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Case Report

Intracranial Malignant Nerve Sheath Tumor in the Middle Cranial Fossa: A Rare Case Report with Review of Literature

Abstract

Intracranial malignant nerve sheath tumors rarely arise in the central nervous system. They usually arise from the cranial nerves, though rare cases of intraparenchymal lesions have also been reported. We report a case of malignant nerve sheath tumor located in the right middle cranial fossa. Preoperatively, the lesion resembled a meningioma arising from the petrous temporal bone. The lesion was completely excised. Postoperatively, the patient developed righted-sided complete facial nerve palsy. Histopathology and immunohistochemistry revealed the lesion to be a malignant nerve sheath tumor. The development of postoperative facial nerve palsy was puzzling but could be explained if we consider the possibility of the lesion arising from the facial nerve near the geniculate ganglion. Intracranial malignant nerve sheath tumor centered over the geniculate ganglion and projecting into the middle cranial fossa is uncommon, and to the best of our knowledge, only one such case has been previously reported. We review the relevant literature, discuss the management and add to the previously reported cases of this rare condition.

Keywords: *Malignant peripheral nerve sheath tumor, malignant intracerebral nerve sheath tumor, malignant neurofibroma, malignant peripheral nerve sheath tumor, malignant Schwannoma*

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Introduction

Intracranial malignant nerve sheath tumors are rare tumors with very few isolated case reports and small case series.

We report a case of an intracranial malignant nerve sheath tumor, mimicking a meningioma, located in the middle cranial fossa, and probably originating from the geniculate ganglion.

Case Presentation

A twenty year old male patient presented with headache and vomiting since 3–4 months. The patient was fully conscious with fundus examination showing papilledema. Cranial nerves were normal on testing. Magnetic resonance imaging (MRI) revealed a large extra-axial tumor in the middle cranial fossa which was hypointense on T1, hyperintense on T2 with contrast enhancement [Figures 1-5]. The base of the tumor was on the petrous temporal bone. A diagnosis of meningioma was made preoperatively, and the patient was taken up for surgery.

Operative technique

The patient was operated in supine position under general anesthesia. The right side temporal craniotomy was made. The tumor was firm grayish red in color. The tumor was completely removed by standard microsurgical technique of internal debulking of the tumor followed by capsular dissection. There was bleeding at the base at the petrous temporal bone which was cauterized. After securing hemostasis and dural closure, drain was kept in the subgaleal space, and the incision was closed in a standard fashion.

Postoperative course

In the immediate postoperative period, total right-sided facial palsy was noted. The patient was observed in the intensive care unit for 2 days. The patient remained fully conscious and was discharged on the 7th postoperative day. Histopathological examination revealed biomorphic tumor composed of sheets and cords of epithelioid cells and bundles of spindle cells [Figure 6]. The epithelioid component showed features of nuclear anaplasia along with frequent mitosis [Figure 7].

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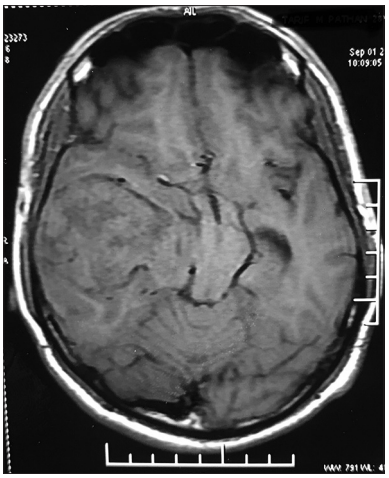

Figure 1: Preoperative T1-weighted image showing isointense lesion in the right middle cranial fossa

