Factors of influence upon overall survival in the treatment of intracranial MPNSTs. Review of the literature and report of a case

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Abstract

Background: Intracranial malignant peripheral nerve sheath tumors are rare entities that carry a poor prognosis. To date, there are no established therapeutic strategies for these tumors.

Methods: We review the present treatment modalities and present the current therapeutic dilemmas. We perform a statistical analysis to evaluate the prognostic factors for Overall Survival of these patients. Additionally, we present our experience with a 64-year-old man with a MPNST of the left cerebellopontine angle.

Results: To our best knowledge, forty three patients with intracranial MPNSTs, including our case, have been published in the international literature. Our analysis showed gross total resection, radiotherapy and female gender to be beneficial prognostic factors of survival in the univariate analysis. Gross total resection was recognized as the only independent predictor of prolonged Overall Survival. In our case, we performed a gross total resection followed for the first time by stereotactically guided radiotherapy.

Conclusion: Considering the results of the statistical analysis and the known advantages of the stereotaxy, we suggest aggressive surgery followed by stereotactically guided radiotherapy as therapy of choice.

Background

Malignant Peripheral Nerve Sheath Tumors (MPNST) usually arise de novo or from a malignant transformation of a neurofibroma. Rarely MPNSTs may arise from schwannoma, ganglioneuroma or phaeochromocytoma [1,2]. Incidence rates of MPNSTs are identified at less than 1/10^6/year, with the majority of cases located in the brachial or lumbal plexus. Their intracranial occurrence is even more sporadic. To date, no generally accepted therapeutic strategies or prognostic factors of intracranial MPNSTs are established.

To our best knowledge, 42 cases of intracranial MPNSTs have been reported in the literature, 16 of them concerning the VIIIth nerve [3-13]. We review the applied therapies and identify prognostic factors of OS for these tumors.

Furthermore, we present a case of a MPNST of the VIIIth nerve, and propose a novel therapeutic strategy consisting of aggressive surgical resection followed by stereotactically guided radiotherapy.

Methods

Twenty case reports and four retrospective clinical studies concerning intracranial MPNSTs were identified using the NCBI PubMed. No limitations regarding the language or time of publication were imposed on the search process. Two studies concerned MPNSTs as a whole, including tumors of the head and neck, without specifying whether the latter were extracranial or intracranial [14,15]. Thus, they were excluded from our review analysis. Similarly excluded were the cases of MPNSTs arising from extracranial trigeminal branches.

Overall survival (OS) was analyzed with the Kaplan-Meier method. Assessments of potential prognostic factors were carried out using log-rank tests. The multivariate analysis was performed using the Cox Regression Hazard Models- Backward Stepwise Procedure. P values ≤ 0.05 were regarded significant.
Results

A total of forty three patients with intracranial MPNSTs, including our case, were identified. The mean age was 37.6 ± 20.3 (3-69) years. A male predominance (30 males, 69.8%) was observed. 63.9% of the MPNSTs arised de novo; the rest derived from benign tumors. NF1 was present in 17.1% of the patients. Gross total resection (GTR) was achieved in 42.9% whereas 51.3% and 2.3% of the patients received postoperative adjuvant radiotherapy (RT) and chemotherapy, respectively (Table 1-[3-5,7-10,12,13,16-26]). When administrated, radiotherapy was usually whole brain radiation with 60 Gy fractioned over 6 weeks.

