Arq Neuropsiquiatr 2007;65(3-B):783-786

CLINICAL ASSESSMENT OF PATIENTS WITH PRIMARY AND POSTPARALYTIC HEMIFACIAL SPASM

A retrospective study

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ABSTRACT - *Objective:* To compared the clinical features of 373 patients with primary and postparalytic hemifacial spasm (HFS). *Method:* Data analyzed were gender, ethnicity, age at symptom onset, disease duration, affected side, distribution of facial spasm at onset, hypertension, family history of HFS, previous history of facial palsy and latency between facial palsy and HFS. *Results:* The prevalence of patients with Asian origin was similar in both groups such as female/male ratio, mean age at symptom onset, disease duration, affected side and distribution at onset of facial twitching. The upper left side of the face was the main affected region at onset. Almost 40% of the patients in both groups had hypertension. A prevalence of vascular abnormalities on the posterior fossa was seen in 7% and 12.5% of both groups. *Conclusion:* The clinical profile and radiological findings of patients with primary and postparalytic HFS are similar. The association of hypertension with vascular abnormalities and HFS was not frequent.

KEY WORDS: hemifacial spasm, facial palsy, neuroimaging, neurovascular contact, hypertension.

Análise clínica de pacientes com espasmo hemifacial primário e pós-paralítico: estudo retrospectivo

RESUMO - *Objetivo:* Comparar características clínicas de 373 pacientes com espasmo hemifacial (EHF) primário e pós-paralítico. *Método:* Os dados analisados foram: sexo, etnia, idade no início dos sintomas, duração da doença, lado afetado, distribuição dos espasmos no início da doença, hipertensão arterial, história familiar de EHF, história prévia de paralisia facial periférica e latência entre a paralisia facial e o início do EHF. *Resultados:* A prevalência de pacientes com origem asiática foi semelhante em ambos os grupos assim como razão homem/mulher, média de idade no início dos sintomas, duração da doença, lado afetado e distribuição dos espasmos no início dos sintomas. O quadrante superior esquerdo da face foi o lado mais afetado no início dos sintomas. Quase 40% dos pacientes em ambos os grupos tinha hipertensão arterial. A prevalência de anormalidades vasculares na fossa posterior foi observada, respectivamente, em 7% e 12,5% dos casos. *Conclusão:* O perfil clínico e também radiológico dos pacientes com EHF primário e pós-paralítico foi semelhante. A associação entre hipertensão arterial e anormalidades vasculares na fossa posterior não foi freqüente.

PALAVRAS-CHAVE: espasmo hemifacial, paralisia facial, neuroimagem, contato neurovascular, hipertensão arterial.

Hemifacial spasm (HFS) is a common movement disorder characterized by involuntary tonic or clonic contractions of peri-ocular and perioral musculature innervated by the ipsilateral facial nerve. It can be classified according to the underlying pathology either into primary or secondary HFS[1].

Few epidemiological studies have estimated the prevalence of HFS[2,3] and it is believed that patients with Asian origin are at greater risk for developing HFS. There is also few data on the literature comparing clinical characteristics of patients with primary and secondary HFS[4].

Thus, our study was aimed to compare clinical features of a tertiary care-center population with primary and postparalytic HFS.

METHOD

Patients and data acquisition – This was a retrospective study (medical record review) from 1985 to 2005 of 373 consecutive Brazilian patients with primary or postparalyt-

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Received 29 January 2007, received in final form 19 April 2007. Accepted 8 June 2007.

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ic HFS were seen in the Movement Disorder Unit (MDU), Escola Paulista de Medicina. The clinical data recording over these 20 years was done in a systematic manner, using a standard protocol. Even though different physicians of varying degree of experience participated on data collection, each case on the first consultation and subsequent follow-up was obligatorily discussed with a movement disorder specialist due to the academic nature of our institution. Incomplete or poorly recorded files were excluded from statistical analysis.

Clinical data – The aim of this study was to compare clinical features of patients with primary or postparalytic HFS using the following data: gender, ethnicity (Caucasian-Brazilian, African-Brazilian or Asian-Brazilian), age at symptom onset, disease duration, affected side, distribution of facial spasm at the onset, triggering factors, family history of HFS, previous history of peripheral facial palsy, latency between peripheral facial palsy and the development of HFS. The prevalence of hypertension was also analyzed reviewing clinical records on the presence or absence of this comorbidity, on the pharmacologic treatment for high blood pressure and on the general physical exam (measurement of blood pressure under standard conditions). Patients with other facial movement disorders such as tics, cranial dystonia or myokymia were excluded such as patients with bilateral HFS (2.6%; 10 patients).

