34-41% in MSA and represents the opening feature in 4% of cases. It is considered a life-threatening condition, leading to subacute episodes of dramatic respiratory failure and death.[7] Two theories try to explain its etiopathogenesis: the first, ''respiratory center damage'' theory, attributes the MSA neurodegenerative process to abnormal outputs from the respiratory network that induce a selective paralysis of the abductor with relative preservation of adductor functions. The second one, ''reflex theory'', hypothesizes a paradoxical activation of the laryngeal closure reflex, that normally protects subglottic space from strong negative pressure; MSA-stridor would result not only from a passive glottis narrowing, attributed both to abductor paralysis and Bernoulli effect, but according to reflex theory,

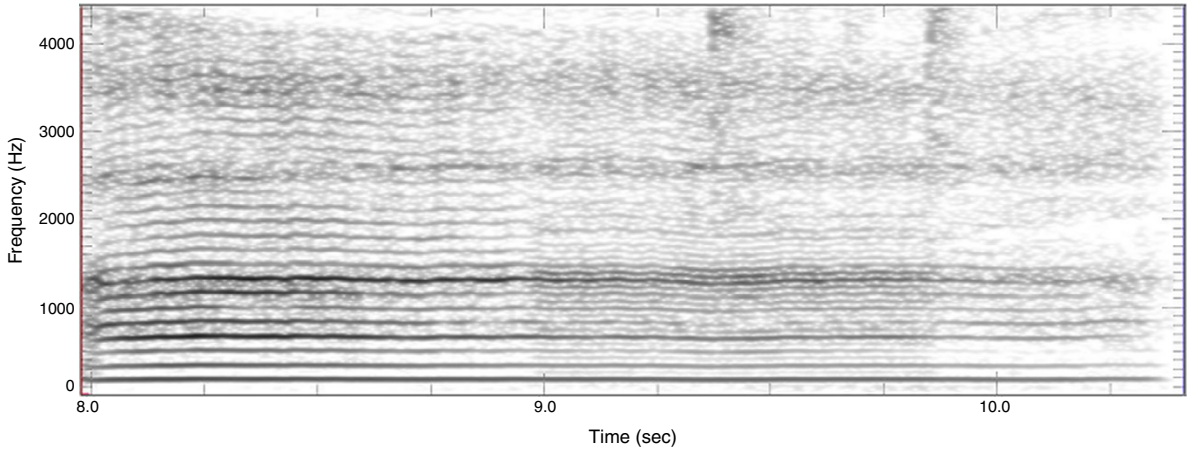

Figure 4 The spectrographic examination executed after surgery points out the presence of noise between the harmonics and of diplophony.