Table 1 Review of published cases of intracranial MPNSTs

| No | Age | Gender | Author, Year [Ref.] | Site | HRT* | NF1 | MT* | Resection | RT | Chemo | OS | Death | DM/R* |
|----|-----|--------|---------------------|------|------|-----|-----|------------|----|-------|----|-------|------|
| 1  | 13  | M      | Ducatman, 1984 [17] | L CN* VII | NR* | no  | NR  | NR         | NR | no    | NR | NR     | NR   |
| 2  | 18  | M      | Bruner, 1984 [30]   | frontal   | NR  | no  | no  | GTR*       | no | no    | 66 | no     | R    |
| 3  | 15  | M      | Stefanko, 1986 [21] | L parietooccipital | NR | NR  | no  | GTR       | yes | yes   | 9  | yes    | NR   |
| 4  | 24  | F      | Best, 1987 [31]     | R CPA*    | NR  | no  | no  | IR*        | no | no    | 4  | yes    | NR   |
| 5  | 54  | M      | Matsumoto,1990 [13] | R CPA, CN VII | no | no  | NR  | GTR       | yes | 9     | yes | NR     |      |
| 6  | 47  | F      | Han, 1992 [32]      | R CPA     | no  | no  | no  | IR        | no | no    | 11 | yes    | NR   |
| 7  | 38  | M      | Maeda, 1993 [33]    | R CPA, CN VII | no | no  | no  | IR        | no | no    | 2  | yes    | NR   |
| 8  | 61  | F      | Singh, 1993 [34]    | R cerebellum | NR | NR  | no  | GTR       | yes | 18    | yes | NR     |      |
| 9  | 8   | F      | Sharma, 1998 [9]    | R temporal lobe | no | no  | no  | GTR       | yes | 17    | no  | NR     |      |
| 10 | 44  | M      | Comey, 1998 [35]    | R CPA, CN VILI | yes | yes | yes | IR        | no | no    | 12 | yes    | R    |
| 11 | 69  | M      | Saito,2000 [12]     | L CPA, CN VIII | no | NR  | NR  | IR        | no | no    | 3  | no     | NR   |
| 12 | 4   | F      | Tanaka, 2000 [36]   | R parietooccipital | NR | no  | no  | GTR       | no | no    | 19 | no     | NR   |
| 13 | 30  | F      | Akimoto, 2000 [37]  | L CN V1    | no  | no  | no  | IR        | yes | 16    | yes | R      |      |
| 14 | 57  | F      | Hanabusa,2001 [10]  | R CPA, CN VIII | yes | yes | yes | IR        | yes | 13    | yes | R      |      |
| 15 | 13  | F      | Stark, 2001 [38]    | L CN V2    | no  | no  | no  | GTR       | yes | 14    | yes | R      |      |
| 16 | 36  | M      | Ueda, 2004 [39]     | R+L CN V   | no  | no  | no  | IR        | no | no    | 10 | yes    | R    |
| 17 | 43  | F      | Gonzalez,2007 [11]  | L CPA, CN VIII | NR | no  | yes | GTR       | yes | 8     | yes | M      |      |
| 18 | 38  | M      | Keyenbuhl, 2007 [4] | ina- suprasellar | yes | yes | yes | IR        | yes | 3     | no  | no     |      |
| 19 | 62  | M      | Miliaras, 2008 [5]  | L temporal lobe | no | no  | no  | GTR       | yes | 13    | yes | R      |      |
| 20 | 40  | F      | Chibbaro, 2008 [3]  | L CN V2    | no  | no  | no  | IR        | yes | 21    | no  | no     |      |
| 21 | 8   | M      | Chen, 2008 [7]      | L CN V     | no  | no  | yes | GTR       | no | no    | 8  | yes    | R    |
| 22 | 43  | M      | Chen, 2008 [7]      | L occipital | no  | yes | yes | IR        | yes | 4     | yes | R      |      |
| 23 | 3   | M      | Chen, 2008 [7]      | L CN V, CS* | NR | NR  | no  | IR        | no | no    | 4  | yes    | R    |
| 24 | 35  | M      | Chen, 2008 [7]      | L CN V, CS | NR  | no  | no  | IR        | no | no    | 2  | yes    | NR   |
| 25 | 46  | F      | Chen, 2008 [7]      | L CN V, CS | NR  | no  | no  | GTR       | yes | 60    | no  | no     |      |
| 26 | 62  | F      | Chen, 2008 [7]      | L CPA, CN VII | no | no  | no  | GTR       | no | no    | 4  | yes    | NR   |
| 27 | 5   | M      | Chen, 2008 [7]      | R V1,orbita | NR  | no  | no  | GTR       | no | no    | 9  | yes    | NR   |
| 28 | 32  | M      | Scheithauer, 2009 [8] | R CPA, CN VILI | yes | no  | yes | IR        | yes | 5     | yes | M      |      |
| 29 | 67  | M      | Scheithauer, 2009 [8] | R CPA, CN VIII | no | no  | yes | IR        | no | no    | 1  | yes    | NR   |
| 30 | 56  | M      | Scheithauer, 2009 [8] | R CPA, CN VIII | no | no  | yes | IR        | no | no    | 2  | yes    | R    |
| 31 | 32  | M      | Scheithauer, 2009 [8] | R CPA, CN VIII | no | no  | yes | IR        | no | no    | 3  | yes    | R    |
| 32 | 26  | F      | Scheithauer, 2009 [8] | R CPA, CN VII | no | no  | yes | IR        | yes | NR    | NR  | NR     |      |
| 33 | 5   | M      | Scheithauer, 2009 [8] | R CPA, CN VIII | no | no  | no  | NR        | no | no    | NR  | NR     |      |
| 34 | 69  | M      | Scheithauer, 2009 [8] | R frontal lobe | no | no  | no  | NR        | no | no    | 4  | yes    | R    |
| 35 | 50  | M      | Scheithauer, 2009 [8] | L CN VII | no | NR  | yes | GTR       | yes | 17    | yes | NR     |      |
| 36 | 26  | M      | Scheithauer, 2009 [8] | posterior fossa | NR | NR  | NR  | NR        | no | no    | NR  | NR     |      |
| 37 | 50  | M      | Scheithauer, 2009 [8] | L CPA | NR  | NR  | NR  | NR        | no | no    | 36 | yes    | R    |
| 38 | 30  | M      | Scheithauer, 2009 [8] | optic chiasma | yes | NR  | NR  | NR        | no | no    | 2  | yes    | R    |
| 39 | 59  | M      | Scheithauer, 2009 [8] | L gasserion ganglion | NR | NR  | NR  | NR        | no | no    | NR  | NR     |      |
| 40 | 41  | M      | Scheithauer, 2009 [8] | posterior fossa | NR | no  | no  | NR        | yes | 5     | yes | M      |      |
| 41 | 32  | M      | Scheithauer, 2009 [8] | CN X        | yes | yes | yes | IR        | yes | no    | NR  | NR     | M    |
| 42 | 62  | M      | Ziadi, 2010 [40]    | L CN V3    | no  | no  | no  | GTR       | yes | 17    | no  | no     |      |
| 43 | 64  | M      | present study       | L CPA, CN VIII | no | yes | yes | GTR       | yes | 12    | no  | no     |      |

