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Article

Intra-cranial malignant peripheral nerve sheath tumor of olfactory nerve: a case report and review of literature

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Intra-cranial malignant peripheral nerve sheath tumor of olfactory nerve: a case report and review of literature

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Abstract: Malignant Peripheral Nerve Sheath Tumors (MPNSTs) are one of the very rare high grade malignancies usually affecting extremities or trunk. Incidence is 1/Lac. Intra-cranial MPNSTs are even rarer, schwannomatous and commonly affecting cranial nerves VIII &VII). Intra-cranial MPNSTs are usually sporadic, arising de novo. The second most common mode of origin is from malignant transformation from pre-existing schwannomas or neurofibroma. We present an extremely rare and probably the first case of intra-cranial malignant peripheral nerve sheath tumor of the olfactory nerve in a non neurofibrosis patient with no prior history of irradiation.

Key words: Intra-cranial malignant peripheral nerve sheath tumor, Olfactory Nerve Tumor, MPNST

Introduction

Malignant peripheral nerve sheath tumor MPNSTs usually arise from peripheral nerves or shows differentiation towards nerve sheath cells. [9] MPNST of cranial nerves is very rare. The incidence of MPNST in the general population is 0.001% [8]. They constitute 5-10 % of all soft tissue sarcomas and 50-70% are associated with Neurofibromatosis type 1 (NF-1).[3,10] MPNSTs involving the Head and neck region comprise only 04-08 % of all MPNSTs.[1] MPNST arising from olfactory nerve without any features of neurofibromatosis type 1 or type 2 as has been noted in our case. In the background of extensive literature search and with the best of our knowledge, probably this is the first case report of MPNST arising from olfactory nerve.

Case history

A 45 years old male presented to us with history of headache and bilateral loss of smell since six months and altered sensorium since seven days. On examination he had frontal lobe signs and features of foster Kennedy syndrome.

His Pre-operative MRI, T1 and T2 weighted images had 8x7x5 cm heterogeneous lesion originating from the anterior cranial fossa. On CEMRI (Figure 1) lesion was heterogeneously enhancing. His metastatic work up was negative. Patient was planned for

surgery with diagnosis of atypical olfactory groove meningioma.

He underwent bifrontal craniotomy and gross total excision of the tumor (Figure 2).

Histopathology was suggestive of malignant peripheral nerve sheath tumor of the olfactory nerve. Post operatively patient was discharged on 7th post op day with no fresh deficit. Patient underwent post op radiotherapy and is recurrence free since last six months.

Discussion

MPNSTs of the cranial nerves are rare tumors, and the literature is limited to isolated case reports and small case series. [9] In general population, the incidence of MPNST is 1/Lac population per year. [1] MPNSTs generally occur typically between ages 20-50 years whereas 10-20% of cases have been reported in the first 2 decades.[4,8] The common sites of origin include the extremities and trunk, usually sciatic nerve, brachial plexus and the sacral plexus MPNSTs of head and neck region comprise 04-08% of all MPNSTs and especially the cranial nerves involvement is extremely rare with only few cases or short case series found in the literature.[9] Most common intracranial MPNSTs are found in VIII & V cranial nerves.[7] In retrospective analysis of intra cranial MPNSTs by Le'Hereux and Saliba which included 31 case reports and short case series comprising 60 cases, the incidence of cranial nerve MNSTs were: VIII CN-60%; V CN-27%, VII CN-10%; III CN-6.7 %; IV CN-1.7% &VI CN-1.7 %.[7] Etiologically, MPNST may occur infrequently when they arise de-novo, occur in association with neurofibromatosis (NF) [5,7], may be consequent to radiation (conventional or SRS) or arise as malignant transformation of schwannoma or neurofibroma.[7] MPNSTs are highly malignant tumors that grow rapidly along nerves, infiltrate surrounding tissues and develop hematogenous metastasis. MPNSTs may develop hematogenous distant metastases (most commonly to Lungs; other sites being Bones and Liver.[5]

MRI study does not allow confident distinction between benign and malignant nerve sheath tumors. A size more than 5 cm, infiltrative margins and marked signal heterogeneity, adjacent structure compression and local invasion are features which favor MPNSTs. The diagnosis of MPNSTs necessitates biopsy and remains controversial due to its lack of specific morphological criteria and immunohistochemical or molecular tests.[6]

