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ORIGINAL ARTICLE

Microsurgical treatment for 55 patients with hemifacial spasm due to cerebellopontine angle tumors

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Abstract Tumor-related hemifacial spasm (HFS) has been found to be rare. During the period from October 1984 to October 2008, we treated 6,910 HFS patients using a microsurgical procedure. Of these HFS patients, 55 cases were associated with cerebellopontine angle tumors. A small craniectomy was performed in order to excise the tumor. All tumors were found to compress the root exit zone (REZ) of the facial nerve to different extents, but concomitant vascular compression of the facial nerve was observed in a majority of cases, and microvascular decompression of the facial nerve at REZ was conducted in 43 of 55 patients (78.2%) by displacing the co-compressing vasculature away from the REZ and retaining it using a Teflon pad. Intraoperative findings and postoperative pathological examinations suggested that the tumors were epidermoid cysts, meningiomas, and Schwannomas. Follow-up in 48 of 55 patients for 4–230 months after surgery showed that the clinical symptoms of HFS disappeared in 43 cases, improved in two cases, and recurred in three cases. Ten patients had sequelae associated with the operation. We concluded from this study that the majority of cases of tumor-related HFS are caused by combined tumor and vascular co-compression at the REZ, and tumor removal and microvascular decompression are required in order to relieve the symptoms.

Keywords Cerebellopontine angle tumor · Hemifacial spasm · Vascular compression · Microvascular decompression

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Introduction

Hemifacial spasm (HFS) is a common cranial nerve disorder characterized by unilateral involuntary paroxysmal contractions of the facial musculature. Generally, HFS initially involves the orbicularis oculi and gradually spreads to other parts of the face and eventually to the platysma. The majority of HFS cases (over two thirds) are caused by compression at the root exit zone (REZ) of the facial nerve mediated by the adjoining vasculature [10, 11]. Facial nerve decompression was proposed for HFS by Gardner and Sava in 1962 [6, 14]. Non-vascular etiologies for HFS have been reported sporadically [1, 5, 7, 9, 12, 18], and tumor-related HFS has been found to be rare [1–4, 8, 19]. A total of 6,910 patients with HFS were surgically treated in our hospital over more than 20 years from October 1984 to October 2008. Careful examination of these cases suggested that 55 patients (0.8%) had associated cerebellopontine angle (CPA) tumors. We report the clinical characteristics of these CPA tumor-related HFSs and the outcome of microsurgical treatment. Permission to use these data was obtained from the Research Ethics Board of Yuquan Hospital, Tsinghua University. All patients gave their informed consent prior to their inclusion in the study.

Materials and methods

The series of patients with tumor-related HFS included 22 men and 33 women. Ages ranged from 19 to 65 years (mean, 37.9 years). There were 25 patients with hemifacial spasm on the left side and 30 on the right side; symptom duration ranged from 2 to 17 years (mean, 6.1 years). All patients had the typical clinical symptoms of HFS, including paroxysmal and repetitive involuntary contrac-

tions of the muscles controlling facial expression. Eleven patients also had ipsilateral tinnitus or hearing impairment. No other neurological deficits were observed (Table 1). Because neuroimaging using computerized tomography (CT) or magnetic resonance imaging (MRI) was not routinely conducted before microsurgery, only 14 patients with intracranial CPA tumors were diagnosed by preoperative neuroimaging.

Surgical techniques

All patients were operated under general anesthesia. Following induction of anesthesia and intubation, the patient was placed in the lateral decubitus position with appropriate padding of pressure points. The head was rotated approximately 10° away from the affected side, and the vertex was lowered by 15° to expose the proximal aspect of the cranial nerve. A small craniectomy (2–2.5 cm in diameter) was performed with the anterior margin at the posterior border of the mastoid sinus. If the tumor was difficult to resect due to the small size of the cranial hole, the hole was enlarged appropriately using gouge forceps. Only three patients underwent hole expansion. Following tumor removal, an artery compressing the REZ of the facial nerve was found in 43 patients (78.2%; Fig. 1). In order to disengage the vasculature compressing the facial nerve