*HRT: History of radiation exposure, MT: malignant transformation of a former benign entity (mainly neurofibroma or schwannoma), DM/R: distant metastasis/recurrence, NR: not reported, GTR: gross total resection, IR: incomplete resection, CN: cranial nerve, CPA: cerebellopontine angle, CS: cavernous sinus.
Median OS was 9 months. Progression free survival was not documented in the majority of the cases, and could not be evaluated.

In the univariate analysis, female gender (p = 0.048), GTR (p = 0.004) and RT (p = 0.010) were significant beneficial factors for OS (Figure 1). Notably, younger age, malignant transformation of a former benign tumor and the presence of NF1 did not significantly influence outcome (p > 0.05) (Table 2).

Some factors of potential influence upon OS, such as histological grade and tumour size, were not estimated due to the lack of reported data.

We included the significant factors above in a multivariate analysis, using the backward stepwise procedure. GTR was found to be an independent beneficial prognostic factor for OS (HR = 0.258, CI 95% 0.102-0.653, p = 0.004) (Table 2).

**Illustrative Case**

A 64-year-old man presented with progressive headache, vertigo, nausea, hypogeusia and ataxia commencing 3 weeks prior to admission. A left hearing loss was known since three decades. A brain MRI approximately 10 years prior to admission revealed a small tumor localized at the left cerebellopontine angle. There were no history or clinical stigmata of Neurofibromatosis types 1 and 2.

Preoperative MRI and CT demonstrate a 3.5*4 cm measuring well delineated contrast-enhancing lesion in the left cerebellopontine angle with mass effect (Figure 2A, B). A thoracoabdominal CT as well as MRI of brachial and lumbal plexus performed ulteriorly excluded other manifestations of the MPNST.

A gross total tumor resection using neuromonitoring of the motor tract and facial nerve function was achieved. Postoperatively, a transient facial nerve palsy House-Brackmann grade III occurred as sole complication.

