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Original article

Hemifacial spasm associated with other cranial nerve syndromes: Literature review

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ABSTRACT

Hyperactive dysfunction may affect all cranial nerves in the posterior fossa. According to literature review and personal experience, hemifacial spasm was found to be associated not only with the most frequent cranial nerve syndromes, namely: trigeminal neuralgia, vago-glossopharyngeal neuralgia or VIIIth nerve disturbances manifested by vertigo, tinnitus, hearing decrease, but also with rarer syndromes like geniculate neuralgia, masticatory spasm etc. Also, a number of publications have pointed out the relatively high incidence of the coexistence of hemifacial spasm and systemic blood hypertension; both can be cured by vascular decompression of the ventrolateral aspect of the medulla and IX-Xth route entry zone (REZ) together with the facial REZ. Even more complex clinical presentations have been encountered, corresponding to disturbances in several cranial nerve nuclei. Some could be attributed to neurovascular conflicts from elongated arteries invaginated into the brainstem, and cured by microvascular decompression surgery. When confronted with such complex, and therefore misleading, syndrome, it is advised to search for vascular conflicts at the brainstem using high-resolution MRI exploration.

Multiple cranial nerve (CN) hyperactive syndromes are rare. Their often complex clinical presentations are misleading prior to diagnosis; their mechanisms should be established with certainty. Well-documented reports on CN syndromes associated with hemifacial spasm (HFS) are limited. Coexistence of HFS and other CN pathologies have been occasionally described in case reports or mentioned within the framework of larger published series, which has made review of literature difficult.

The association of trigeminal neuralgia (TN) to ipsilateral HFS can be estimated at 1%, ranging from 0.5 to 3% of the patients affected with HFS according to literature [1]. We encountered association of HFS and TN in 3 patients (0.88%) in our series of 340 patients with HFS (unpublished data). For those patients, the vascular compression was related to an ectatic vertebrobasilar arterial system (Fig. 1) and symptoms were relieved by micro-vascular decompression (MVD) of both CNs. In some rare patients when HFS and TN are simultaneous the clinical presentation corresponds to the so-called "tic convulsif", as firstly described by Cushing [2]. We observed one such case in our series of 340 HFS operated on.

Vertigo and/or ipsilateral disturbances in hearing are rather frequently associated with HFS. As early as the seventies, the University of Montreal team pointed out the logical occurrence of their associations as follows: "during the investigations of patients presenting symptoms related to the Vth and VIIth nerves, frequently associated symptoms relevant to the VIIIth nerve were found, such as tinnitus, disequilibrium or vertigo. On the other hand, there also came to our attention patients whose major complaints were of VIIIth origin, especially of the vestibular component, and who occasionally presented associated symptoms of Vth or VIIth nerve involvement. Following the successful treatment of trigeminal neuralgia and hemi-facial spasm by liberation of the Vth and the VIIth nerves from the mechanical irritative lesion, frequently a vascular loop anomaly, it was postulated that the same cause could be responsible of tinnitus and vertigo secondary to irritation of the VIIIth nerve in some cases" [3].

In a review of HFS epidemiology, association of VIIIth nerve symptoms to HFS was reported to be 7.4%, ranging from 4.7 to 16% according to reports [1]. In our series of 340 patients referred for HFS and assessed for our report, association rate was at 6%. Briefly,

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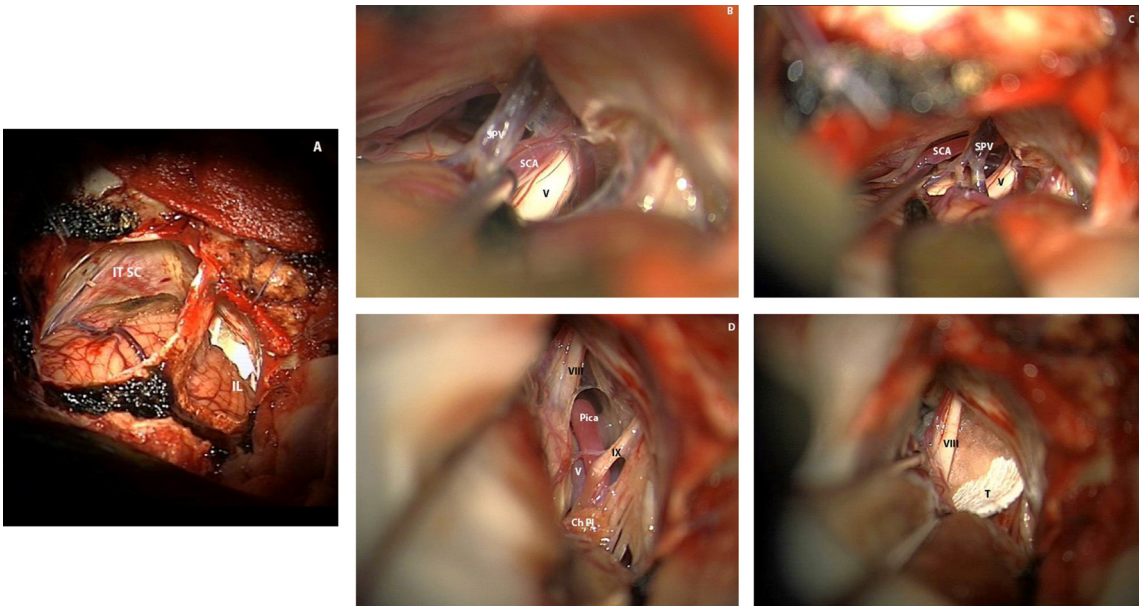

