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Review Article

Intracranial malignant peripheral nerve sheath tumor: A case report and comprehensive literature review

Brandon Michael Wilkinson[1], Michael A. Duncan[1], Richard Davila[2], Brian Nicholas[3], Harish Babu[1]

Departments of [1]Neurosurgery and [2]Otolaryngology, State University of New York Upstate Medical University (SUNY), Syracuse, [3]Department of Otolaryngology, White Plains Hospital, White Plains, New York, United States.

E-mail: *Brandon Michael Wilkinson - wilkinsb@upstate.edu; Michael A. Duncan - duncanm@upstate.edu; Richard Davila - davilar@upstate.edu; Brian Nicholas - nicholab@upstate.edu; Harish Babu - babuh@upstate.edu

***Corresponding author:**
Brandon Michael Wilkinson, Department of Neurosurgery, SUNY Upstate Medical University, Syracuse, New York, United States.

wilkinsb@upstate.edu

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ABSTRACT

Background: Malignant peripheral nerve sheath tumors (MPNSTs) are rare malignant soft-tissue sarcomas arising from peripheral nerves. Little data exist regarding MPNST originating intracranially. Here, we present a 7th/8th nerve complex MPNST, discuss the treatment strategy and patient outcome, and provide a comprehensive review of existing literature.

Methods: Using Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines, PubMed and crossed references were queried, yielding 37 publications from 1952 to the present. Fifty-three cases of primary intracranial and extra-axial MPNST were identified.

Results: We additionally report a 40-year-old female presented with acute onset dizziness and subsequent hearing loss with associated right-sided facial numbness. Magnetic resonance imaging revealed a 0.5 cm × 1.7 cm enhancing lesion within the right internal auditory canal extending into the cerebellopontine angle. The patient was initially treated with retro sigmoid craniotomy for tumor resection followed by a trans labyrinth approach for residual tumor resection. She completed adjuvant fractionated radiation therapy and underwent facial nerve transfer to restore complete hemifacial paralysis. The most common cranial nerves involved were V and VIII (43.4% each), with 66% of patients male and 34% female. The average age was 43.4 ± 17.4 years. The mean survival time for reported non-survivors after tissue diagnosis was 15 ± 4 months. Two-year survival for patients receiving gross total resection was 33.3% versus 22.8% with subtotal resection.

Conclusion: MPNSTs comprise a group of highly aggressive neoplasms that rarely arise intracranially. Gross total surgical resection should be pursued when feasible.

Keywords: Cerebellopontine angle, Intracranial, Malignant peripheral nerve sheath tumor, retro sigmoid craniotomy

INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNSTs) are a rare and poorly defined heterogeneous group of malignant neoplasms comprising 5–10% of all malignant soft-tissue sarcomas.[39] Primary risk factors for development include neurofibromatosis type 1 (NF1) and prior radiotherapy. The differential diagnosis is wide and includes neurofibroma, schwannoma, and various soft-tissue sarcomas. NF1-associated MPNST is thought to derive from neoplastic Schwann cells that undergo further mutation to progress to MPNST. Sporadic MPNST has also

been described with a later age of onset and smaller size; however, mechanisms of neoplastic progression are less well known.[33]

Intracranial MPNST is exceptionally rare, with little data guiding diagnosis and treatment strategies. Reported cases show a male predilection with age of onset predominantly in the 5th-6th decades, although widely variable. Rapid tumor growth, lesion size >5 cm, and marked heterogeneity on magnetic resonance imaging (MRI) may suggest MPNST.[43] The most effective treatment involves gross total surgical resection, as adjuvant chemo- and radiotherapy have shown to be largely ineffective in terms of disease progression and survival. Head-and-neck MPNST carries a 5-year cause-specific mortality rate of 56–67%.[44]

CASE REPORT

A 40-year-old female presented initially with acute onset dizziness. She later developed progressive right-sided hearing loss with the right face and tongue numbness. Initial MRI showed a 0.5 cm enhancing lesion within the right internal auditory canal (IAC). Six-month surveillance imaging demonstrated rapid tumor growth with extension into the cerebellopontine angle, measuring 1.7 cm in diameter [Figure 1]. Preoperative hearing testing showed profound right sensorineural hearing loss (AAO-HNS class D). She developed House Brackmann II right facial weakness during the surveillance period. In March 2021, she underwent a right retro sigmoid craniectomy for tumor resection.

