

Hemifacial spasm can be the presenting symptom of a fourth ventricle tumour. A short case-illustrated review and pathogenetic considerations

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Abstract In this short case-illustrated review we aimed to analyse the possible nuances of hemifacial spasm (HFS) as the presenting symptom of a tumour of the fourth ventricle. The issue is remarkable since HFS can be secondary to a fourth ventricle tumour, even when no other neurological signs are reported. In addition, the possible presentation with only upper facial muscle involvement, as in the presented case, can be deceitful because this is characteristic of the benign and much more frequent “typical” form. Based on our intra-operative data and on the previously reported cases, we think that pathogenesis could be referable to the facial nerve nucleus involvement and that clinical nuances could be related to the specific somatotopy of the nucleus under the fourth ventricle floor that, as in our case, can be infiltrated by tumour. Resolution of the disorder can usually be obtained after the complete resection of the tumour that in the reported case resulted a subependymoma (WHO grade I), so far never described in literature associated with HFS.

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Introduction

Hemifacial spasm (HFS) is a neurological disorder characterized by unilateral involuntary paroxysmal contractions of the facial musculature. It can be primary or secondary. The most common aetiology of primary HFS is the mechanical compression of the cisternal portion of the seventh cranial nerve at the root entry zone by one or more arteries or veins [10]. Primary HFS can present with both the “typical” and the “atypical” forms. However, for anatomical reasons, the usual presentation is the selective involvement of the orbicularis oculi muscle (“typical HFS”). Conversely, the early contraction of the lower muscles of the face or, alternatively, of all the hemifacial muscles at the same time (“atypical HFS”) is much less frequent. Secondary HFS can be generally the effect of space-occupying lesions (tumours, aneurysms or arteriovenous malformations) located in the cerebello-pontine angle (CPA) close to the root entry zone. HFS in patients with masses in anatomical regions different from CPA has to be considered exceptional and its pathogenesis remains unknown. Secondary HFS commonly presents with the atypical form [3], but very rarely with the typical form. The authors report the rare case of a patient with a subependymoma of the fourth ventricle presenting with a typical HFS.

Illustrative case

A 45-year-old woman came to our attention with a 2-month history of left HFS. Spasm initially involved the periorbital area, then progressively spreading to all ipsilateral facial expression muscles and worsening in frequency and intensity. Her medical history was unremarkable, no previous infections or traumatism, no metabolic disorders,

Fig. 1 Preoperative MRI. Sagittal T1-weighted image (a) and axial T2-weighted image (b) before gadolinium administration. Sagittal (c) and axial (d) T1-weighted images after gadolinium administration. Tumour is located in the inferior part of the fourth ventricle and presents focal infiltration of the brain stem (floor of the fourth ventricle)