Fig. 1. Operative view of a patient who presented with the association of a right hemifacial spasm and a right trigeminal neuralgia. A. Approach of the right trigeminal cerebello-pontine angle via an infratentorial-supracerebellar (ITSC) route to access the trigeminal nerve, and via an inferolateral cerebellar (ILC) route to access the facial root exit zone (REZ). B. Trigeminal nerve (V) compressed by the superior cerebellar artery (SCA). SPV: superior petrosal vein. C. Trigeminal nerve (V) after decompression from the superior cerebellar artery (SCA), transposed upwards. SPV: superior petrosal vein. D. The cochlear-vestibular nerve (VIII) masks the facial nerve compressed at REZ by an atheromatous megadolicho-vertebral artery (VA) and the posterior-inferior cerebellar artery (Pica). Ch Pl: choroid plexus emerging from the lateral foramen of Luschka. IX: glossopharyngeal nerve. E. View after decompression by pushing away, inferolaterally, the VA and Pica, and inserting a piece of (knitted, rigid) Teflon (T) in between the facial REZ ventrally (not seen) and the VIIIth nerve dorsally on one side and the compressing vessels on the other side.

vertigo was favorably influenced by MVD in most cases whereas tinnitus rather not (unpublished data).

The cochleovestibular complex and its vascular supplies have an anatomical relationship so close with the facial nerve that a pre-operative otologic examination to search for vestibulocochlear disturbances is crucial. Exploration recommended by the American Academy of Otolaryngology-Head and Neck Surgery (AA0-HNS) are the following: Air and bone audiometry with calculation of Pure Tone threshold Averages (PTA), Speech Discrimination Scores (SDS) and if necessary BAEPs [4]. Also, depending on the symptoms, an assessment of vestibular reflexes is recommended [5].

Surprisingly, only very few cases have been published regarding the combination of geniculate neuralgia and HFS. We found only two such reports in the literature [6,7]. In both of them the elongated anterior inferior cerebellar artery that was considered responsible was transposed with complete relief of symptoms. The intermediate nerve was not sectioned. The presence of paroxysmal pain or even burning pain with a trigger area at the posterior wall of the external auditory canal should bring attention to this diagnosis according to the recommendation of the International Headache Society [8].

To our knowledge, there are only limited published studies on the dual association of vago-glossopharyngeal neuralgia (VGPN) and HFS. This is quite surprising as the vessels involved in the compression of the facial nerve at the Root Exit Zone (REZ) frequently have a close relationship with the Xth and IXth CN REZ before reaching the ventro-caudally located facial REZ. Wang et al. reported six patients with coexistence of HFS with TN and VGPN, three of them on same side [9]. In all of their patients, a small posterior fossa was found, which was crowded with CN roots and cerebellar vessels. All symptoms were relieved in all patients either immediately or secondarily after combined MVD. We observed one case of an association of HFS with VGPN in our series of 340 patients operated on for HFS (Fig. 2). Such a rare association could well be explained by the low incidence of VGPN in the patient population, in the order of one VGPN for one hundred TN.

In contrast, the association of blood hypertension (HT) to HFS has been the subject of a number of reported studies. Several authors have addressed the question of the prevalence of hypertension in patients with HFS compared with the general population of the same age range, with some authors investigating the side of the potentially responsible neurovascular compression (NVC) by means of MRI. While some studies showed a higher prevalence of hypertension in patients with HFS than in control groups [10,11], as much as in 67% of patients with HFS [11], others did not find any statistically significant differences [12–14]. Discrepancies between these studies might be due to bias in methodology: prevalence studies were retrospective and/or lacked proper matching with controls. In our global series of patients referred for surgery for treating their HFS, the percentage of patients with hypertension was 23.8% ($n = 48$) [15]. Hypertension was present in 26.11% of the patients with left-sided HFS and in 20% of those with right-sided HFS, which is not a statistically significant difference.

In a major study by Nakamura et al. there was no significant difference found in prevalence of hypertension between patients with HFS ($n = 82$) and a control group without HFS ($n = 82$, matched for age and sex): 39% versus 29.3% ($P = 0.19$) [16]. There was no difference in the prevalence of hypertension between the patients with left-sided ($n = 44$) and right-sided ($n = 38$) HFS: 32% versus 47%. However a significant difference was found in a MRI study when the side of the HFS and the presence or absence of hypertension were considered together. Vascular compression at the ventro-lateral medulla on imaging was observed in 86% of the patients with left HFS who had hypertension ($n = 14$) versus only 33% of the patients with left HFS without hypertension ($n = 30$) ($P = 0.0012$). There was no similar significant difference in patients with right HFS ($P = 0.18$). Nakamura et al. concluded that these findings were of clinical importance, at least for the patients harboring left HFS and hypertension, and also for those with essential hypertension alone provided that MRI shows vascular compression at the brainstem.

Furthermore, a large number of studies were launched to evaluate the validity of MRI to identify NVC that might be causing

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