



Surgical management of collision-tumors between vestibular schwannoma and meningioma in the cerebellopontine angle in patients with neurofibromatosis type 2

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Abstract

Introduction When vestibular schwannoma (VS) collides with meningioma at the cerebellopontine angle (CPA), a particular threat occurs. Sudden acceleration of tumor growth and unpredictable involvement of cranial nerves results in a special environment that aggravates management. The goal of this study was to analyze the extent of resection, postoperative facial and cochlear function, surgical strategy, and survival rates in patients with neurofibromatosis type 2 (NF2) with meningioma–vestibular schwannoma (M-VS) collision tumors.

Methods A total of 1284 VS, including 165 NF2 VS were operated at our department between January 2004 and May 2018. Out of these cases, a group of six NF2 patients with seven M-VS collision tumors was found following careful analysis of neuro-radiological data and pathological and surgical reports. Patients were evaluated for extent of tumor resection and, furthermore, postoperative facial and hearing function.

Results Six patients with NF2 with seven M-VS collision tumors were included in this study. Mean age was 32 ± 8.2 years. A gross total resection (GTR) of both colliding tumors was achieved in only one case, a GTR of the meningioma and a subtotal resection (STR) of the VS in four cases and in two cases only, the meningioma was removed. In five of the cases, facial function was intact or good (House and Brackmann grades I–II) at long-term follow-up (mean follow-up 22 months). No mortality occurred during follow-up.

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Conclusions Collision between M and VS at the CPA is a particular phenomenon in NF2 patients that may aggravate the situation with less favorable surgical outcome than NF-2 VS without meningioma.

Keywords Cerebellopontine angle · Collision tumor · Meningioma · Neurofibromatosis type 2 · Vestibular schwannoma

Introduction

Vestibular schwannomas (VS) and meningiomas are the most common lesions of the cerebellopontine angle (CPA), which comprise 80% [2, 3, 6] and 6–15% [1], respectively, of all tumors of the CPA.

The occurrence of both tumors in the same patient is a characteristic of neurofibromatosis type 2 (NF2) [3], and they also exist in rare cases in patients without NF2.

In particular situations, a meningioma can meet a VS within the CPA, forming a tumor conglomerate, the so-called collision tumor [4]. Collision tumors are defined as “histologically distinct tumors arising at independent topographical sites, simultaneously growing, meeting together, and eventually intermingling” [4]. They are very uncommon even in patients with NF2 and are found in only 6% of multiple CPA tumors [5]. Collision tumors appear to be an extraordinary lesion with special features. Its extraordinary growth rate might be explained by the interrelated stimulation between the collision tumors [9] and their behavior appears to be more aggressive than that of single VS in NF 2. Due to this particular growth pattern, collision tumors represent a formidable task in surgical management.

The goal of this retrospective single-center study is to analyze the surgical management of collision tumors between vestibular schwannoma and meningioma in the cerebellopontine angle in patients with NF-2, especially with focus on the surgical strategy, extent of resection, postoperative facial and cochlear function, complications, and survival rates.

Methods and materials

Data collection and inclusion criteria

A total of 1284 VS, including 165 NF2 VS, were operated at our department between January 2004 and May 2018. From these 1284 cases, a group of six patients with M-VS collision tumor was found following careful retrospective analysis of neuroradiological data, pathological reports, and surgical reports. The study was approved by the ethics committee. Patients were considered to have NF2 when they fulfilled the Manchester criteria. Inclusion criteria are shown in Table 1.

Surgical technique and strategies

All patients had been positioned under general anesthesia in a semi-sitting position according to our standards. The anesthesiologic setup included transesophageal echocardiography for the detection of air emboli. All patients underwent a retrosigmoid craniectomy, and a transmeatal tumor removal when the VS removal was attempted [12]. Intraoperative monitoring included sensory evoked potentials (SEP) and motor evoked potentials (MEP) of the upper and lower extremities and MEP and electromyography (EMG) recordings of the facial nerve and the lower cranial nerves, and auditory evoked potentials (AEP) when hearing preservation was attempted.

Surgical strategies included the following: (1) Single removal of the meningioma, (2) removal of the meningioma and partial removal of the VS, or (3) removal of both tumors.

Patients were evaluated for preoperative and postoperative facial and hearing function (according to the House and Brackmann and Gardner–Robertson scale, respectively), extent of tumor resection, complications, and survival rate. Furthermore, the surgical strategy was analyzed.