Histopathological examination revealed a highly cellular tumor with considerable cytologic atypia. (Figure 3). Immunohistochemical examinations revealed only focal immunoreactivity for antibodies against S-100-protein and p75. Tumors cells were strongly immunopositive for vimentin and variable immunoreative for CD99 and

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**Table 2 Statistical Analysis**

|                      | Univariate Analysis* | Multivariate Analysis *** |
|----------------------|----------------------|--------------------------|
|                      | gender | resection | RT | age** | NF1 | MT          |
| Log Rank             | p      | p         | p  | p     | p   | p           |
| Overall Survival     | 0.048  | 0.004     | 0.010 | p     | p   | p           |
| Resection (GTR vs IR)| p = 0.004 | HR = 0.258 | CI 95% (0.102-0.653) | 0.756 | 0.132 | 0.140 |
| Gender (female)      | p = 0.059 | HR = 0.401 | CI 95% (0.155-1.037) |                   |       |             |

*Kaplan-Meier method and Log Rank test
** over or under 37.6 years old (mean age)
***Cox proportional hazards model
Figure 2 Preoperative (A+B) and postoperative (C+D) MRIs: (A+C)Axial T1Wse without and (B+D)with contrast. MRI findings: Enlargement of the left IAC. In non-contrast T1w homogeneous intermediate signal mass in the CPA-IAC cistern on the left with displacement of the middle cerebellar peduncle and strong enhancement after contrast administration. No intramural cysts and no dural tail. C+D, no residual tumor is shown.

Figure 3 Histopathological examination revealed a highly cellular tumor with considerable cytologic atypia. The cytomorphological aspect was dominated by spindle cells with eosinophilic cytoplasm and nuclear enlargement as well as hyperchromasia. Brisk mitotic activity was present, whereas necrosis was no significant feature of the tumor (bar graph - 200 μm).
Bcl-2. The tumor was classified as grade II according to FNCLCC grading system [27].

Four weeks after surgery, the patient underwent fractionated stereotactic and image guided radiotherapy using single isocentre dose delivery. A total of 60 Gy was delivered in 30 fractions. The treatment was performed using the Novalis(r) system with micro-multileaf-collimator and ExacTrac(r). The patient was immobilized using a relocatable stereotactic frame with an aquaplast mask (all components by BrainLAB(r), Germany). Because there was no detectable residual tumour on post operative MRI (Figure 2C, D), the CTV (clinical target volume) was defined as the former tumour cavity which was delineated by fusing the pre- and post-op T1 MRI sequences with contrast enhancement. The safety margin was set to 2 mm receiving the PTV (planning target volume) of 19.026 cc, (Figure 4A, B). By using 8 non-coplanar conformal static beams the 90% isodose encompassing PTV with a conformity index of 1.52. All delivery parameters were according to the guidelines of RTOG (Figure 4C, D, E, F).

The radiotherapy was well tolerated without acute toxicities. Clinical and MRI follow up at 12 months is without any hints of tumour recurrence.

**Discussion**

In contrast to their benign counterparts, neurofibromas or schwannomas, intracranial MPNSTs carry a poor prognosis with a median OS of 9 months, (range 1 to 66 months, present review). In combined series of intracranial and extracranial MPNSTs, Zou et al report a 5-year survival rate of 38.7%, whereas Anghileri et al described a 5-year cause-specific mortality of 39.9%. When the influence of tumor site is considered, Anghileri reported an increased 5-year mortality of head and neck MPNSTs of 66.7%, as compared to 48.8% and 27.5% of trunk and extremities MPNSTs, respectively. The rarity of intracranial MPNSTs hampers the establishment of evidence based strategies for their optimal treatment. Thus, the management of the intracranial MPNSTs should also consider the experience gained from the treatment of extracranial MPNSTs.

Anghileri et al conducted a study of 205 patients with MPNSTs, of which 9 cases were head and neck tumors, and found that GTR, achieved in 62% of the patients, correlated significantly with longer OS, and inversely with local recurrence on multivariate analysis [14]. Zou et al carried out another study of 140 patients with MPNSTs, including 20 tumours of the head and neck,
and showed that a complete surgical resection was inversely related to local recurrence on univariate analysis [15]. The results of the present review verify for intracranial MPNSTs the statistically significant influence of GTR upon OS in the univariate and multivariate analysis. Thus, a main goal in the treatment of the intracranial MPNSTs should be the complete surgical tumour resection with preservation of neurological function, whenever applicable.

The role of adjuvant radiotherapy remains controversial. Some studies suggest that radiation may be implicated in the pathogenesis of MPNSTs [8,28]. Foley et al suggested that ionizing radiation may cause chromosomal injury and induce proliferation as well as cytologic atypia in Schwann cells, resulting in radiation-induced MPNSTs [29]. In our review series, 41.7% of patients harbouring a malignant transformation to MPNST received radiation in their history. Other studies haven't shown any positive effects of radiotherapy on patients outcome[30-32], while the recent literature indicates the beneficial role of the radiotherapy in local control of disease after a total or a near total resection of extracranial MPNSTs [14,33-38]. Anghileri et al found adjuvant radiotherapy to be significantly related to longer OS on multivariate analysis, while no correlations with local recurrence or distant metastases were observed [14].