On identification of the lesion, hemosiderin deposits were noted as indicative of prior hemorrhage. The tumor appeared highly vascular without clear delineation of the 7th/8th nerve complex. Frozen pathology showed numerous mitotic figures concerning malignant tumors. The decision was made to proceed with maximal total resection with 7th/8th nerve complex sacrifice. Stimulation identified the proximal nerve complex, and dissection proceeded medially to laterally. The firm tumor engulfed the nerves at the porus acusticus; thus, the petrous bone was drilled to expose the mid and lateral IAC. An additional tumor was noted laterally within the vestibule, again without clear delineation from the cranial nerves (CNs). The visible tumor mass was resected en bloc. A 45° endoscope was utilized better to visualize additional tumors within the IAC and vestibule. Further, abnormal-appearing tissue was removed without obvious evidence of gross residual tumor.

Final pathology revealed low-grade MPNST. Postoperatively, the patient had expected complete right facial paralysis (House-Brackmann VI). MRI showed a small 2 mm nodular enhancement within the vestibule concerning for residual tumor [Figure 2]. Two days postoperatively, she underwent right eyelid gold weight implantation. Given the final pathology results, four weeks postoperatively, she underwent a trans labyrinth approach for resection of the residual tumor. Simultaneously, a right masseteric-to-facial and hypoglossal-to-facial nerve transfer was performed to restore facial function. A sternocleidomastoid muscle flap was utilized for labyrinthectomy and eustachian tube closure. As opposed to a fat graft, the muscle flap is raised in the same incision and utilizes vascularized tissue, avoiding fat necrosis. The case was discussed in a multidisciplinary tumor board, and adjuvant radiation therapy (RT) was pursued. The patient

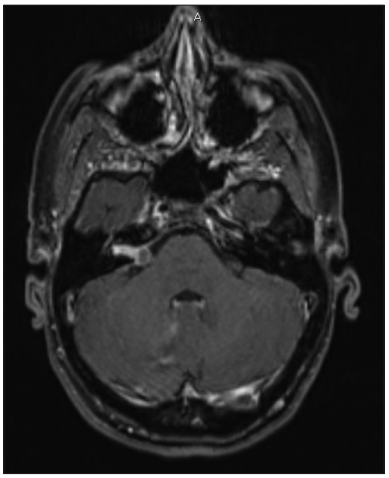

Figure 1: Pre-operative axial T1-weighted magnetic resonance imaging with contrast demonstrating a 1.7 cm enhancing lesion within the right internal auditory canal extending into the right cerebellopontine angle cistern.

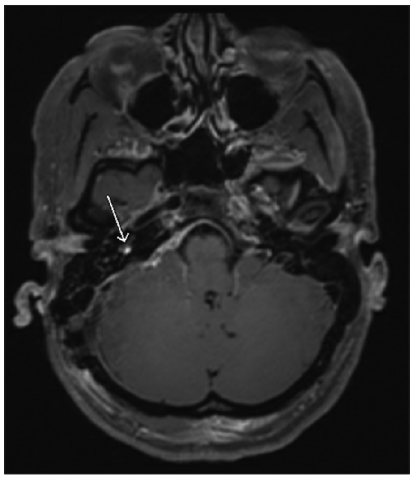

Figure 2: T1-weighted magnetic resonance imaging with contrast following right retro sigmoid craniectomy for tumor resection. Of note a small nodular area of enhancement is seen near the right vestibule (arrow).

completed fractionated RT at 5400 cGy in 30 fractions with a simultaneous integrated boost to 6000 cGY in June 2021. No obvious tumor recurrence was seen on posttreatment MRI after six months [Figure 3]. Meanwhile, the patient underwent genetic testing, and no NF1 or NF2 mutations were detected.