Neuroimaging and therapeutic data – Although it was not the purpose of this study, we listed neuroimaging findings focusing on vascular abnormalities seen on the posterior fossa for patients who underwent scans (computed tomography - CT and magnetic resonance imaging - MRI). Due to the retrospective nature of this study imaging techniques used were not optimized to detect these abnormalities. All patients who underwent MRI were scanned on the same machine mostly from the years 2000 to 2005. Three different machines of CT were used to scan our patients over this 20 year period. We also listed the oral medicines or botulinum toxin type A (BTX-A) patients from this series took. The outcome of the different pharmacological treatments was not assessed.

Statistical analysis – Differences between groups were assessed using the Student's *t* test or the Chi-square test and were considered statistically significant if $p<.05$.

This protocol was submitted to and approved by the Local Ethical Committee.

RESULTS

The main demographic features of both groups (primary and postparalytic HFS) are presented on Table. The largest group was represented by female Caucasian-Brazilian above 50 years-old at symptom onset and with long disease duration (more than five years). The upper left side of the face was the main affected region at onset for both groups. The latency between peripheral facial palsy and subsequent development of HFS was of 28.9 months. Almost 40% of patients in both groups were diagnosed as having high blood pressure. Interestingly, if we consider only patients with Asian-Brazilian origin the prevalence of hypertension was identical (40%). Family history of HFS was seen, respectively, in 3.3% and 2.5% of patients with primary and postparalytic HFS.

Among patients with primary HFS one trigger factor was reported in 96 occasions (29%) and other 17 ones (5%) reported two or more triggering factors. In patients with postparalytic HFS triggering factors were reported in 7 cases (18%). The main triggering factors seen were anxiety or stress, reading, light and chewing for both groups.

Approximately 30% (101 CTs and 41 MRIs) and 23% (nine CTs and three MRIs) of patients, respectively, with primary and postparalytic HFS underwent neuroimaging procedures without statistical difference on the presence of vascular abnormalities on the posterior fossa (Table). In a subgroup analysis of 13 Asian-Brazilian patients who were scanned only one was positive (left vertebral artery loop).

Mean follow-up was, respectively, 41.9 and 21.2 months in both groups. Benzodiazepines (44%, primary HFS vs. 29%, postparalytic HFS), antiepileptic (43% vs. 67%) and antidepressant (6% vs. 7%) drugs corresponded to the majority of medications used. BTX-A was the treatment of choice, respectively, in 74% of patients with primary HFS and 59% of patients with postparalytic HFS (Table).

DISCUSSION

In this study data from patients with primary HFS were not significantly different from patients with postparalytic HFS suggesting that clinical presentation are similar between these groups, even if we take into account that pathophysiological mechanisms in both situations are different. Despite the possibility of assessment bias due to the retrospective nature of this study, this is to our knowledge the largest case series comparing primary and secondary cases of HFS.

Whether high blood pressure is associated to HFS and neurovascular compression of the facial nerve is still a matter of debate[5-8]. According to one retrospective study[9], arterial hypertension was significantly more common in Brazilian patients with HFS than in patients with blepharospasm. Thirty-eight percent of our patients with primary HFS had hypertension. A subgroup analysis of Asian-Brazilian patients showed similar prevalence. Our finding is similar with another study which compared the prevalence of hyper-

Table. Clinical features of patients with either primary or postparalytic hemifacial spasm.