Follow-up

Clinical examination for facial nerve (according to House and Brackmann’s classification) and hearing function (according to Gardner–Robertson scale) was performed in all cases 3 days after surgery and repeated at the last date of follow-up (within a maximum follow-up of 3 years) after surgery (mean follow-up 22 months, range between 3 and 36 months). In two patients, the longest follow-up was less than 1 year. MRI examinations were performed 3 months after surgery and after 3 months yearly.

Table 1 Inclusion criteria

Inclusion criteria
1. NF 2 according to the Manchester criteria
2. Radiologically confirmed collision between 2 different tumors
3. Two different tumor components confirmed intraoperatively
4. Histopathological finding of meningioma (and schwannoma)

NF 2 neurofibromatosis type 2

Results

Six patients with NF2 (four women and two men) harboring seven M-VS collision tumors were included in this study. Mean age of the patients was 32 ± 8.2 years. Three M-VS collision tumors were located on the right side and four on the left side.

Meningioma was histologically confirmed in all seven cases (Table 2). In five cases, the VS was histologically confirmed (in two cases, VS were not removed). The histopathological examination of the meningiomas showed a single meningotheliomatous meningioma in two cases, a single transitional meningioma in one case, a single psammomatous meningioma in one case, and a combination of two different meningiomas (meningotheliomatous together with atypical, transitional together with psammomatous, and meningotheliomatous together with psammomatous) in three cases.

Radiological findings

The M-VS collision tumor was radiologically confirmed in all cases as a tumor conglomerate containing at least two different tumor characteristics (Fig. 1) with an intermingling zone between them (according to Frassanito et al.'s criteria [4]). All patients had bilateral VS and additional intracranial tumors. In all patients, the M-VS collision tumor resulted in significant brain stem compression or displacement.

Presenting symptoms

The majority of patients (4/6) presented with rapid loss of hearing and symptoms of brain stem compression such as ataxia and difficulty in walking, followed by vestibular symptoms such as nausea and dizziness (3/6). None of the patients had a facial palsy on the side of the collision tumor before surgery, but two of six patients had a facial palsy on the contralateral side before surgery (due to a contralateral VS). Other

symptoms included headache, vomiting, weakness, tinnitus, hemiparesis, and dysphagia.

Tumor resection strategy

There were three different strategies:

1. Only the large meningioma was removed in case of a small VS (with the aim of certain hearing and/or facial nerve preservation).
2. The meningioma was removed, while the VS was only partially removed (with the aim of hearing and/or facial nerve preservation).
3. Both tumors were completely removed due to hearing loss and infiltration of the facial nerve, followed by facial nerve reconstruction.

Extent of resection

A gross total resection (GTR) of both tumors of the M-VS collision tumor was achieved in one case only (1/7). In four cases (4/7), a GTR of the meningioma and a subtotal resection (STR) of the VS was performed, and in two cases, only the meningioma was removed with the aim of preserving the cochlear and facial nerve function.

Radiotherapy

None of the patients received radiotherapy before or after surgery.

Facial nerve function

In five of seven cases, no functional impairment of the facial nerve was detectable (Table 3) at the last follow-up examination within a maximum of 3 years after surgery (mean follow-up after surgery 22 months, range between 3 and 36 month). In one case of total removal of the tumors, no cleavage plane was found, and dissection was continued until a distal and

Table 2 Histopathological results

Case number	VS	Meningioma 1	Meningioma 2
1	Schwannoma (WHO I)	Meningotheliomatous (WHO I)	Atypical (WHO II)
2	Schwannoma (WHO I)	transitional (WHO I)	Psammomatous (WHO I)
3	Schwannoma (WHO I)	Psammomatous (WHO I)	–
4	Schwannoma (WHO I)	Transitional (WHO I)	–
5	No resection of VS	Meningotheliomatous (WHO I)	–
6	No resection of VS	Meningotheliomatous (WHO I)	–
7	Schwannoma (WHO I)	Meningotheliomatous (WHO I)	Psammomatous (WHO I)