The radiation dosage administrated in the majority of the cases was 50 - 60 Gy. Our review revealed the beneficial prognostic significance of adjuvant radiotherapy for OS in the univariate analysis. However, the multivariate analysis failed to show an independent influence of RT on OS. This could be related to the limited sample of patients. Considering the above findings and the highly malignant histological appearance of the tumour, in our patient we decided for adjuvant radiotherapy with stereotactic guidance due to its precise dosage delivery while sparing the adjacent healthy brain tissue. This strategy provides the possibility to apply an adequate high dose of 60 Gy despite of nearby sensitive risk structures like the brainstem. Thus, we were able to take advantages of both stereotactic radiotherapy and conventional fractionation while minimising the risks of RT-inducing brain injury like radiation necrosis and cognitive decline.

The optimal radiation dose has not yet been defined. We decided for a total dose of 60 Gy balancing the relatively high radiation dose to the highly malignant histological tumour appearance.

Some authors consider MPNSTs to be chemotherapy-resistant [28] while others suggest that surgery followed by combined radiochemotherapy results in improved survival [39]. Two recent studies of large series of peripheral MPNSTs failed to show any benefit of chemotherapy [7,34]. Therefore, in our patient, chemotherapy was decided to be spared for the case of tumour relapse or metastatic disease.

In the present patient the MPNST seems to have resulted from the malignant transformation of a pre-existing benign schwannoma. 36.1% of the review cases experience a progression of benign tumor to malignancy, having a worse OS compared to MPNSTs arising de novo. The latter difference though did not reach statistical significance (8.46 vs 22.95 months, p = 0.140). These observations point out the importance of a thorough long-time follow-up of all benign intracranial schwannomas and neurofibromas that have not been resected. However, it is not clear whether MRI follow-up can reliably indicate the exceptional transition of a schwannoma to a MPNST. Approximately, 25 to 50% of MPNSTs are associated with NF-1. The overall lifetime risk of genesis of MPNST in patients with NF-1 is estimated to be from 8 to 13% [14,40]. In the present review 17.1% of intracranial MPNSTs were related to NF-1.

It is noteworthy, that the female gender is less likely to present with intracranial MPNST and that females harbouring this tumour have a significant longer OS than men. Further studies are needed to enlighten the background of these observations.

Conclusion
In conclusion, we propose as therapeutic strategy for intracranial MPNST consisting of the maximal surgical resection feasible with preservation of neurological function, followed by adjuvant stereotactically guided radiotherapy. This strategy minimises the possible complications of surgery as well as of brain radiation. Chemotherapy should probably be spared for relapsed or metastasized disease.

Abbreviations
CIV: Clinical target volume; GTR: Gross total resection; MPNST: Malignant peripheral nerve sheath tumor; NF1: Neurofibromatosis 1; OS: Overall survival; PTV: Planning target volume; RTOG: Radiation therapy oncology group for stereotactic radiotherapy.

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Authors’ contributions
All of the authors have been involved in drafting this paper and have read and approved the final manuscript. KG conceived the idea of the paper, reported the case, performed the literature research and statistical analysis, wrote the paper; was the attending physician-resident during the stay of the patient at Hospital and follow up the patient through tel.interviews each month. JB managed the patient concerning the stereotactically guided...
radiotherapy (in another clinic), wrote the part of the paper concerning radiotherapy and followed up the patient at his outpatient clinic. AK was the pathologist who examined the tissue and wrote the part of the pathology evaluation. IW performed the ETN examination preoperatively and postoperatively, as well as performed with KG the relevant literature research. RK was the neurosurgeon who operated the patient, was the supervisor of the clinic admitted the patient, decided for the therapy procedures and revised the manuscript. All authors read and approved the final draft.

Competing interests
The authors declare that they have no competing interests.