Table 1 Clinical summary of patients with tumor-associated HFS

Characteristics	Number of cases (%)
Sex	
Male	22 (40.0)
Female	33 (60.0)
Side of facial nerve involvement	
Left	25 (45.5)
Right	30 (54.5)
Clinical manifestations	
Hemifacial spasm	55 (100)
Hearing decrease or loss	11 (20.0)
Tumor type	
Epidermoid cyst	41 (74.5)
Meningioma	7 (12.7)
Schwannoma of cranial nerve VIII	2 (3.6)
Schwannoma of cranial nerve IX	3 (5.6)
Schwannoma of cranial nerve X	2 (3.6)
REZ compression factors	
Combined tumor and vascular compression	43 (78.2)
Epidermoid cyst	29 (67.4)
Other tumors	14 (32.6)
Tumor compression alone	12 (21.8)
Epidermoid cyst	12 (100)

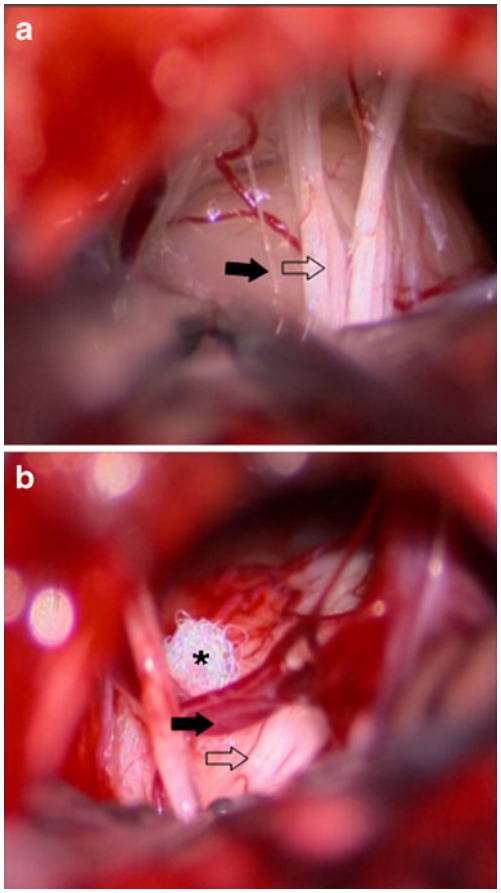

Fig. 1 Digital micrographs of a representative patient with left hemifacial spasm showing a Schwannoma (*solid arrow*) from cranial nerve X (*open arrow*) before tumor removal (**a**). An artery (*solid arrow*) was found to be compressing the REZ of the facial nerve (*open arrow*) after tumor removal (**b**)

REZ, we detached and laterally moved the vasculature away from the REZ by about 3 mm. A small piece of polytef (Teflon) pad was then placed between the vasculature and the brainstem; care was taken to ensure that the pad did not contact the REZ. We term this operation "tumor resection plus microvascular decompression" (MVD; Fig. 2).

Results

Anatomical–pathological findings

During the operation, we found that the lesions were "pearl-like tumors" at the CPA in 41 patients, meningioma arising from dura mater of the posterior petrosum or petral-clivus area in seven, or Schwannomas of the cranial nerves VIII (in two cases), IX (three), or X (three; Table 1). These lesions were postoperatively verified by standard histopathological examination. Tumor sizes measured intraoperatively with a soft ruler were between 1.5 and 5 cm in