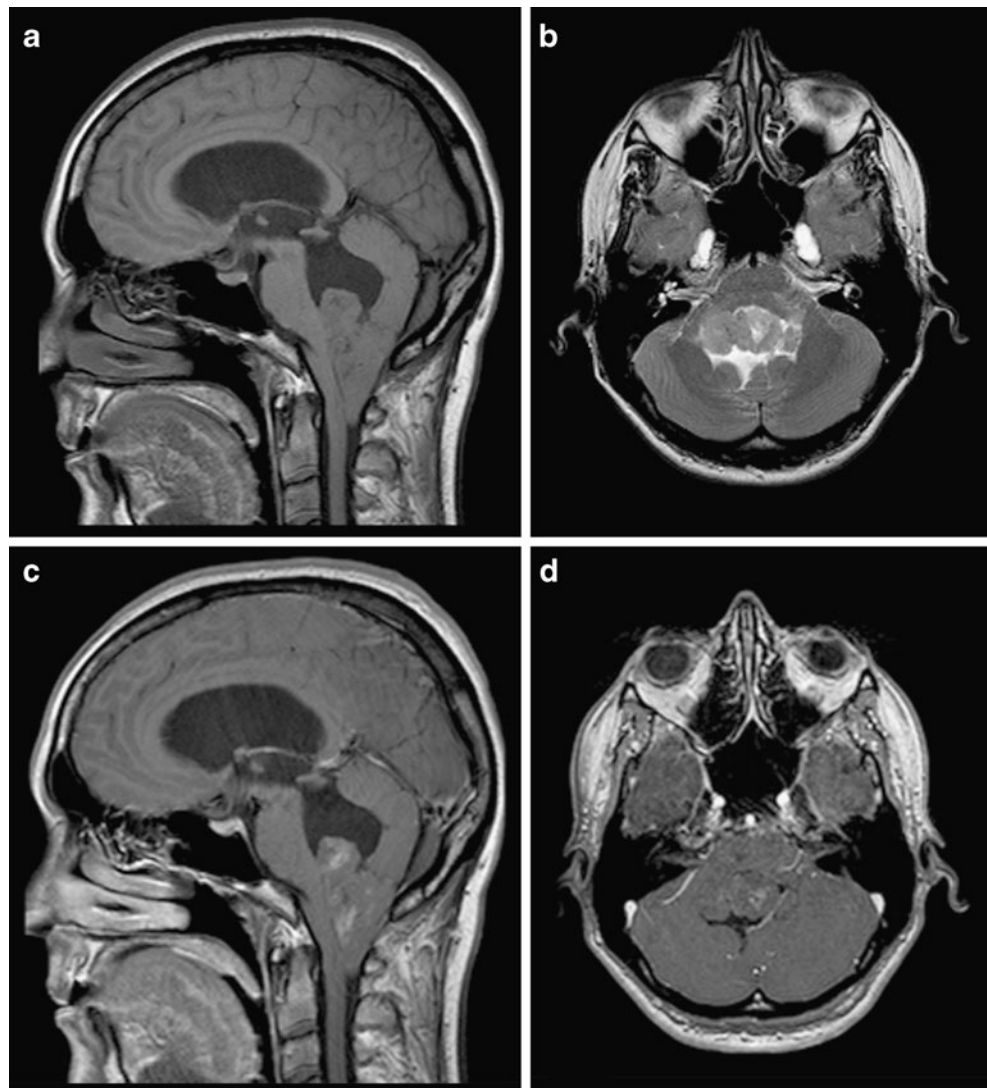
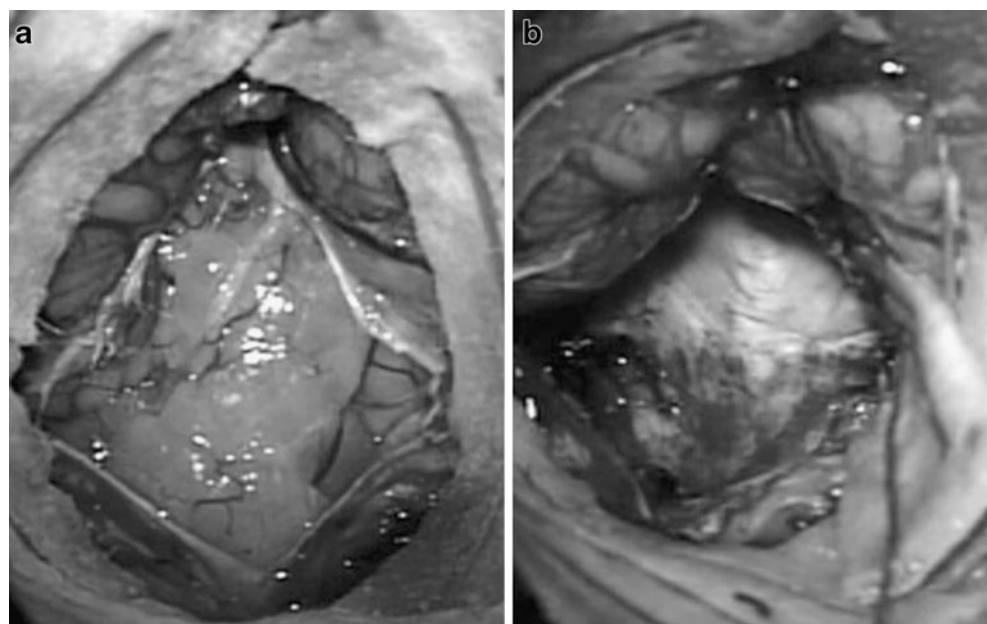