Demographic and clinical features	Primary HFS (n=334)	Postparalytic HFS (n=39)	p value
Gender, female/male ratio	1.85 (217/117)	6.8 (34/5)	1.00; NS
Ethnicity			
Caucasian Brazilian	221 (66.1%)	26 (66.6%)	
African Brazilian	81 (25.2%)	10 (25.6%)	0.92; NS
Asian Brazilian	31 (9.2%)	4 (10.2%)	
Age symptom onset, y	55.6±13	51.9±15.5	0.16; NS
Disease duration, mo	63.5±76	49.3±53.9	0.15; NS
Affected side			
Right	142 (42.5%)	17 (43.5)	0.88; NS
Left	192 (57.4%)	22 (53.4)	
Distribution of facial spasm at onset			
Upper face	243 (82.9%)	21 (84%)	4.16; NS
Lower face	50 (17%)	4 (16%)	
High blood pressure	130 (38.9%)	15 (38.4%)	0.14; NS
Family history of HFS	11 (3.2%)	1 (2.5%)	0.47; NS
Neuroimaging (CT and MRI)	101 (30.2%)	9 (23%)	0.56; NS
Vascular abnormalities on posterior fossa	10 (7.0%)	1 (12.5%)	
Oral medicine			
Benzodiazepines	95 (44%)	4 (29%)	
Antiepileptic drug	93 (43%)	8 (67%)	
Antidepressant drug	13 (6%)	1 (7%)	
Other	16 (7%)	1 (7%)	
Botulinum toxin type A	246 (74%)	23 (59%)	

HFS, hemifacial spasm; n, number of patients; y, years; mo, months; CT, computer tomography; MRI, magnetic resonance imaging; NS, without statistical significance.

tension in Asian patients with HFS and controls and found negative correlation[10].

In our series vascular abnormalities on the posterior fossa were found in 10% of patients who had neuroimaging scans and only four cases were associated with hypertension, which reinforces that in our population perhaps HFS, hypertension and vascular abnormalities in the posterior fossa may not be interdependent. We believe we have underestimated the prevalence of vascular abnormalities in our sample since our protocol was not aimed at the identification of neurovascular compression at the root exit zone of the facial nerve and a large amount of patients only performed CT scans. The association between neurovascular contact of the facial nerve and HFS is supported by high-resolution MRI (constructive interference in steady state – CISS) and magnetic resonance angiography techniques[5,9] and also the efficacy of microvascular decompression procedures.

The postparalytic syndrome, which includes HFS, is a late consequence of axonal damage and aberrant regeneration. This process generally begins three months after injury[11]. According to our data, however, the latency for developing HFS after PFP was of 28.9 months, a longer period than expected.

Our results are somewhat different from an Italian multicenter study that evaluated 214 patients with HFS[4]. Although we also found a similar median age at onset and a more prevalent left-sided involvement of the facial nerve, their data suggest a similar prevalence of males and females but significant differences with regard to muscle spasm distribution at onset.

We do not know yet why HFS is more prevalent on the left side, but one may speculate that this is probably related to the anatomy of the vertebrobasilar system. Generally the anterior inferior cerebellar artery (AICA) or posterior inferior cerebellar artery (PICA), which arises, respectively, from the basilar and vertebral arteries, are the most implicated vessels in the neurovascular contact theory. The two vertebral arteries are usually unequal in size, the left one being generally larger than the right one[12,13]. It is not clear why this asymmetry exists. One of the hypotheses is that this asymmetry is related to the vascular requirements of the brain. Thus, it might be that this

left-sided preponderance of the vertebral artery is implicated on the left-sided prevalence of HFS.

Patients with Asian origin (Asian-Brazilian) corresponded to 9.4% of the total patients in our sample, with similar distribution among groups with primary and postparalytic HFS. It seems that narrowing of the posterior fossa may be a possible facilitating factor for HFS in Asian populations[14]. Although it was not our aim to systematically evaluate neurovascular contact compression rates we found only one Asian-Brazilian patient with vascular abnormality on posterior fossa.

Family history of HFS was to some extent higher among our patients. There are few case reports in the literature on familial HFS, but evidence suggests that there are no clinical differences between the sporadic and familial forms of HFS[15-20].

HFS is not considered as a life-threatening disease but functional and also psychological impact may seriously impair daily life of these affected subjects[21]. Therefore, triggering factors for facial spasm should be recognized and whenever possible avoided. In our series anxiety or stressful situations were the main precipitating factors in 32.1% of patients (n=120) with either primary HFS or postparalytic HFS.

Oral medicines were taken by a large amount of patients before attending to our Movement Disorder Unit (MDU). Benzodiazepines were the most frequent used medications in the primary HFS group, whereas patients with postparalytic HFS used to take antiepileptic drugs. BTX-A was the treatment of choice, respectively, in 74% and 59% of the patients in our MDU.

In summary, our results supports that the clinical profile of patients with primary and postparalytic HFS are similar, despite remarkable etiologic differences. Additionally, it seems that in our sample the association of high blood pressure with vascular abnormalities and HFS is not as strong as in other case series. Further prospective large case series are needed to address these questions.

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