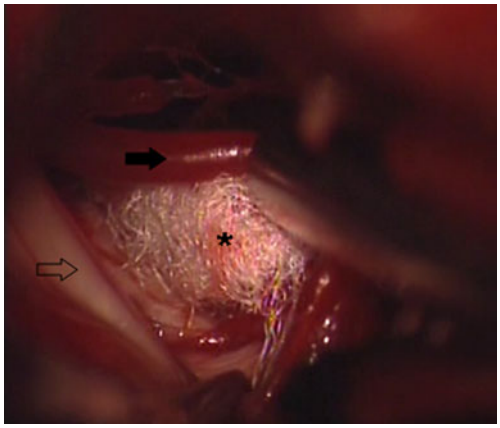

Fig. 2 Digital micrograph after tumor (at right CPA) resection. The co-compressing vasculature was detached and moved away from the REZ, and a small piece of Teflon pad was placed between the vasculature and the brainstem, taking care to prevent contact between the REZ and the pad

diameter and compressed the REZ of the facial nerve to different degrees. As summarized in Table 2, tumors were removed totally in 51 patients (92.7%) and subtotally in four (7.3%, meningioma in two cases and Schwannomas of the cranial nerves VIII in two). Of the 43 patients with tumors accompanied by vascular co-compression at the facial nerve REZ, the anterior inferior cerebellar artery was the artery responsible in 29 patients and the posterior inferior cerebellar artery in 14. No arterial compression of the facial nerve REZ was found in 12 of the 41 patients with an epidermoid cyst (Table 1).

Outcome and complications

Clinical outcomes were assessed at 1-year intervals after treatment. Cure was defined as the disappearance of facial spasms. Improvement was defined as a significant decrease in the frequency or degree of facial spasms. Inefficacy was defined as no change in facial spasms. Recurrence implied that facial spasms disappeared shortly after surgery but relapsed after 1 year. Shortly after surgery, 11 patients had peripheral facial paralysis and eight had hearing impairment. Other surgical complications included dysarthria in two patients and light dysphagia in one. No patient died after surgery. MRI examination 3 months after surgery revealed the presence of residual tumor tissue in four patients (two with meningioma and two with Schwannoma of cranial verve VIII) who then received gamma knife therapy.

Follow-up in 38 of the 43 patients treated by "tumor resection plus MVD" (group A) and 10 of the 12 patients who had tumor resection only (group B) was performed through correspondence including telephone interviews and return visits. The period of follow-up ranged from 4 to 230 months (mean 73 months). In group A, 37 patients were cured and one recurred; in group B, six patients were cured, two improved, and two recurred. There were ten patients with sequelae associated with the operation: hearing impairment in six patients (these all had preoperative tinnitus or impaired hearing on the ipsilateral side), light peripheral facial palsy in two, and hoarseness in two. Contact was lost with seven patients; of these, five had epidermoid cysts, one meningioma, and one had cranial verve IX Schwannoma. The remaining patients all recovered fully.

Discussion

The reported frequency of tumor-related HFS is 0.3% to 2.5% of all HFS [15, 17, 21], depending upon the type of

Table 2 Outcomes after microsurgery of patients with tumor-associated HFS

		No. of cases (%)
Tumor removal		
Total		51 (92.7)
Subtotal		4 (7.3)
Meningioma		2
Schwannoma of cranial nerve VIII		2
Clinical manifestation (48 patients followed-up)		
	Group A	Group B
Cure	37 (97.4)	6 (60.0)
Improved	-	2 (20.0)
Recurred	1(2.6)	2 (20.0)
Sequelae		
Hearing impairment		6 (12.6)
Light peripheral facial palsy		2 (4.2)
Hoarseness		2 (4.2)

tumor, race, and geographical location. In the Chinese population, only 35 of 4,260 HFS cases (0.82%) were reported to be associated with tumors [24]. Here, we report a similar rate of morbidity (0.8%) for HFS resulting from cerebellopontine angle tumors. Symptoms of these tumors (in addition to HFS in all patients) were ipsilateral tinnitus or hearing impairment in 11 of the 55 patients. No other neurological deficits were observed.