Fig. 2 Intra-operative view (sub-cerebellar approach). Initial microscopic view of tumour before surgical removal (a). Final microscopic view after complete surgical removal of tumour. Fourth ventricle floor presented multiple sites of tumour infiltration (b)



no hypertension. Neurological examination was unremarkable, except for the left HFS. Magnetic resonance imaging (MRI) (Fig. 1) revealed a large tumour of the fourth ventricle. It was irregularly shaped, and it inhomogeneously enhanced after gadolinium administration. It displaced the medulla and cerebellum. The fourth ventricle was completely filled by the tumour and supratentorial ventricles were enlarged. A cerebral angiography study excluded vascular abnormalities. Surgical resection was performed through a midline sub-occipital route. Intra-operatively, the tumour appeared strictly connected to the inferior part of the floor of the fourth ventricle, which appeared partially infiltrated (Fig. 2). No direct relationship of the tumour with vascular structures was detected. The histopathological report was of subependymoma (WHO grade I). Lobular architecture with microcystic degeneration and clustered nuclei were observed. Neither calcifications nor haemorrhage were detected. The MIB-1 labelling index was low (less than 1%) .

A post-operative MRI confirmed the total resection of tumour and the disappearance of hydrocephalus (Fig. 3). No adjuvant treatments were required because of the tumour's grade and the extent of removal. Immediately after the operation the left hemifacial spasm disappeared. One year after surgery the patient was symptom-free and MRI investigation did not show tumour recurrence (Fig. 3).

Literature review

The Medline database (PubMed, <http://www.ncbi.nlm.nih.gov/pubmed/>) was examined using the following terms: “hemifacial spasm”, “IV ventricle tumours”, “subependymoma”, “adulthood”. It was possible to identify nine cases of tumours in adulthood which presented at the onset with an HFS that could be directly or indirectly referable to fourth ventricle involvement. [1, 6, 10–13, 15, 16] (Table 1). In particular, they were two cases of schwannoma and one

Fig. 3 Post-operative MRI. T1-weighted images after gadolinium administration showed complete removal of tumour (a) and hydrocephalus resolution (b) in the post-operative stage. T1-weighted images after gadolinium administration at 1 year follow-up (c, d)



Table 1 Non-CPA tumours causing HFS at the onset reported in the literature

Reference	Age at onset, sex	HFS at onset	Duration of symptoms before HFS diagnosis	Site of lesion	Histology	Reported ethiopathogenesis	Surgical outcome on HFS
Nagata et al. 1992 [10]	61 years, female	Hemifacial	1 year	Tentorial	Meningioma	Unknown	+
Weiner et al. 1993 [15]	61 years, male	Hemifacial	Several years	IV ventricle	Schwannoma	Root entry zone involvement	-
Weiner et al. 1993 [15]	78 years, female	Hemifacial	7 months	IV ventricle	Schwannoma	Root entry zone involvement	-
Ogasawara et al. 1995 [11]	76 years, female	Hemifacial	3 years	Cerebellar convexity	Transitional meningioma	Stressed vascular conflict "pre-existing"	+
Rapanà et al. 1999 [13]	43 years, male	Orbicularis oculi	7 years	Obex	Ependymoma	Stressed vascular conflict "pre-existing"	+
Wrobel-Wisniewska et al. 2003 [16]	47 years, female	Not specified	1 year	Not specified	Meningioma	Not specified	+
Cancelli et al. 2005 [1]	42 years, female	Orbicularis oculi	3 years	Tentorial	Meningioma	Vascular conflict	+
Gómez-Perals et al. 2005 [6]	44 years, female	Hemifacial	15 months	Cerebellar convexity	Meningioma	Displacement/distorsion of medulla resulting in a secondary vascular conflict	+
Park et al. 2009 [12]	31 years, female	Orbicularis oculi	3 years	Cerebellar convexity	Meningioma	Vascular conflict and VII nc stretching	+

case of obex ependymoma located into the fourth ventricle. Conversely, six cases of meningioma (two tentorial, three of cerebellar convexity, one not specified) were close to but not into the fourth ventricle.

Subependymoma of the fourth ventricle was never described as a cause of HFS.

The two patients affected by fourth ventricle schwannoma presented an atypical form of HFS [15]. Conversely, patients affected by fourth ventricle ependymoma presented selective involvement of the orbicularis oculi muscle [13]. In all cases, HFS disappeared only when total tumour removal was achieved. All three patients did not present additional neurological signs.