Her right facial weakness gradually improved, and she regained the ability to close her eyes after nine months (House-Brackmann III). Surveillance imaging two years after surgery remains unchanged. She has since returned to work and resumed her day-to-day life.

LITERATURE REVIEW

A systematic review was performed according to the Preferred Reporting Items for Systematic Reviews and Meta-analyses guidelines. A systematic search was conducted in PubMed using the criteria "malignant peripheral nerve sheath tumor," "intracranial," "nerve sheath neoplasms," and combinations of the variables "malignant," "nerve," "sheath," "neurogenic sarcoma," and "schwannoma." The search strategy is outlined in Figure 4.

Inclusion criteria were as follows: (1) must be a case report, case series, or systematic review published between 1952 (first reported case of intracranial MPNST) and November 2023 when the search was performed; (2) study must report intracranial MPNST involving CN(s); (3) study must have reported patient age, sex, and specific CN involved; and (4) study must not have overlapping patients and have full-text article available through SUNY Upstate Medical University. Cases lacking information on specific CN involved, metastatic spread from distant site locations, or solely intra-axial lesions without CN involvement were excluded from the study.

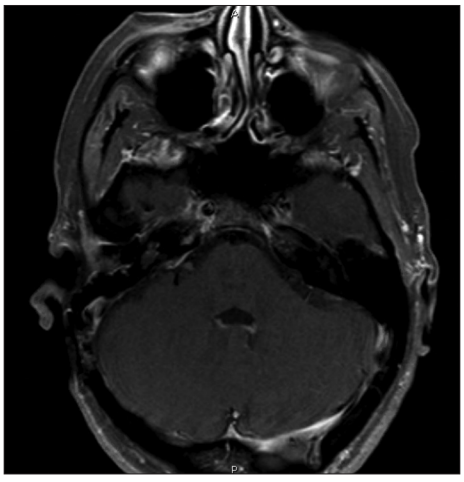

Figure 3: T1-weighted magnetic resonance imaging with contrast obtained 6 months following translabyrinthine approach for resection of residual tumor. Expected postsurgical changes are seen without clear evidence of recurrent enhancing tumors.

Further, data extraction was performed using the following parameters: (1) author, (2) year of publication, (3) patient age and gender, (4) tumor location and specific CN involved, (5) prior surgical treatment or RT, (6) surgical approach, (7) use of adjuvant chemotherapy or radiation postoperatively, and (8) patient outcome including morbidity and mortality.

Our review yielded 37 publications from 1952 to the present describing 53 cases of intracranial MPNST originating from CNs, including our own. Patient demographic data, tumor location, treatment, and outcome are described in Table 1. Thirty-five patients were male (66.0%) and 18 female (34.0%). Eleven patients (20.6%) received cranial RT before the time of pathologic diagnosis, either for nerve sheath tumors or unrelated intracranial tumors. The most common CNs involved were CN V and CN VIII, with an equal number present in our review (43.4% each). Twenty-eight patients (52.8%) received postoperative adjuvant radiotherapy, while only one patient received chemotherapy. Eight patients had a reported history of NF1 or NF2 mutation (15.1%). Results are summarized in Table 2. For reported nonsurvivors on publication, the mean survival time after tissue diagnosis was 15.5 ± 4 months.

DISCUSSION

MPNSTs are aggressive lesions arising from peripheral nerve or nerve sheath cells. Intracranial MPNST remains extremely rare and can be divided into two categories: extra-axial and intraparenchymal tumors. We have provided a comprehensive review of all primary intracranial extra-axial MPNST cases to date.

In addition to radiation exposure, both NF-1 and NF-2 mutations have been implicated in pathogenesis.[7,43] About 15.1% of patients in our review had proven NF mutations, whereas 20.6% had received prior radiotherapy. Neither were present in our patient, suggesting these lesions may arise *de novo,* and their pathogenesis remains poorly understood. Several case reports have described significant pre- and post-operative tumor-associated hemorrhage, leading to increased morbidity and mortality.[16-18] In our patient, hemosiderin deposits were readily apparent on tumor identification, suggesting intracranial MPNST may carry a potential hemorrhage risk.