Diverse lesions have been reported to cause HFS via direct or indirect mass effects; these include meningioma, acoustic Schwannoma, dermiod, epidermoid, lipoma, glioma, arachnoid cyst, aneurysm, and arteriovenous malformation [3, 6, 13, 15, 20]. The lesions may be located at the ipsilateral or contralateral CPA or at the intrinsic pontine. Large meningiomas at the distant ipsilateral posterior fossa have also been reported as a cause of HFS [8]. In the present study, the type and morbidity of the tumors is summarized as follows: epidermoids in 41 patients (74.5%), meningiomas in seven (12.7%), acoustic Schwannomas in two (3.6%), glossopharyngeal nerve Schwannomas in three (5.6%), and vagus nerve Schwannomas in two (3.6%). Schwannomas from the ninth and tenth nerves that present as ipsilateral HFS have not been reported previously.

HFS associated with tumors may be caused by direct or indirect mass effects and/or chemical irritation of the facial nerve. All tumors in the present study were located at the extra-brain ipsolateral CPA. After tumor removal, the REZ of the facial nerve was explored in all patients; this revealed 43 patients (78.2%) with vascular compression of the REZ. All 14 patients with meningiomas or Schwannomas at CPA had two compressive factors, and in the 41 patients harboring epidermoid cysts, 29 (70.7%) were accompanied by vascular compression. This demonstrated that two-factor compression of the ipsolateral facial nerve REZ may be an important etiological factor in tumor-associated HFS. Nevertheless, 12 patients (29.3%) in this series harbored epidermoid cysts, and there was no evidence of compression due to the vasculature. In these patients, HFS might be caused by chemical irritation to the REZ, possibly by sterols secreted from the tumor that could cause demyelination in the REZ, or by some indefinite etiological factors such as the inflammatory adhesion of the arachnoid membrane around the REZ as an intraoperation finding in some rare cases. We suggest that CPA tumors are unlikely to cause HFS without simultaneous vascular compression of the facial nerve REZ, with the exception of chemical irritation associated with epidermoid cysts or inflammatory adhesion around the REZ in rare cases.

In cases where compression is mediated both by the tumor and by the adjoining vasculature, it is difficult to assess which plays the more important role in causing HFS. Intraoperative monitoring of the abnormal muscle response (AMR) has been employed to identify the cause of HFS and for prognostic evaluation [16, 22, 23]. This series of patients was not monitored intraoperatively for AMR, and it was not possible to identify whether the tumor or the vasculature was the primary cause of HFS: It may only be concluded that "tumor resection plus microvascular decompression" was responsible for the disappearance of symptoms.

Preoperative neuroimaging may help distinguish between compression and vascular lesions around the facial nerve. However, for our patients with HFS (6,910 cases), preoperative neuroimaging was not prescribed for most patients with the exception of those whose accompanying symptoms included tinnitus, impaired hearing, or the involvement of other cranial nerves. This was not only due to financial constraints but also because only 0.8% of HFS cases are caused by tumors, and most patients whose HFS is caused by a tumor can be treated via the same cranial hole as those without tumors. In the present study, only 14 of the 55 patients with tumor-related HFS were preoperatively evaluated using CT or MRI. There were no significant differences in tumor removal or complication rates between the neuroimaging group and the remaining cases ($p>0.05$). Nevertheless, an option for preoperative neuroimaging may be provided in the future.

In many cases, there was evidence that compression of the facial nerve REZ was caused both by the tumor and by the surrounding vasculature. During microsurgery for tumor removal, it is therefore important to evaluate whether there is additional vascular compression. In the present patient series, 43 (78.2%) were found to have vascular co-compression of the REZ following tumor resection. The vessel responsible was detached from the REZ, and a small piece of polytef pad was introduced between the vasculature and the brainstem; care was taken to ensure that the pad did not contact the REZ. Of the 43 patients in group A treated with "tumor resection plus MVD," 38 were followed up. Of these, 37 (97.4%) were cured and one (2.6%) recurred. Ten of the 12 patients in group B who had tumor resection only were followed up; of these, six (60.0%) were cured, two (20.0%) improved, and two (20.0%) recurred. The difference in cure rate between the two groups was significant ($p<0.005$). The procedure of "tumor resection plus MVD" was therefore successful in that with very few exceptions, all HFS patients treated improved markedly; the majority were cured of their condition. Because the REZ of the facial nerve was carefully explored after tumor removal in all patients, it seems unlikely that the vasculature responsible would be missed. Compared with group A subjects, HFS patients with tumor compression without evident vascular compression are less likely to be cured. This may be due to a difference in the pathophysiology of the disease process between the two groups.