VS vestibular schwannoma

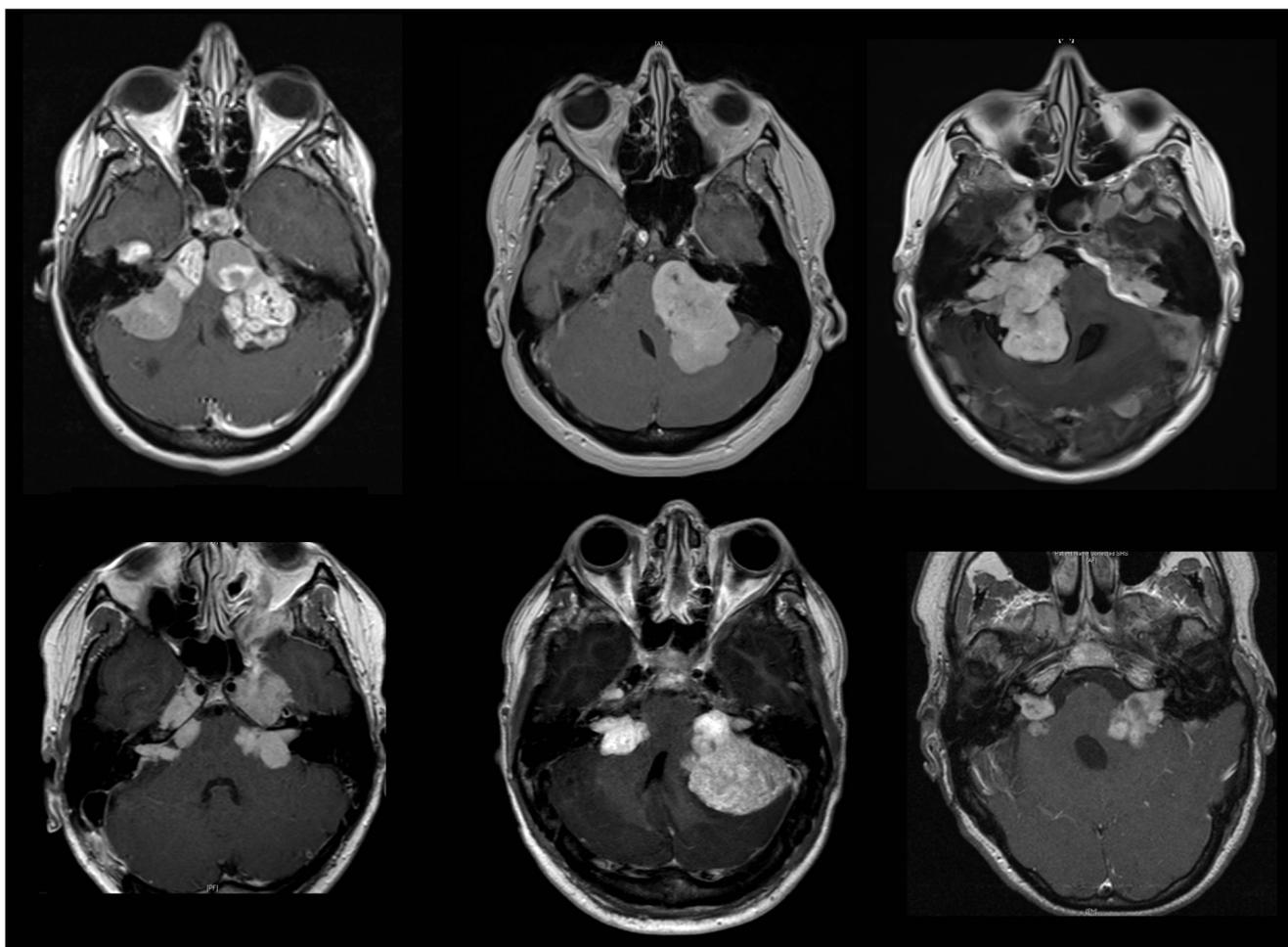


Fig. 1 Preoperative MRI scans of M-VS collision tumors

proximal nerve stumps were identified, and then a facial nerve reanimation using an end-to-end interposed sural nerve graft was indicated. The last follow-up examination (33 months after surgery) showed a partial facial nerve recovery up to HB grade III. A further patient had also a HB grade III palsy in long-term follow-up (31 months after surgery).