Intraparenchymal, non-CN-associated MPNST has also been described. Rubino *et al*. reviewed 26 primary cerebral cases of MPNST arising in both supra- and infratentorial regions.[28] As with nerve-associated MPNST, NF mutation appears to be the primary risk factor, with three patients reported having received prior RT as well. The origin of parenchymal MPNST

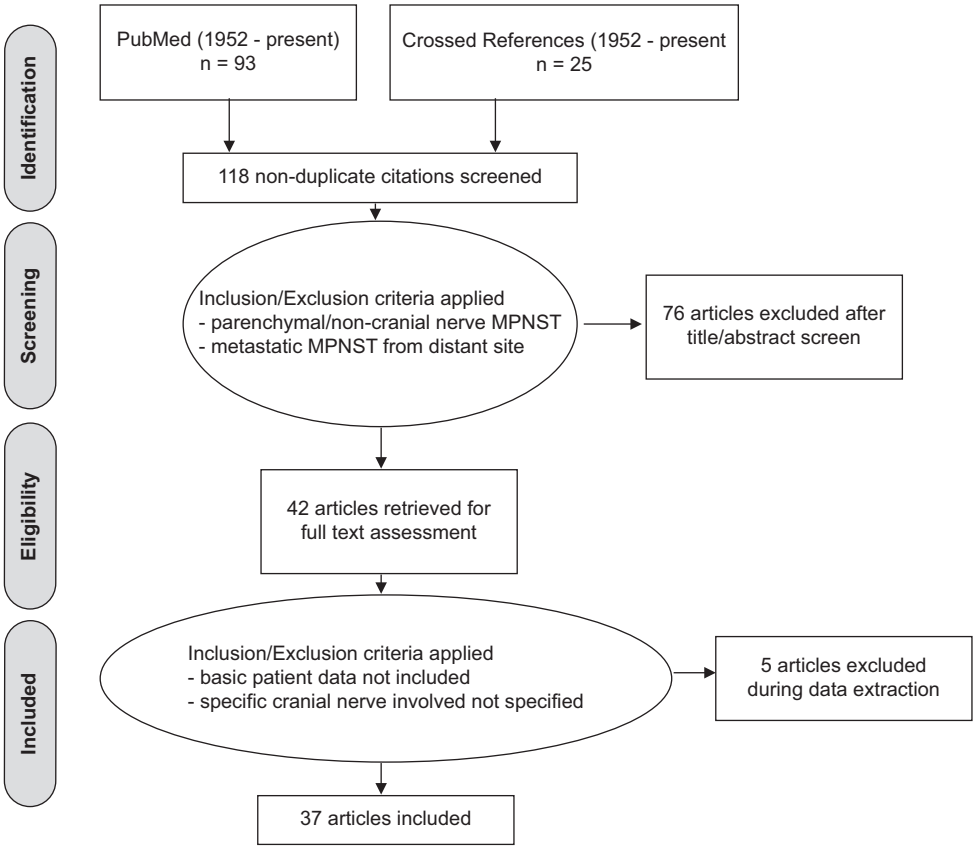

Figure 4: Preferred Reporting Items for Systematic Reviews and Meta-Analyses flow diagram. MPNST: Malignant peripheral nerve sheath tumor.

has been speculated to derive from the nevi vasorum, autonomic nerves deriving from the peripheral nervous system that lies within the adventitial layer of small and large pial arteries. Two-year survival remains poor, with a survival benefit in patients undergoing gross total versus subtotal tumor resection (53% vs. 22% at two years, respectively). A similar trend was noted in our review of extra-axial cases [Table 3]. Also described were 29 cases of MPNST originating outside the brain and spine with metastatic spread to the intracranial space. The prognosis for these patients, in particular, was especially poor, likely attributed to the advanced stage of disease, with 0% alive two years after surgery.

