

# Secondary or Symptomatic Hemifacial Spasm Caused by a Tumor of Cerebellopontine Angle: Case Report and Review of Literature

Mohammad-Javad Mirzayan <sup>1\*</sup>, Morteza Faghih-Jouibari <sup>2</sup>, Ahmad Aoude <sup>2</sup>, Seyed-Mohammad Ghodsi <sup>2</sup>

<sup>1</sup> Department of Neurosurgery, Medical School Hannover, Germany

<sup>2</sup> Department of Neurosurgery, Shariati Hospital, Teheran University of Medical Sciences, Tehran, Iran

\* Corresponding Author: Department of Neurosurgery, Medical School Hannover, Germany. Tel: +49-1739148646, Fax: +49-511-6407079, E-mail: mirzayan@hotmail.com

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## Abstract

**Background & Importance:** Primary hemifacial spasm is usually caused by microvascular compression of the facial nerve at its root exit zone at brainstem without any space-occupying pathology. Secondary or so-called symptomatic hemifacial spasm has an additional underlying pathology e.g. tumors leading to the microvascular compression. We review and discuss the pathophysiology and the literature of secondary hemifacial spasm.

**Case Presentation:** We report on a 23 year-old man with a secondary right-sided hemifacial spasm in association with an ipsilateral epidermoid tumor of the cerebello-pontine angle.

**Conclusion:** Intraoperatively, no offending vessel was observed near the root exit zone of the facial nerve. Microsurgical tumor removal relieved the patient also from his hemifacial spasm.

**Keywords:** Cerebello-pontine angle; Epidermoid; Facial nerve; Microvascular decompression; Skull base

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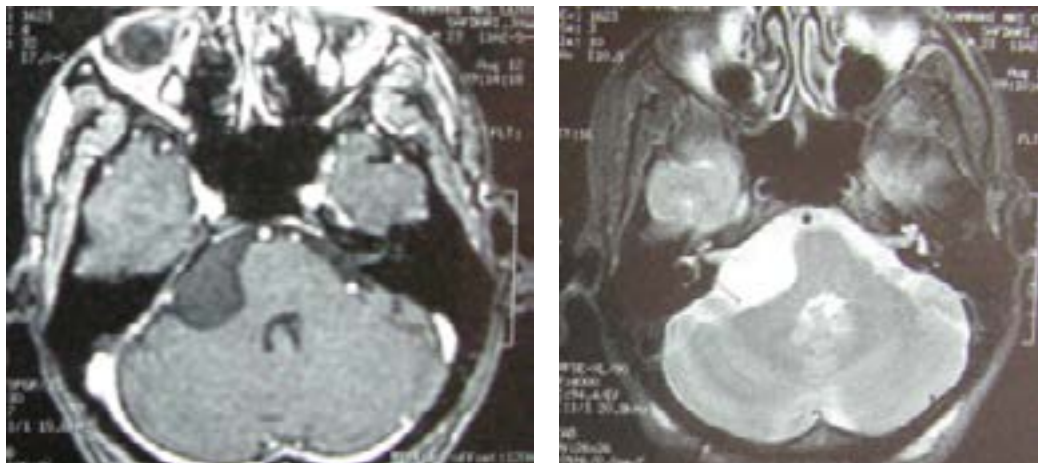
## Introduction

Epidermoid tumors arise from remnant ectopic epithelial cells and account for 0.3-1.8 % of all intracranial tumors (1). Arising in the cerebellopontine angle (CPA), they usually present with deficits of the lower cranial nerves, cerebellar and brainstem symptoms (2,3). Hemifacial spasm (HFS) has been primarily well understood as a result of microvascular compression of the facial nerve at its root exit zone at the brainstem (4). Secondary or so called symptomatic HFS has been reported in few case reports to be associated with additional pathologies like tumors of the CPA (5). While the incidence of epidermoid tumors are far away from those of schwannomas and meningiomas (as the most common CPA tumors), secondary HFS has been reported rather for epidermoid tumors than schwannomas and meningiomas (6). We report on an additional patient with an epidermoid tumor of the CPA who presented with a secondary HFS without obvious microvascular compression, and summarize the so far existing explanations and add our understanding of the pathophysiology.

## Case Presentation

This 23-year-old man suffered from right-sided HFS involving the platysma. Starting 18 months prior surgery the frequency increased within the last 6 months. Neurological examination did not reveal any deficit of other cranial nerves, in particular no hearing loss, tinnitus or trigeminal hypesthesia. Computed tomography and magnetic resonance imaging (MRI) of the brain showed a large mass of the CPA which was hypointense in T1, hyperintense in T2, without contrast enhancement (Figure 1). Diffusion-weighted imaging was not available. The patient underwent surgery via a retrosigmoid approach in semisitting position. Microsurgical appearance of the tumor and histopathological examination confirmed the diagnosis of an epidermoid tumor. The tumor was removed totally except the capsule leaving at the surface of brainstem. Neither during tumor removal nor at the end of tumor removal a microvascular compression of the facial nerve could be observed. Microsurgical exploration of the nerve root exit zone was done before closure of the dura.