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References
1. World Health Organization. Pathology and Genetics of Tumors of the Nervous System. Lyon: IARC Press; 2000.
2. Al-Ghantamy M, Midha R, Guha A, Jacobs WB. Malignant periphere nerve tumors. In Textbook of Neuro-oncology. Edited by: Beyer MS, Prados MD. Philadelphia, Elsevier Saunders; 2003:364-571.
3. Chibbaro S, Heiman P, Povilka M, George B. Malignant trigeminal schwannoma extending into the anterior skull base. Acta Neurochir (Wien) 2008, 150:599-604.
4. Krayenbuehl N, Heppner F, Yonekawa Y, Bernays RL. Intranasal malignant peripheral nerve sheath tumor. Acta Neurochir (Wien) 2007, 149:201-206.
5. Milianas G, Tsiopoulos P, Aspros I, Tsietsis P, Polyzoidis K. Malignant orbital schwannoma with massive intracranial recurrence. Acta Neurochir (Wien) 2008, 150:1291-1294.
6. Kumar P, Jaiswal S, Agrawal T, Datta NR. Malignant peripheral nerve sheath tumor of intracranial trigeminal nerve. A case report and review of the literature. Neurosurg Rev 2003, 16(1):1334-1335.
7. Chen L, Mao Y, Chen H, Zhou LF. Diagnosis and management of intracranial malignant peripheral nerve sheath tumors. Neurosurgery 2008, 62(4):825-832.
8. Scheithauer BW, Ergodan S, Rodriguez FJ, Burger PC, Woodruff JM, Kros JM, Golden M, Spinner RJ. Malignant Peripheral Nerve Sheath Tumors of Cranial Nerves and Intracranial Contents: A Clinico-pathologic Study of 17 Cases. Am J Surg Pathol 2009, 33:525-338.
9. Sharma S, Abbott R, Zaggag D. Malignant Intracerebral Nerve Sheath Tumor. A case report and review of the literature. Cancer 1998, 82(3):545-552.
10. Hanabusa K, Morikawa A, Murata T, Hanabusa K, Morikawa A, Murata T, Taki Y. Acoustic neuroma with malignant transformation. J Neurosurg 2001, 95:518-521.
11. Gonzalez LF, Lekovic GP, Eschbacher J, Coons S, Spetzler RF. Malignant peripheral nerve sheath tumor with divergent cartilage differentiation from the acoustic nerve case report. No To Shinkei Geka 1990, 18(1):59-62.
12. Saito T, Oki S, Mikami T, Kawamoto Y, Hayashi Y, Yuki K. Malignant peripheral nerve sheath tumor with divergent cartilage differentiation from the acoustic nerve: case report. No To Shinkei Geka 2000, 52B(8):734-739.
13. Matsumoto M, Sakata Y, Sanpei K, Onagi A, Terao H, Kudo M. Malignant schwannoma of the ean nerve: case report. No To Shinkei Geka 1990, 18(1):59-62.
14. Anghieri M, Miceli R, Fiore M, Mariani L, Ferrari A, Mussi C, Lozza L, Collini P, Olmi P, Casali PG, Pilotta S, Gronchi A. Malignant peripheral nerve sheath tumors: Prognostic factors and survival in a series of patients treated at a single institute. Cancer 2006, 107:1065-1074.
15. Zou C, Smith KD, Liu J, Lahat G, Myers S, Wang WL, Zhang W, McCutcheon IE, Slopis JM, Lazzar AJ, Pollock RE, Lev D. Clinical, Pathological, and Molecular Variables Predictive of Malignant Peripheral Nerve Sheath Tumor Outcome. Annals of Surg 2009, 249(6):1014-1022.
16. Bruner JM, Humphreys JH, Armstrong D. Immunocytochemistry of recurring intracranial nerve sheath tumor. J Neuropathol Exp Neurol 1984, 43:296.
17. Best PV. Malignant triton tumour in the cerebellopontine angle. Acta Neuropathol 1987, 74:92-96.
18. Han DH, Kim DG, Chi JG, Park SH, Jung HW, Kim YG. Malignant triton tumor of the acoustic nerve. Case report. J Neurosurg 1992, 76:874-877.
19. Maeda M, Joso S, Baba S, Muro H, Hirasawa H, Ichihashi T. Malignant nerve sheath tumor with rhabdomyoblastic differentiation arising from the acoustic nerve. Acta Pathol Jpn 1993, 43(4):198-203.
20. Singh RV, Syas S, Campbell DA, Broome JC. Malignant schwannoma of the cerebellum: Case report. Surg Neurol 1993, 39:128-132.