Histopathologically, the distinction between low-grade MPNST and atypical nerve sheath tumors is challenging. Standardized grading systems for MPNST are largely lacking. Many authors use the Scheithauer *et al*. morphological criteria for diagnosis of low-grade MPNST, which include hypercellularity, nuclear enlargement, and hyperchromasia,[31] all of which were present in our patient's tumor. A fascicular pattern of growth was also seen, which is usually lacking in conventional neurofibromas.[27] The tumor exhibited loss of p16 and patchy S100 staining, further suggesting MPNST in contrast to the diffuse S100 staining seen in cellular schwannoma. Additional low-grade MPNST features may include a collagenous interface between precursor lesion and MPNST, absence of p27, homozygous CDKN2A/B deletion, and EGFR amplification. Higher-grade lesions are categorized based on cytologic atypia, increased mitotic activity, and regions of necrosis.[27]

Although surgical resection remains the mainstay of treatment, gross total resection was achieved in only 35.3% of cases, likely attributed to both technical challenges with resection and attempts to limit significant morbidity. Even with gross total resection, 2-year survival remains poor at 33.3% (vs. 23.8% with subtotal resection). Particularly in tumors involving the 7th/8th nerve complex, a thorough discussion should be had with the patient regarding potential cosmetic and functional consequences of nerve injury/sacrifice. In deciding on a retrosigmoid approach, our initial goal was to obtain a tissue diagnosis and attempt to preserve the remaining hearing function, given the small size of the tumor. Once frozen pathology suggested a malignant process, we then opted for radical dissection, which ultimately required a trans labyrinth approach. During the initial surgery, the use of a 45° endoscope assisted with the

Table 1: Clinical characteristics and treatment of 53 patients with cranial-nerve associated MPNST.

Case	Author	Date	Age	Sex	Tumor location	Previous lesion or RT	Surgical approach	Adjuvant RT (±)	Adjuvant CT (±)	Outcome
1	Cueno and Rand[9]	1952	37	M	CN V, gasserian ganglion and mandibular n.	-	Temporal, GTR	-	-	Not reported
2	Hedeman *et al.*[14]	1978	53	M	CN V	-	Temporal, STR	+	-	Alive at last follow-up of 18 month.
3	Liwnicz[20]	1979	49	M	Bilateral CN V	-	Retromastoid, STR	-	-	Second operation for re-resection, mortality 9 month. after surgery
4	Levy *et al.*[19]	1983	61	M	CN V (middle fossa, cav sinus)	-	Subtemporal, STR	+	-	Functional after 10 month.
5	Levy *et al.*[19]	1983	67	F	CN V (middle fossa, parasellar)	-	Temporal, STR	-	-	Postoperative IPH; mortality after 1 month
6	Kudo *et al.*[17]	1983	54	M	CN VIII	-	N/A - autopsy case	-	-	N/A - autopsy case
7	Muhlbauer *et al.*[26]	1987	47	M	CN VII	-	Translabyrinth, GTR	+	+	Recurrence, suboccipital craniectomy at 16 month., metastasis to cervical, thoracic, lumbar spine, mortality at 2 year
8	Best[5]	1987	24	F	CN VIII	-	STR, not specified	-	-	Mortality after 4 month.
9	Horie *et al.*[16]	1990	18	M	CN V	-	N/A - autopsy case	-	-	Autopsy case
10	McLean *et al.*[24]	1990	75	M	CN VIII	Prior vestibular schwan., GTR	Suboccipital, GTR	-	-	Recurrence at 11 month., re-resection; mortality 2 month. After re-operation
11	Matsumoto *et al.*[23]	1990	54	M	CN VIII	-	STR, not specified	-	-	Recurrence, repeat resection ×2, mortality at 16 month.
12	Han *et al.*[12]	1992	47	F	CN VIII	-	Retromastoid, STR	+	-	Recurrence with metastasis to cervicothoracic spine, re-operation with mortality 11 month. After initial surgery
13	Maeda *et al.*[21]	1993	38	M	CN VIII	-	N/A - autopsy case	-	-	Autopsy case
14	Mrak *et al.*[25]	1994	40	M	CN VIII	-	STR, not specified	+	-	Re-resection for recurrence at 10 month., recurrence 11 month. After re-operation, metastasis to cervical/thoracic spine
15	Yamashiro *et al.*[41]	1994	46	M	CN V, cavernous sinus	-	Subtemporal, STR	+	-	Recurrence, re-resection ×2; alive after 5 year
16	Tegos *et al.*[36]	1997	66	M	CN V	-	GTR, not specified	+	-	Alive at 7-year follow-up
17	Higami *et al.*[15]	1998	45	F	CN III, IV, V, VII	NF2 history	N/A	N/A	N/A	N/A
18	Comey *et al.*[8]	1998	44	M	CN VIII	Acoustic schwannoma, prior GKRT	Translabyrinth, GTR	+	-	Recurrence, subsequent retro sigmoid approach for re-resection, mortality 1 year after initial surgery
19	Balasubramaniam[3]	1999	6 mo.	M	CN IX, X, XI	-	Retromastoid, STR	-	-	Hydrocephalus requiring EVD, respiratory failure with mortality at 20 days