Conclusions

The majority of cases of tumor-related HFS are caused by combined tumor and vascular co-compression at the REZ. In our experience, both tumor removal and microvascular decompression are required in order to relieve the symptoms.

Acknowledgments This work was supported by the Tsinghua-Yue-Yuen Medical Sciences Foundation (no. 20240000513) and the Beijing Natural Science Foundation (no. 7062036).

Permission A total of 6,910 patients with HFS were surgically treated in our hospital over more than 20 years from October 1984 to October 2008. Careful examination of these cases suggested that 55 patients (0.8%) had associated cerebellopontine angle (CPA) tumors. We report the clinical characteristics of these CPA tumor-related HFSs and the outcome of microsurgical treatment. Permission to use these data was obtained from the Research Ethics Board of Yuquan Hospital, Tsinghua University. This study was performed in accordance with the ethical standards laid down in 1964.

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Comments

Marc Sindou, Lyon, France

The authors present a considerably numerous series of hemifacial spasm (HFS) cases related to the presence of a cerebellopontine angle tumor: 55 patients treated over the last 24 years! This represents a 8‰ percentage of their overall series of 6,910 HFS who underwent microsurgical intervention. This is likely the biggest such experience in the world.

The anatomical–pathological findings described in the article are of the highest interest, especially the fact that the tumor was responsible of the spasm through a compressing vascular mechanism at the root exit zone (REZ) of the facial nerve. Therefore, the authors insist to complete the removal of the tumor by "displacing the co-compressing vasculature away from the REZ."

Louis J. Kim, Seattle, USA

This article describes an extensive experience of hemifacial spasm in the setting of concomitant cerebellopontine angle tumors. Given the relative rarity of this clinical setting, the series of Han et al. is impressively large, and the favorable clinical outcomes are a reflection of this superb neurosurgical group. Of interest, tumor patients were categorized into groups A and B based on whether there was concomitant vascular compression (A) or not (B). The outcomes from HFS were statistically significant between them, with group B HFS patients faring worse in follow-up. This suggests that the mechanism

and severity of injury in tumor-related HFS differs based on whether or not there is a vascular compression component. Notably, the only tumor type associated with group B patients was epidermoid cysts. Based on these data, identification and treatment of simultaneous vascular compression in this setting is paramount to optimal long-term outcomes.

Shinya Sato, Yamagata, Japan

Tumor-related hemifacial spasm (HFS) is a relatively rare disease. Although there are some reports about this disease, precise evaluation about tumor and vascular co-compression has not been enough. In this paper, the authors report 55 cases of the tumor-related HFS and discuss an extent of participation in facial spasm between tumor compression and vascular compression. For this reason, this paper seems to be beneficial to many readers of "Neurosurgical Review" journal.

Because majority of their cases are caused by tumor and vascular co-compression at the root exit zone (REZ), the authors concluded that tumor removal and microvascular decompression are required in order to get good outcome. In their series, 12 patients (29.3%) harbored epidermoid cysts, and there was no evidence of vascular compression. For these cases, the authors consider new hypothesis that the HFS might be caused by chemical irritation to the REZ, possibly sterols secreted from the tumor that could cause demyelination in the REZ. However, this hypothesis seems to be controversial because there is no obvious pathological evidence of inflammation around the REZ. Further discussion is necessary in this issue.

In addition, unfortunately, the locations of tumor and the compressing vasculature are ipsilateral in all cases in this paper. There are some reports about the HFS caused by contralateral tumors as mentioned in this paper. In our personal series, there is a case of the HFS associated with a contralateral tumor. In such cases, the decision of surgical tactics is more complex than in ipsilateral tumors. To establish surgical treatment for the tumor-related HFS, further evaluation including the HFS associated with the contralateral tumors is necessary.