Surgery and postoperative course were unremarkable without any neurological morbidity. The HFS disappeared completely during the early postoperative period and remained unchanged at 6 months follow-up.



**Figure 1.** Contrast enhanced, T1-weighted, axial MRI (left) demonstrates the mass within the right cerebello-pontine-angle. T2-weighted axial MRI (right) demonstrates the hyperintense signal of the mass.

## Discussion

Secondary or so-called symptomatic HFS has been occasionally reported as the presenting symptom for several pathologies inside or surrounding the CPA in few case reports. These varies from arachnoid cyst, aneurysm, meningioma, lipoma, to even bony narrowing of the posterior fossa as an anatomical variant (5,7-9). Among the tumors of the CPA, epidermoid tumor seems to have a disproportionate affinity causing a secondary HFS (6). This is remarkable as meningioma and schwannoma are the more common tumors of CPA. Thus, beside the plain gross occurrence of a mass other “microanatomical” features may be crucial in development of secondary neuro-vascular compression syndromes. Occurrence of an epidermoid tumor within the subarachnoid space may result in direct open exposure of the facial nerve by the tumor. This absent arachnoid interlayer results in reduced protection of the facial nerve. Both capsule and content of the epidermoid tumor might be irritant to the facial nerve (10). Many authors report that epidermoid tumors involve the facial nerve early (11). The hypothesis of possible direct irritation on the facial nerve is in accordance with the missing evidence for a correlation of tumor size with occurrence of the secondary HFS. As schwannomas and meningiomas arise between the dura and arachnoidea, the facial nerve is separated by an additional protecting interlayer. This interlayer makes them obviously less sufficient to cause symptomatic HFS more often. Schwannomas do involve very late the facial nerve. Consequently, huge vestibularis schwannomas are regularly seen without facial nerve deficits. Samii M et al.(1995) has reported an incidence of 0.6% for secondary HFS caused by schwannomas within the cerebello-pontine angle (12). The occurrence of HFS in some cases should be most likely related to the narrowing of the cisterns and displacement of the vessels against the facial nerve. Comparing the bony size of posterior fossa in patients with or without idiopathic HFS Kamiguchi et al. found the infratentorial bony border of patients with HFS to be smaller than patients without HFS (9). Recently, microvascular compression has been reported in a case of vestibular schwannoma to be responsible for dizziness and not

only the tumor itself (13). As HFS is an impressive obvious clinical presentation, some of other symptoms can remain unregistered by the patients. This issue has been observed by Auger RG who found that in patients presented with secondary HFS additional deficits of cranial nerves were evident (14). Detailed clinical examination of the patients is of major and conclusive importance. In case of simultaneous deficits of other cranial nerves, cerebellar or brainstem symptoms, or headache a mass lesion would be more probable than isolated microvascular compression. Independently from additional deficits MRI is mandatory prior to any kind of therapy like botulin toxin injection or surgery. If additional neurological symptoms are present MRI should be available more promptly. This is obviously not self-evident as reported recently (6). They have reported that only 14 of their 55 patients with a secondary HFS had a preoperative imaging (6). Although they discuss that only 0.8% of their HFS cases (from 1984 to 2008) were caused by a tumor, in the meantime, a treatment like botulinum toxin or surgery prior to cranial imaging has to be avoided in every single case. Atypical evolution of HFS has been reported by some authors to be suggestive for secondary HFS. However, this might be not so clear to give a strong suspicion in every case. It would be crucial to have information whether a recurrent epidermoid tumor appears again with a recurrence of the HFS. So far, the literature does not provide any information about such a possibility. Regarding microvascular decompression following tumor removal differing experience exist as microvascular compression has not been detected in all cases so in our patient. By opening the dura and releasing CSF the configuration can change and be undetectable even during first intradural steps of microsurgery. So even resolution of idiopathic hemifacial spasm after posterior fossa exploration without vascular decompression has been reported (15).

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## Conflicts of Interest

The authors claim no conflict of interest.