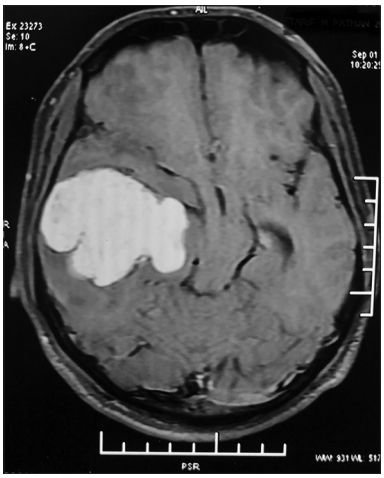

Figure 2: Preoperative T1 contrast axial image showing the brilliantly enhancing lesion in the right middle cranial fossa

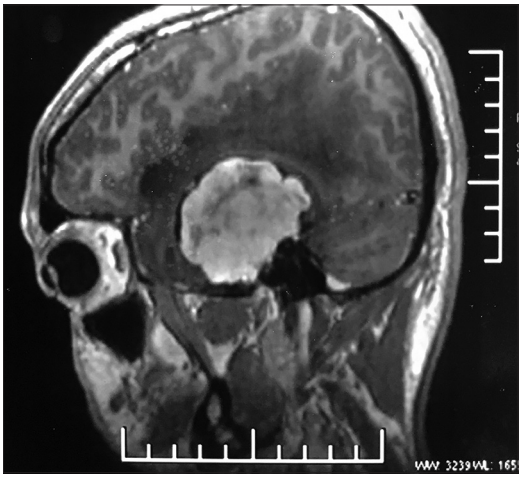

Figure 3: Preoperative T1 contrast sagittal image showing the tumor arising from the base of the middle cranial fossa and the petrous bone

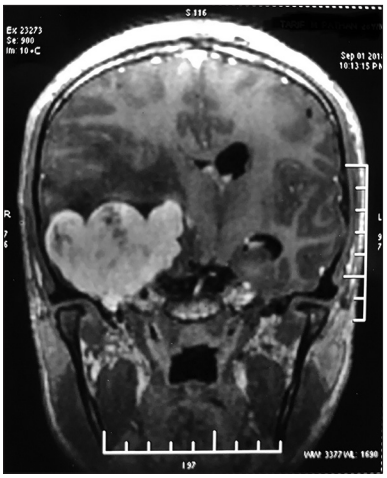

Figure 4: Preoperative T1 contrast coronal image showing the tumor

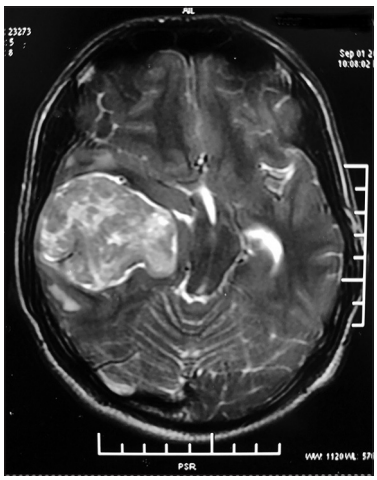

Figure 5: Preoperative T2 axial image showing the hyperintense tumor in the right middle cranial fossa

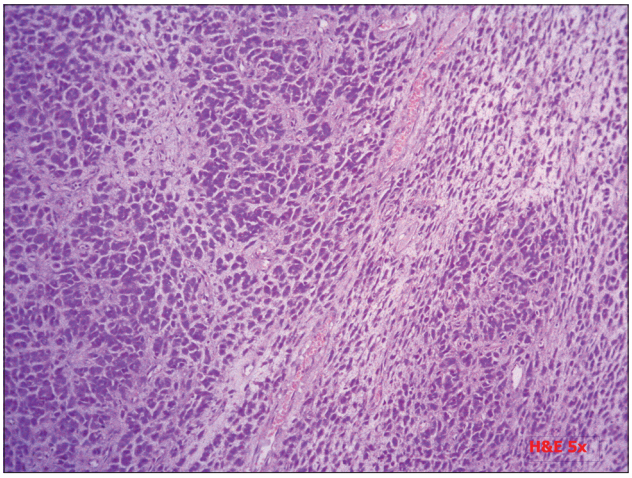

Figure 6: Scanner view of the pathology specimen shows biomorphic tumor composed of sheets and cords of epithelioid cells and bundles of spindle cells

Immunohistochemistry (IHC) revealed immunopositivity for vimentin, confirming the mesenchymal differentiation of the tumor [Figure 8]. The tumor cells showed patchy nuclear and cytoplasmic immunostaining for S-100, confirming the neural differentiation [Figures 9 and 10].