(*Contd...*)

Table 1: (*Continued*).

Case	Author	Date	Age	Sex	Tumor location	Previous lesion or RT	Surgical approach	Adjuvant RT (±)	Adjuvant CT (±)	Outcome
20	Akimoto *et al.*[2]	2000	30	F	CN V (middle fossa, lateral cavernous sinus)	-	Frontotemporal, STR	+	-	Rapid recurrence; reoperation, mortality after 10 months
21	Saito *et al.*[29]	2000	69	M	CN VIII	-	STR, not specified	-	-	Recurrence, re-operation at 2 months, longer follow-up N/A
22	Hanabusa *et al.*[13]	2001	57	F	CN VIII	Acoustic schwannoma, prior GKRT	Combined retrosigmoid translabyrinth, STR	+	-	Mortality after 6.5 years
23	Stone *et al.*[35]	2001	71	M	CN V	-	Biopsy of extension in infraorbital n.	+	-	Mortality after 3 year
24	Bari *et al.*[4]	2002	28	F	CN VIII	NF2, prior radiosurgery	GTR, not specified	+	-	Mortality after 5 year
25	Wilkinson *et al.*[40]	2004	53	M	CN VIII	Acoustic schwannoma, prior to RT	Not specified	N/A	N/A	N/A
26	Ueda *et al.*[37]	2004	36	M	CN V, anterior skull base	NF1	Bifrontal craniotomy, STR	+	-	Spread to CPA, subsequent suboccipital craniotomy; mortality at 10 month.
27	Fisher and Dennis[10]	2006	41	M	CN V, VII - extension to IT fossa	-	Frontotemporal, STR	+	-	Recurrence at 38 month., mortality after 7 years
28	Maire *et al.*[22]	2006	63	F	CN VIII	Prior vestibular schwannoma, STR, adjuvant RT	Translabyrinth, GTR	+	-	No long-term follow-up was reported, and no significant surgical morbidity.
29	Stark *et al.*[34]	2006	59	M	CN V	-	Retrosigmoid, GTR	+	-	N/A
30	Gonzalez *et al.*[11]	2007	43	F	CN VIII	-	Retrosigmoid, GTR	+	-	Second staged operation for GTR, metastasis to thoracic and lumbar spine, mortality at 8 month.
31	Chen *et al.*[6]	2008	8	M	CN V	-	Middle fossa approach, STR	-	-	Recurrence in CPA, subsequent retro sigmoid approach for re-resection; mortality at 8 month.
32	Chen *et al.*[6]	2008	62	F	CN VII, VIII	-	GTR, not specified	-	-	Mortality after 4 month.
33	Chen *et al.*[6]	2008	3	M	CN V, cavernous sinus	-	STR, not specified	-	-	Mortality after 4 month.
34	Chen *et al.*[6]	2008	35	M	CN V, cavernous sinus	-	STR, not specified	-	-	Mortality after 2 month.