## References

1. Ghaemi K, Mirzayan MJ, Krauss JK (2010). Interdural Epidermoid of the Cavernous Sinus. *Cen Eur Neurosurg.* 2010; 71(3): 155-6.
2. Dufour H, Fuentes S, Metellus P, et al. [Intracavernous epidermoid cyst. Case report and review of the literature]. *Neurochirurgie.* 2001; 47: 55-59.
3. Tatagiba M, Iaconetta G, Samii M. Epidermoid cyst of the cavernous sinus: clinical features, pathogenesis and treatment. *Br J Neurosurg.* 2000; 14: 571-575.
4. Jannetta, P.J. (1977). Observations on the etiology of trigeminal neuralgia, hemifacial spasm, acoustic nerve dysfunction and glossopharyngeal neuralgia. Definitive microsurgical treatment and results in 117 patients. *Neurochirurgia (Stuttg).* 1977; 20: 145-154.
5. Lee SH, Rhee BA, Choi SK, et al. Cerebellopontine angle tumors causing hemifacial spasm: types, incidence, and mechanism in nine reported cases and literature review. *Acta Neurochir (Wien).* 2010; 152(11): 1901-1908.
6. Han H, Chen G, Zuo H. Microsurgical treatment for 55 patients with hemifacial spasm due to cerebellopontine angle tumors. *Neurosurg Rev.* 2010; 33: 335-339.
7. Nagata S, Matsushima T, Fujii K, et al. Hemifacial spasm due to tumor, aneurysm, or arteriovenous malformation. *Surg Neurol.* 1992; 38: 204-209.
8. Han IB, Chang JH, Chang JW, et al. Unusual causes and presentations of hemifacial spasm. *Neurosurgery.* 2009; 65: 130-137.
9. Kamiguchi H, Ohira T, Ochiai M, et al. Computed tomographic analysis of hemifacial spasm: narrowing of the posterior fossa as a possible facilitating factor for neurovascular compression. *J Neurol Neurosurg Psychiatry.* 1997; 62: 532-534.
10. Wakabayashi T, Tamaki N, Satoh H, et al. Epidermoid tumor presenting as painful tic convulsif. *Surg Neurol.* 1983; 19: 244-246.
11. de Souza C, de Souza R, da Costa S, et al. Cerebellopontine angle epidermoid cysts: a report on 30 cases. *J Neurol Neurosurg Psychiatry.* 1989; 52(8): 986-990.
12. Samii M, Matthies C. Acoustic neurinomas associated with vascular compression syndromes. *Acta Neurochir (Wien).* 1995; 134: 148-154.
13. Wuertenberger C, Rosahl S. Vertigo and tinnitus caused by vascular compression of the vestibulocochlear nerve, not intracanalicular vestibular schwannoma: review and case presentation. *Skull Base* 2009; 19(6): 417-424.
14. Auger RG, Piepgras DG. Hemifacial spasm associated with epidermoid tumors of the cerebellopontine angle. *Neurology.* 1989; 39: 577-580.
15. Aoki N, Nagao T. Resolution of hemifacial spasm after posterior fossa exploration without vascular decompression. *Neurosurgery.* 1986; 18: 478-479.

## Comments

I read with interest the report of a case of hemifacial spasm due to Epidermoid tumor of CPA by Mirzayan MJ et al. The kind authors have reviewed some of the reports relevant to this pathology briefly and added their case. The main point highlighted in this report is that there was no vascular compression upon the REZ of the 7th. nerve. The association of CPA tumors, especially the epidermoids with hemifacial spasm is reported by different authors as the kind researchers have included in their reference list. The issue worthy of consideration might be that P Jannetta insisted upon the association of tumors and hemifacial spasm to be the result of a kind of vascular compression in most of the cases. This has been reminded and published by several authors further on (1,2). It has always been demanding to look delicately around all the course of the 7th nerve (or the 5th, nerve in patients with trigeminal neuralgia and epidermoid tumors) for a kind of vascular compression which might be present and needs decompression, otherwise there will be a failure for long term symptom free period and it will recur even without tumor recurrence. If this search is done after appropriate tumor decompression, finding adherence between the offending vessel and the nerve can be somehow misinterpreted.

*Kazem Abbasioun, MD, Professor of Neurosurgery, Vice-president of Iranian Association of Surgeons, Tehran, Iran*

### Reference:

- 1-Alemdar M. Epidermoid cyst causing hemifacial spasm. *J. Of Rural Neurosciences in Rural Practice.* 2012;3;344-6.
- 2-Microvascular decompression in 55 patients with hemifacial spasm due to CPA tumors. Keu K et al. *Neurosurgical Review* 2012;33;335-9.

It is a great pleasure to see the first issue of Iranian journal of neurosurgery with international authors. Dr mirzayan et al are reporting a case of Cerebellopontine Angle (CPA) epidermoid tumor presenting with hemifacial spasm (HFS) (1). They are correctly discussing the various intraoperative findings in such cases in the literature. Micro vascular compression although commonly present in such cases nevertheless may be absent in some cases (as in the current case). The authors have not removed the tumor capsule, they have drained cafe and distorted morbid anatomy, this may have affected the anatomical vicinities and the micro environmental change

could have alleviated the HFS. Further cases with more detailed anatomical report may be necessary to discuss the pathophysiology of HFS.

*Hooshang Saberi, MD, MPH, Associate Professor of Neurosurgery, Tehran University of Medical Sciences, Tehran, Iran*

### Reference:

- 1-Mirzayan M.J, Faghih-Jouibari M, Aoude Ahmad, Ghodsi S.M. Secondary or Symptomatic Hemifacial Spasm Caused by a Tumor of Cerebellopontine Angle: Case Report and Review of the Literature. *IrJNS.* 2015;1(1):53-55.