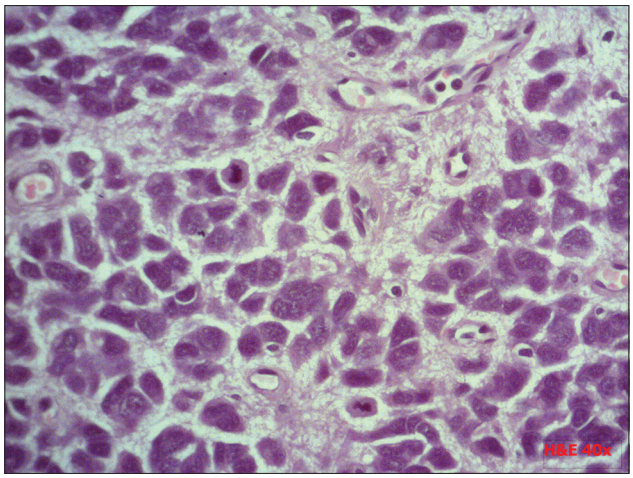

Figure 7: High power view of the epithelioid component shows features of nuclear anaplasia along with frequent mitosis

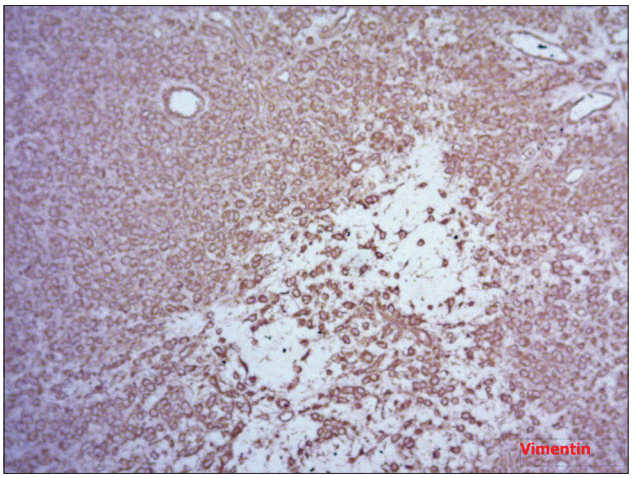

Figure 8: Scanner view section shows tumor cells are diffusely immunopositive for vimentin, confirming the mesenchymal differentiation of the tumor

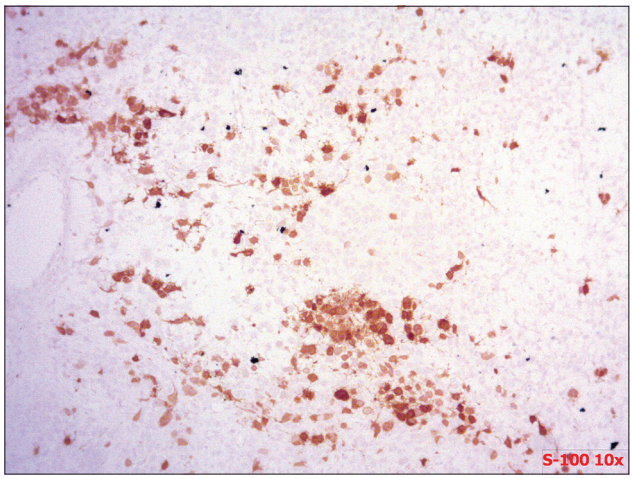

Figure 9: ×10 view showing tumor cells show patchy nuclear and cytoplasmic immunostaining for S-100, confirming the neural differentiation