21. Comey CH, McLaughin MR, Jho HD, Martinez AJ, Lunsford LD. Death from a malignant cerebellopontine angle triton tumor despite stereotactic radiosurgery. Case report. J Neurosurg 1998, 89:653-658.
22. Tanaka M, Shibui S, Nomura K, Nakanishi Y, Hasegawa T, Hirose T. Malignant intracerebral nerve sheath tumor with intratumoral calcification. Case report. J Neurosurg 2000, 92:338-341.
23. Akimoto J, Ito H, Kudo M. Primary intracranial malignant schwannoma of trigeminal nerve. A case report with review of the literature. Acta Neurochir [Wien] 2000, 142(S):591-595.
24. Stark AM, Bult H, Hugo H, Mehboob HM. Malignant Peripheral Nerve Sheath Tumours-Report of 8 Cases and Review of the Literature. Acta Neurochir (Wien) 2001, 143:357-364.
25. Ueda R, Saito R, Horihachi T, Nakamura Y, Ichikizaki K. Malignant peripheral nerve sheath tumour in the anterior skull base associated with neurofibromatosis type 1-case report. Neuroradiol Med Chir 2004, 44(1):38-42.
26. Zaidi A, Saliba I. Malignant peripheral nerve sheath tumor of intracranial trigeminal nerve: A case series review. Auras Neurology 2010, 37:539-545.
27. Fédération Nationale des Centres de Lutte Contre le Cancer. [http://www.fnclcc.fr].
28. Ducatman BS, Scheithauer BW, Piepgras DG, Reiman HM, Istrup DM. Malignant peripheral nerve sheath tumors. A clinopathological study of 120 cases. Cancer 1986, 57:2006-2021.
29. Foley KM, Woodruff JM, Ellis FT, Posner JB. Malignant peripheral nerve sheath tumor: A review of 29 cases. J Neurosurg Sci 2000, 44(1):67-72.
30. Shin M, Ueki K, Kurita H, Kinjo T. Malignant transformation of a vestibular schwannoma after gamma knife radiosurgery. Lancer 2002, 27:309-310.
31. Vathey J, Woodruff JM, Brennan MF. Extensive malignant peripheral nerve sheath tumors (neurogenic sarcomas): A 10-year experience. Ann Surg Oncol 1995, 2:126-131.
32. Stefanko SZ, Vuzenski PD, Maas AI, van Vroonhoven CC. Intracerebral malignant schwannoma. Acta Neurochir (Berl) 1986, 71:321-325.
33. Carl M, Ferrari A, Mattek A, Zanetti I, Casanova M, Bisogno G, Cecchetto G, Alaggio R, De Soo L, Koscielnik E, Sott G, Treuner J, Carl M, Ferrari A, Mattek A. Pediatric malignant peripheral nerve sheath tumor: The Italian and German Soft Tissue Sarcoma Cooperative Group. J Clin Oncol 2005, 23:8422-8430.
34. Gachiani J, Kim D, Nelson A, Kline D. Surgical management of malignant peripheral nerve sheath tumors. Neurosurgery 2007, 2(2):E13.
35. Basco-Rico S. Therapy of malignant schwannomas: Usefulness of an integrated radiologic. Surgical therapy. J Neurosurg Sci 1989, 33:253-257.
36. Ferrer RE, Gutmann DH. International consensus statement on malignant peripheral nerve sheath tumors in neurofibromatosis 1. Cancer Res 2002, 62:1573-1577.
37. Wilson AN, Davis A, Bell RS, O’Sullivan B, Catton C, Madadi F, Kandel R, Formisani VL. Local control of soft tissue sarcoma of the extremity: The experience of a multidisciplinary sarcoma group with definite surgery and radiotherapy. Eur J Cancer 1994, 30:746-751.
38. Wong WW, Hirose T, Scheithauer BW, Schild SE, Gunderson LL. Malignant peripheral nerve sheath tumors. J Radiat Oncol Biol Phys 1998, 42:351-360.
39. Minovi A, Baster O, Hunter B, Draf W, Bockmühl U. Malignant peripheral nerve sheath tumors of the head and neck: management of 10 cases and literature review. Head Neck 2007, 29:439-444.
40. Evans DG, Baser ME, McLaughan J, Sharif S, Howard E, Moran A. Malignant peripheral nerve sheath tumors in neurofibromatosis 1. J Med Genet 2002, 39:311-314.

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