(*Contd...*)

Table 1: (*Continued*).

Case	Author	Date	Age	Sex	Tumor location	Previous lesion or RT	Surgical approach	Adjuvant RT (±)	Adjuvant CT (±)	Outcome
35	Chen *et al.*[6]	2008	46	F	CN V, cavernous sinus, extracranial extension	-	GTR, not specified	+	-	Survival >5 year
36	Chen *et al.*[6]	2008	5	M	CN V1, orbit	-	GTR, not specified	-	-	Mortality after 9 month.
37	Scheithauer *et al.*[30]	2009	32	M	CN VIII, IX, X, XI	NF1, chiasmatic glioma, RT	STR, not specified	+	-	Mortality 5 month. after surgery
38	Scheithauer *et al.*[30]	2009	67	M	CN VIII	Prior CN VIII schwannoma, STR	STR, subsequent GTR, not specified	-	-	Repeat surgery for cerebellar hemorrhage, mortality 1 month. after re-operation
39	Scheithauer *et al.*[30]	2009	50	M	CN VII	NF2, contralateral vest schwan 1 y prior	GTR, not specified	+	-	Recurrence at 12 month., mortality at 17 month.
40	Scheithauer *et al.*[30]	2009	56	M	CN VIII	Vestibular schwannoma, same site	STR, not specified	-	-	Recurrence, mortality at 2 month.
41	Scheithauer *et al.*[30]	2009	32	M	CN VIII, X	NF1	STR, not specified	-	-	Severe brainstem infarction, mortality after 3 month.
42	Scheithauer *et al.*[30]	2009	30	M	CN II	Hypothalamic pilocytic astrocytoma at age 7, adjuvant RT/chemo	STR, not specified	-	-	Mortality 1.5 month. after surgery
43	Scheithauer *et al.*[30]	2009	26	F	CN VII	Low-grade MPNST at 13 y, recurrence at 15 y surgically treated	STR, not specified	+	-	Hydrocephalus after 3rd surgery requiring ventricular drainage.
44	Scheithauer *et al.*[30]	2009	5	M	CN VIII	-	GTR, not specified	-	-	Stable CN VII/VIII dysfunction at last follow-up
45	Scheithauer *et al.*[30]	2009	32	M	CN X	NF1; hypothalamic pilocytic astrocytoma at age 5, adjuvant RT	Retrosigmoid, STR	+	-	Leptomeningeal spread
46	Voorhies *et al.*[38]	2013	30	F	CV VI	-	Retrosigmoid, STR	-	-	Alive a 1 y follow-up

(*Contd...*)

Table 1: (*Continued*).

Case	Author	Date	Age	Sex	Tumor location	Previous lesion or RT	Surgical approach	Adjuvant RT (±)	Adjuvant CT (±)	Outcome
47	Abou-Al- Shaar *et al.*[1]	2019	67	M	CN V	-	Combined temporal, infratemporal, GTR	+	-	Symptomatic improvement, no complications at 3 month. follow-up
48	Shutran *et al.*[32]	2019	53	F	CN V, cavernous sinus	-	Right frontal inter-hemispheric (metastasis), STR	+	-	Corpus callosum metastasis, recurrence with spread to pons, mortality at 16 month.
49	Yeole *et al.*[42]	2021	52	F	CN VIII	NF2; bilateral vestibular schwan., prior GKRT	STR, not specified	+	-	Stable tumor size, alive after 11 month. follow-up
50	Yeole *et al.*[42]	2021	39	F	CN V	Prior STR of trigeminal Schwan., GKRT	GTR, not specified	+	-	Alive without new deficits 8 month. postoperatively
51	Yeole *et al.*[42]	2021	18	M	CN V	-	Kawase approach, STR	+	-	Recurrence with extension to posterior fossa at 6 month., subsequent retromastoid craniotomy; alive at 8 month. follow-up
52	Lai *et al.*[18]	2023	53	F	CN VIII	-	Suboccipital, STR	-	-	Severe pre-operative posterior fossa IPH, mortality after several days
53	Our case	2023	40	F	CN VII, VIII	-	Retrosigmoid, STR; subsequent trans labyrinth, GTR	+	-	Translabyrinth approach after 1 month. For residual tumor, alive at 2 years