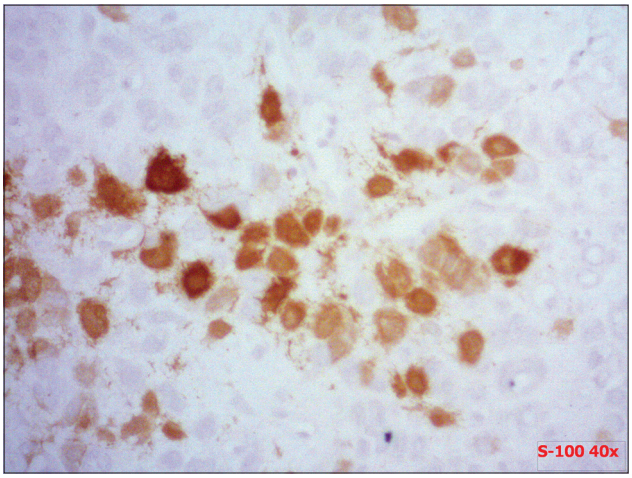

Figure 10: ×40 view showing tumor cells show patchy nuclear and cytoplasmic immunostaining for S-100, confirming the neural differentiation

Glial fibrillary acidic protein, smooth muscle actin, desmin, HMB-45, epithelial membrane antigen, and cytokeratin were negative.

MRI 1 month after the surgery confirmed the total excision of the lesion [Figure 11]. The patient was referred for radiotherapy. At 6 months, there was an improvement in the right-sided facial palsy [Video 1].

Discussion

Malignant peripheral nerve sheath tumors (MPNSTs) are very rare tumors with an incidence of 0.001%.[1]

Their intracranial counterparts are even more sporadic.

They can be classified into two types:

1. Extra-axial lesions: Intimately associated with the cranial nerves and more commonly originating in the posterior fossa. The site of origin is from the eighth cranial nerve in (60%), fifth nerve in (27%), seventh nerve in (10%), and from others in remaining cases[2]
2. Intraparenchymal lesions: Also called malignant intracerebral nerve sheath tumors (MINST), not associated with any cranial nerve and more common in the supratentorial region[3-5]

Our literature search for MPNST revealed two case series reviews. Ziadi *et al*. reported 32 cases of intracranial MPNST in 2010 while Lebeau *et al*. could find 60 reported cases in 2013. These reviews did not include MPNSTs arising within the brain parenchyma (MINST). MINST is more uncommon and literature search revealing review of 15 reported cases by Shweikh *et al*. 2013 and a review of 25 reported cases till 2016 by Fevre *et al*.

Malignant nerve sheath tumors can arise sporadically (47%) or from malignant transformation of benign lesions either Schwannoma (40%) or neurofibroma (8%).[2] They have also been reported in patients who have received radiation to the head and neck region for some other condition. Some association with NF1 and NF2 has been reported.

Surgery followed by radiotherapy has been the mainstay of treatment. Radical resection may not be possible in all cases because of its close association with critical brain structures. These tumors carry a poor prognosis with a 1-year survival rate of 33%.[2] Drop metastasis to spine has been reported in 22% of the cases.[1] Hence, spinal MRI should be done during the follow-up of these patients.

In our patient, the preoperative diagnosis was that of a middle fossa meningioma arising from the petrous bone. It was the histopathology and IHC report, showing immunopositivity for vimentin and S-100, which revealed it to be an MPNST.