Cochlear nerve function

Four of six patients were deaf before surgery (Table 4). The remaining two patients had preoperatively some degree of a hearing impairment (grade II regarding the Gardner–Robertson scale). In those cases, a conservative removal was

Table 3 Facial nerve results

Case number	Extent of resection	Facial function (HB grade)		
		Preop FF	Postop FF	Last FU
1	GTR of M-VS collision tumor	I	VI	III
2	GTR of M, STR of VS	I	I	I
3	GTR of M, STR of VS	I	II	I
4	GTR of M, STR of VS	I	IV	III
5	GTR of M	I	II	I
6	GTR of M	I	I	I
7	GTR of M, STR of VS	I	I	I

HB House-Brackmann, *FF* facial function, *GTR* gross total resection, *STR* subtotal resection, *M* meningioma, *VS* vestibular schwannoma

Table 4 Preoperative and postoperative cochlear function

Case number	Extent of resection	Preop cochlear function (GR grade)	Postop cochlear function (GR grade)	ABI
1	GTR of M-VS collision tumor	V	V	No
2	GTR of M, STR of VS	V	V	Useful auditory sensation
3	GTR of M, STR of VS	II	II	No
4	GTR of M, STR of VS	V	V	No
5	GTR of M	II	II	No
6	GTR of M	V	V	No
7	GTR of M, STR of VS	V	V	No

preop preoperative, *postop* postoperative, *GR* Gardner–Robertson, *ABI* auditory brainstem implant, *GTR* gross total resection, *M* meningioma, *VS* vestibular schwannoma, *STR* subtotal resection

preferred to preserve hearing (in one case, single removal of the meningioma, and in the second case, complete resection of the meningioma with partial resection of the VS was done). In both patients, the hearing function remained stable according to the Gardner–Robertson scale 3 days after surgery.

In the patient with a bilateral M-VS collision tumor and deafness, an auditory brain stem implant (ABI) was placed following removal of the second M-VS collision tumor (Table 4). This patient regained auditory sensation following the ABI placement.

Complications

Complications included one case of pneumonia due to transient impairment of lower cranial nerves. One patient developed impairment of vision due to a meningioma of the optic canal and needed decompression of the optic canal and partial removal of the meningioma. Vision improved immediately. No mortality occurred during follow-up.

Discussion

Different theories of pathophysiology of M-VS collision tumors have been discussed so far, including the autocrine–paracrine growth-stimulatory mechanism [4, 8, 9]. In M-VS collision tumor, special problems arise due to the accelerated tumor growth, unpredictability of anatomical displacement of the cranial nerves, and more severe attachment of the tumor to the cranial nerves.

Surgical strategy

Samii et al. [11] reported that the indication and the timing of tumor resection in patients with NF 2 are in some aspects different from normal VS handling and are dependent on the tumor extension and the related necessity of brain stem decompression and on the auditory function. The management

of M-VS collision tumors in patients with NF 2 involves some further aspects.

In our study, a GTR of both tumors of the M-VS collision tumor was achieved in one case only, while in four cases (4/7), a GTR of the meningioma and a subtotal resection (STR) of the VS was performed, and in two cases, only the meningioma was removed with the aim of preserving the cochlear and facial nerve function.

Kim et al. [7] concluded for the management of VS in case of NF 2 that usually complete tumor removal is the aim of the surgery, but not disregarding functional worsening.

In Samii et al.'s study, the total resection rate in 210 VS was 85% [10]. Graffeo et al. [5] on other hand reviewed that in case of multiple cerebellopontine angle tumors, a GTR was achieved in 50%, while in the specific subgroup of synchronous tumors, it was 52%.

The management of the facial nerve in case of M-VS collision tumors comprises different strategies. In five of seven cases of our study, no functional impairment of the facial nerve was detectable at the last follow-up examination, while two cases showed a HB grade III palsy in long-term follow-up. In one of these cases, a facial nerve