Two out of six cases of meningioma occurred with orbicularis oculi spasm at the onset [1, 12]. However, only an indirect anatomical relationship with the fourth ventricle was detected.

Finally, in eight out of nine cases the etiology of HFS was attributed to a pre-existing neurovascular conflict that became symptomatic when the tumour arose. This was the case of the three patients affected by upper HFS. In one case, displacement and distorsion of the medulla resulting in a secondary vascular conflict was suggested. Actually, all cases were explained by the authors as "peripheral" disorders.

Discussion

Secondary HFS is not frequent, ranging from 0.3% to 5.1% [5, 7] of all cases reported in the literature on this

neurological disorder. CPA tumours have been described from 0.3 to 0.6 of cited cases (including epidermoid tumors, schwannomas, meningiomas, and cysts) [8, 10]. Also, demyelinating disorders, traumatism and infections could cause HFS [4]. Furthermore, four cases of HFS as a feature of Chiari malformation are illustrated in the literature [9]. Recently some authors have suggested that a smaller posterior fossa CSF space may be a risk factor for HFS [2]. Based on these data, it is clear that HFS diagnosis is challenging.

Even if primary and secondary HFSs share common demographic and clinical features, sex distribution, age at onset, synkinesis and rarity of familial cases, the two forms usually differ in facial muscle involvement at onset and in neurological signs' association. Habitually, patients with secondary HFS show contractions at onset of the lower muscles of face or of the hemifacial muscles at the same time. The involvement of the orbicularis oculi muscle at onset is typical of primary HFS but extremely rare among secondary HFS (none out of 241 patients with secondary HFS reported by Colosimo et al. [3]). The difference in facial muscle recruitment in the two forms reflects the particular topographic arrangement of cranial nerve VII fibres. In fact in the cerebello-pontine cistern the nerve fibres located in the anterior/caudal side of facial nerve innervate the upper facial muscle, while the fibres located in the posterior/rostral side innervate the lower muscles of the face. For this reason, normally, typical HFS starts with orbicularis oculi spasm, because blood vessels compression occurs at the anterior/caudal side of facial nerve. On the

other hand, atypical HFS is caused by a compression of the posterior/rostral fibres as it could happen in CPA lesions [14].

In our case, the tumour was located in the fourth ventricle and it did not apply any direct pressure on the cisternal tract of the facial nerve. Furthermore, at the end of surgical procedure, we did not find any neurovascular conflict that could be exacerbated by the presence of the tumour; again, no anatomical features predisposing HFS, such as a small posterior fossa, were detected. Instead, since the tumour was infiltrating the floor of the fourth ventricle, it appeared more likely that the HFS was due to the involvement of the facial nucleus. In fact, nucleus neurons that are designated to innervate the upper part of the face are located in the lower part of the nucleus. In our patient the lower part of the nucleus was just closely to the tumour. In our opinion this is the anatomical explanation of the presented case and it could be, maybe partially, the anatomical explanation of others reported in the literature of secondary HFS presenting with the typical form [12, 13].

The involvement of the orbicularis oculi muscle alone, characteristic of the much more frequent primary HFS, could be considered the earliest symptom of fourth ventricle tumour when it is infiltrating the inferior part of the ventricle floor. Subsequent turning into the atypical form could be the sign of a growing facial nucleus involvement. Central origin of HFS in relation to fourth ventricle tumours is innovative, since until now all similar cases were explained as “peripheral” disorders. The hypothesis could be further validated by the clinical similarity between our case and the case described by Rapanà et al. [13]; in fact, in both cases the tumours (ependymoma and subependymoma) presented a similar infiltrative attitude towards the fourth ventricle floor (Fig. 3).

The absence of neurological signs could be deceitful because brain tumours responsible for HFS are usually located in the ponto-cerebellar angle and in that case usually associated with multiple cranial nerve involvement.

We believe that the considerations presented in this short case illustrated review should be taken in consideration when a patient with the typical form turns into the atypical form of HFS, even if no other neurological signs are detected. In this case, fourth ventricle investigation should be mandatory. In our experience, the surgical removal of the tumour enabled the prompt disappearance of HFS. Concerning the nature of the tumour, this appears to be the first case presented in the literature of HFS related to a subependymoma of the fourth ventricle.

Conflicts of interest None.

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