Clinically and radiologically, it is difficult to point to any association with the facial or any other cranial nerve. Only the development of immediate postoperative facial nerve palsy points toward a facial nerve involvement. It is very well possible that the origin of the tumor might be from the facial nerve near the geniculate ganglion. In order to lend credence to this hypothesis, postoperative computed tomography (CT) scan of the temporal bone was done [Figures 12 and 13] which showed mild erosion of tegmental wall of middle ear cavity (epitympanum) along with focal dehiscent wall of temporal bone near geniculate ganglion, supporting the hypothesis of origin of the tumor from the geniculate ganglion (the petrous was not drilled during the surgery. Preoperative CT scan was not done as the lesion appeared to be a meningioma and because of financial constraints). This explains the postoperative facial nerve palsy which probably occurred due to traction injury to the nerve or due to heat of the diathermy during cauterization from the tumor base, from where there was troublesome bleeding intraoperatively.

Another possibility is that the tumor might have been adherent to the greater superficial petrosal nerve and traction over the tumor during surgery may have led to the facial palsy.

We could find one such reported case in the literature where the tumor was arising from the facial nerve and was centered over the geniculate ganglion.[5] Postoperative radiotherapy is recommended in MPNSTs to delay recurrence and improve survival. However, the prognosis for this condition remains poor.

Conclusion

Intracranial malignant nerve sheath tumors are rare without any preoperative distinguishing clinical and radiological features. They may usually occur as extra-axial lesions and rarely within the brain parenchyma. In our case, it is difficult to exactly determine the site of origin of the lesion, but the postoperative facial nerve palsy points to the origin from the facial nerve.

Our case adds to the previously reported cases of this rare condition.

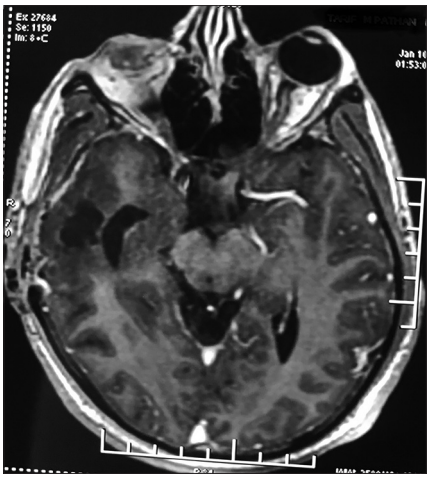

Figure 11: Postoperative image T1 contrast axial image confirming total tumor removal

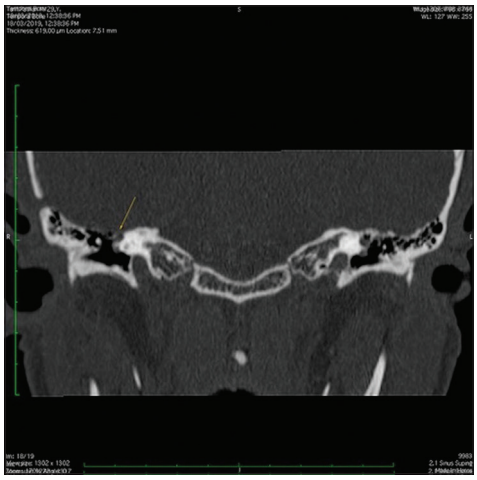

Figure 12: Postoperative computed tomography scan of temporal bone showing erosion of the epitympanum of the right petrous bone (arrow)

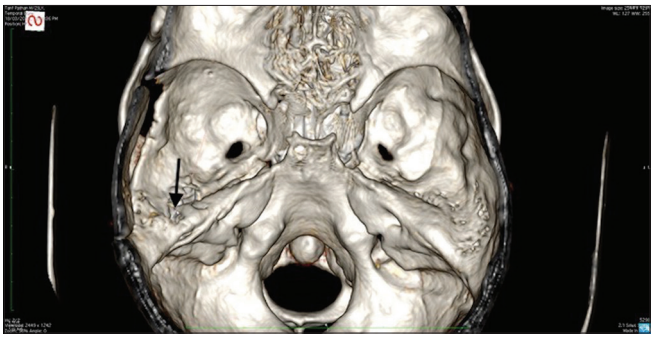

Figure 13: Three-dimensional computed tomography reconstruction of showing the erosion of the petrous bone (black arrow)

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The

patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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