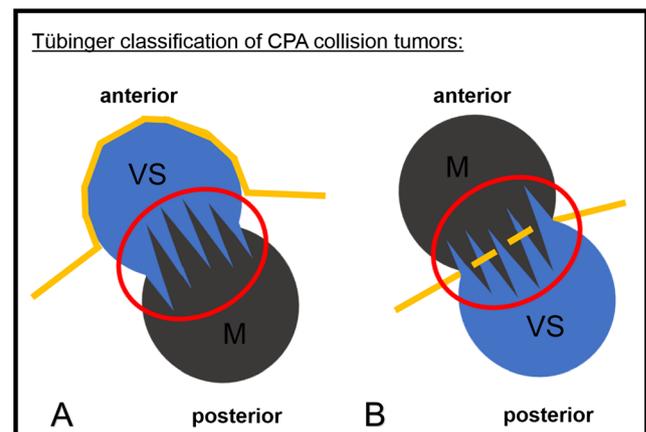
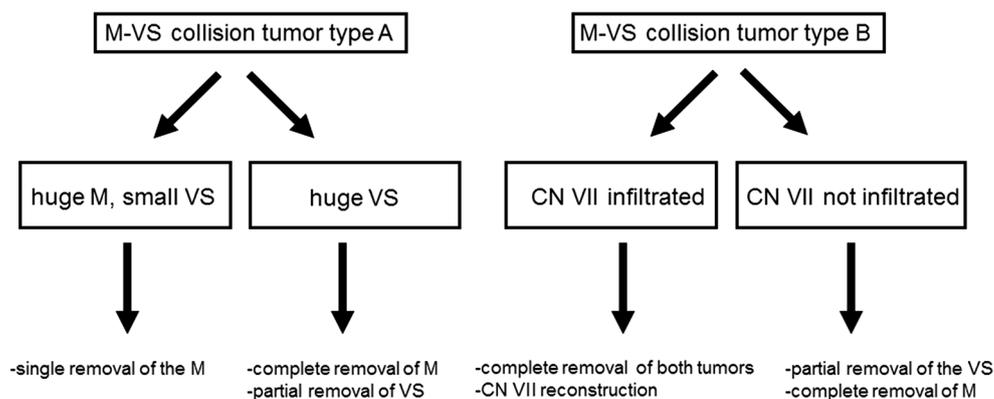


Fig. 2 Classification of CPA collision tumors. **a** Type A: nerves are displaced by one of the tumors. **b** Type B: nerves are compressed/infiltrated between both tumors (red, intermingling zone; yellow, facial nerve; VS, vestibular schwannoma; M, meningioma)

Fig. 3 Algorithm for the management of M-VS collision tumor (M, meningioma; VS, vestibular schwannoma; CN, cranial nerve)



reanimation using an end-to-end interposed sural nerve graft had been performed before.

The anatomic facial nerve preservation in Samii et al.'s study [11] was achieved in 85% and in a later study from 2008 [10] in 89%. In 14% of the cases, a facial nerve reconstruction by sural grafting was indicated [11].

Graffeo et al. [5] concluded that in case of collision tumors, there is a higher risk of poor facial nerve outcomes, with an overall incidence of unfavorable postoperative function in 27%. Moreover, they [5] reported that among patients whose multiple tumor combination included VS unfavorable HB grade III–VI function was documented in 33% of patients.

The topography of the meningioma is also of paramount importance when selecting the correct strategy.

We defined two types of M-VS collision tumor, regarding their topography. In type A, the nerves are displaced by one of the tumors (Fig. 2a), while in type B, the nerve is compressed between both tumors and, in some cases, also is infiltrated by them (Fig. 2b). From a surgical perspective, it is more difficult to remove a meningioma, which is located anterior to the VS than that located posterior to it, because in these cases, the cranial nerves are situated between both tumors.

In the situation of a type A collision tumor with a small vestibular schwannoma and a large meningioma, a single removal of the meningioma will allow brainstem decompression without endangering facial and hearing functions. This strategy was used in two of seven cases. In the case of type A collision tumor with a large VS, treatment may include meningioma removal and partial resection of VS in order to preserve both the facial nerve and cochlear function. This strategy was used in four of seven cases.

In the case of a type B constellation with a collision between an extremely large vestibular schwannoma and a large meningioma, the facial nerve may be damaged between the tumors. In such cases, complete removal of both tumors and a facial nerve reconstruction using a sural nerve graft can be performed. This strategy was necessary in one of seven cases.

Figure 3 summarizes the surgical management according to the different collision tumor types.

In our opinion, an early resection of two nearly touching tumors should be discussed in future, to prevent the tumors stimulating each other and triggering more aggressive behavior.

Limitations

The main limitation of this study is the small number of patients, due to the rare prevalence of this tumor constellation. For that reason, it is important to share the experience between different centers. A further limitation is the retrospective design of the study and the high range of long-term follow-up.

Conclusions

The surgical management of M-VS collision tumor, particularly in patients with NF 2 is very complex. The facial nerve may be infiltrated or damaged from “both sides.” Different strategies (depending on the preoperative symptoms of the patient and the topography of the tumors) include (1) single removal of the meningioma, (2) removal of the meningioma and partial removal of the VS, or (3) complete removal of both tumors with (if necessary) facial nerve reconstruction.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval This study was performed in accordance with the ethical standards of the institutional research committee (University of Tuebingen/Ethic Commission) and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent was obtained from all individual participants included in the study.

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