CN: Cranial nerve, STR: Subtotal resection, GTR: Gross total resection, RT: Radiation therapy, CT: Chemotherapy, IPH: Intraparenchymal hemorrhage, GKRT: Gamma Knife radiation therapy, NF: Neurofibromatosis, EVD: External ventricular drain, CPA: Cerebellopontine angle, MPNST: Malignant peripheral nerve sheath tumor, N/A: Not available

visualization of additional tumors within the IAC and may be useful in similar cases, although ultimately, a small 2 mm residual tumor remained within the vestibule.

Table 2: Summary of patient demographic data, risk factors, tumor location, and treatment.

Variables	MPNST cases
Age (year)	43.4±17.8
Number of patients (*n*)	53
Male (%)	35 (66.0)
Female (%)	18 (34.0)
Risk factors (%)	
NF mutation	8 (15.1)
Prior RT	11 (20.6)
CN Involved (%)	
II	1 (1.9)
III	1 (1.9)
IV	1 (1.9)
V	23 (43.4)
VI	1 (1.9)
VII	6 (11.3)
VIII	23 (43.4)
IX	2 (3.8)
X	4 (7.5)
XI	2 (3.8)
Treatment (%)	
STR	14 (27.4)
STR+RT	15 (29.4)
GTR	6 (11.8)
GTR+RT	12 (23.5)
Biopsy only	1 (2.0)
Autopsy case	3 (5.9)

MPNST: Malignant peripheral nerve sheath tumor, GTR: Gross total resection, STR: Subtotal resection, RT: Radiation therapy, NF: Neurofibromatosis, CN: Cranial nerve

MPNST may be indistinguishable from benign nerve sheath tumors radiographically. Tumor growth seen on short-interval surveillance imaging necessitates timely surgical evaluation and tissue diagnosis. Despite the sacrifice of the proximal 7th nerve, our patient regained significant facial function following nerve transfer. In such cases, a multidisciplinary approach should be utilized to achieve optimal patient outcomes. Two years following surgery, our patient remains without signs of disease recurrence; however, strict surveillance remains essential. Our practice is to surveillance images every three months in the first two years, then every six months after that.

CONCLUSION

MPNSTs comprise a group of highly aggressive neoplasms typically arising from peripheral nerve cells. While their intracranial involvement is rare, prompt diagnosis and treatment are essential. Gross total resection should be attempted, and a multidisciplinary approach should be utilized in similar cases where facial nerve function is compromised. Further, research on intracranial MPNST will continue to improve treatment and patient outcomes.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

Patients' consent not required as patients' identities were not disclosed or compromised.

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Nil.

Table 3: Comparison of patient factors, treatment, and survival between primary extra-axial, primary intraparenchymal, and secondary MPNST. Adopted from Rubino *et al*.

Variable	Primary extra-axial MPNST	Primary intraparenchymal MPNST	Metastatic/secondary MPNST
Age (years)	43.4±17.4	31.2±23	41.8±22.4
Number of patients (*n*)	53	26	29
Male (%)	35 (66)	13 (50)	19 (65.5)
Female (%)	18 (34)	13 (50)	10 (34.5)
Risk factors			
NF mutation (%)	8 (15.1)	6 (23)	8 (27.5)
Prior radiotherapy (%)	11 (20.6)	0 (0)	3 (10.5)
Treatment			
GTR (%)	18 (35.3)	16 (61.5)	7 (24.1)
STR or biopsy (%)	30 (58.8)	10 (38.5)	5 (17.3)
2-year survival			
GTR (%)	4/12 (33.3)	53	0 (0)
STR or biopsy (%)	5/21 (23.8)	22	0 (0)

GTR: Gross total resection, STR: Subtotal resection, NF: Neurofibromatosis, MPNST: Malignant peripheral nerve sheath tumor

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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