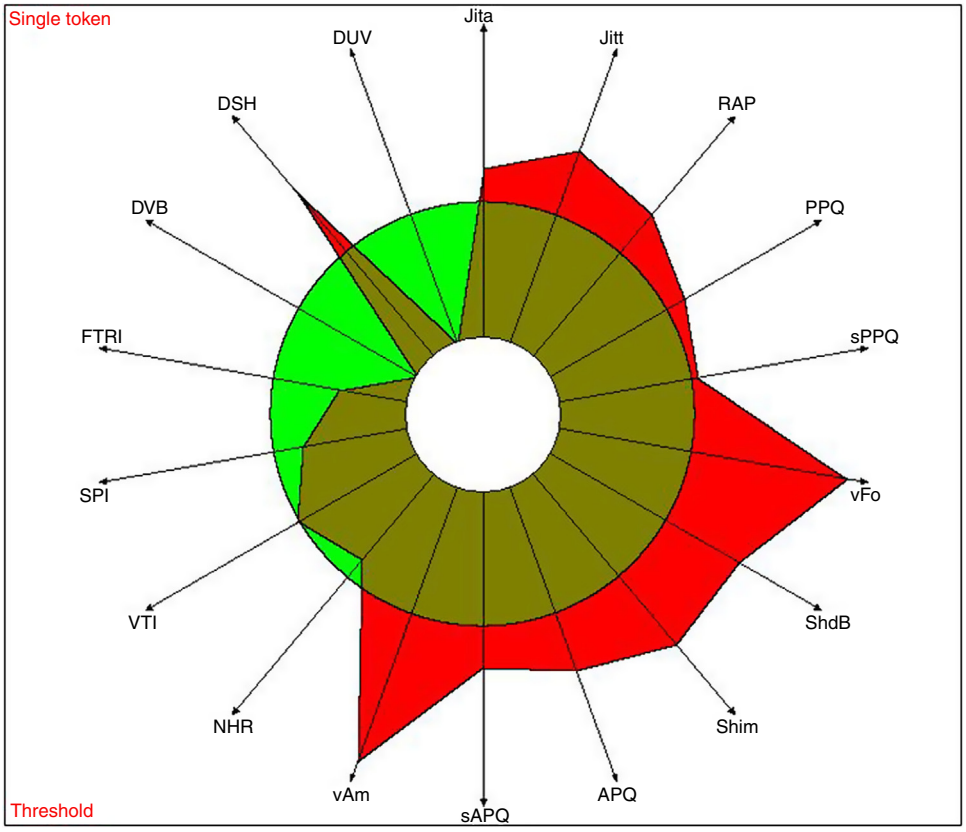

Figure 5 The multidimensional voice program evidences the postoperatory mild worsening of the perturbations in frequency and amplitude.

from an active narrowing of vocal fold. Hyper-activation of laryngeal closure reflex is triggered by increasing drop of pressure during voluntary inspiration. In MSA patients, the glottic space is reduced by passive forces, therefore laryngeal closure reflex builds up a vicious cycle that promotes an active vocal cords narrowing and, then stridor. In fact, once voluntary inspiration and negative airways pressure are erased (through tracheostomy or CPAP), stridor disappears.[2] Stridor and laryngeal dysfunction develop during the course of disease: in the first stages, during wakefulness, only a slightly impaired vocal cord abduction or flicker or ataxic movements of the vocal cords, and periodic or persistent involuntary adductions or abductions of the vocal cords may be present[2]; in intermediate and late stages, the restricted glottis and abductor paralysis develop causing daytime stridor.[2] Stridor associated with a decreased survival is the only independent predictive survival factor, but it is not the only one cause of sudden death in parkinsonian syndromes. CPAP and tracheotomy increase survival rates, although there are some reports of sudden death even after these treatments, probably due to central sleep apnoea. Laser treatment of the posterior glottis in adduction bilateral vocal fold palsy can be an appropriate way to solve the respiratory problem because, preserves the phonatory function, penalizing the posterior glottis whose main function is the respiratory one and whose influence is not determinant on voice quality.[8] Among the laser techniques, Remacle sub-total aritenoidectomy, while increasing the glottic respiratory space, ensures a good phonatory outcome, and with a good fixation of the arytenoid region, minimizes the risk of aspiration. We believe that, in selected cases of MSA with subtotal glottic pattern of restriction,[9] if respiratory stridor is present during wakefulness and the patient doesn't tolerate the C-PAP and refuses a possible tracheotomy, it is possible to perform a CO2 laser subtotal arytenoidectomy to restore an adequate airflow through the glottis. To our knowledge, in the literature only two authors[9,10] have investigated the surgical options for the treatment of glottic obstructions in MSA patients, but they proposed a complete removal of arytenoid cartilage with partial sacrifice of thyroarytenoid muscle or the Ejnell technique[3]; their evaluation of voice quality after was limited, in one case to Girbas scale, and, in the other one, to limited parameters of voice. Our evaluation based on self-perception (VHI), perceptive evaluation of voice quality (GIRBAS scale), on spectral analysis of phonatory results as well on evaluation of deglutition, shows that Remacle technique is effective and ensures only a mild worsening of voice quality, while a lower impact on swallowing is guaranteed compared to the previous proposals.

Conclusion

We recommend laser subtotal arytenoidectomy in MSA patients affected by nocturnal stridor due to a paradoxical adduction inspiratory movement, but selection of the patients is mandatory paying attention to dysphagia: if

alterations in the normal process of deglutition are present surgical procedures altering the glottic plane must be avoided.

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Conflicts of interest

The authors declare no conflicts of interest.

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