Treatment of MPST is primarily surgical. Gross total excision of intracranial MPNSTs significantly influences overall survival rates and relates inversely with local recurrence. Complete surgical tumor resection with preservation of neurological function should be the goal of the treatment. However, critical location and stubborn nature of tumor, complete surgical resection can be challenging and is not always feasible.[1,7] The role of adjuvant Radiotherapy remains controversial.[7] Numerous chemotherapy regimens have been used against MPNSTs but results have been unconvincing even when combined with surgery and radiotherapy.[3,7] In early stages of MNST Surgical resection of

the tumor provides survival benefits. Whereas radiotherapy is useful in improving survival in metastatic MPNST. Combined therapy with surgery and radiotherapy are prognostically important in patient with tumors >5 cm. [2]

Conclusion

Intra-cranial olfactory MPNST is extremely rare. This is the probably first such reported case. Complete excision should be the goal of surgery which is the mainstay of treatment. However, due to the stubborn nature of tumor and intimate relation with critical neurovascular structures, this may not always be feasible. Combined therapy surgery with post op radiotherapy has some survival benefits in metastatic MPNST.

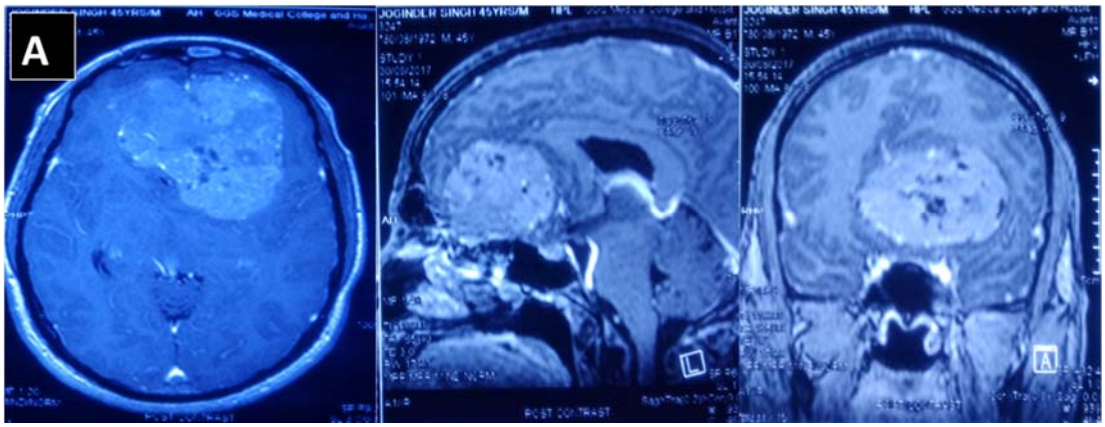

Figure 1 - Pre Op contrast enhance MRI brain (axial, sagittal and coronal view) Showing anterior cranial fossa SOL

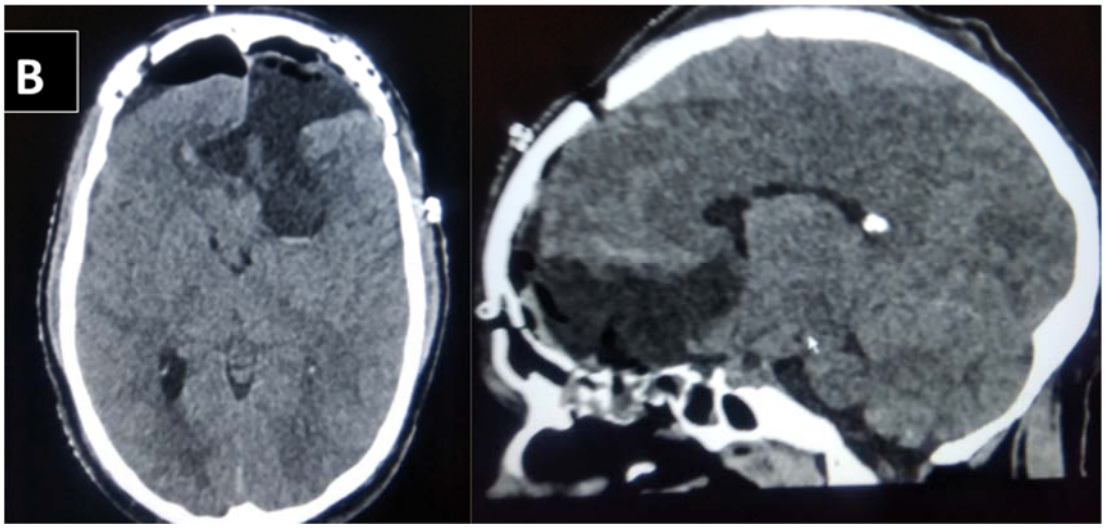

Figure 2 - Post op NCCT Head axial and (sagittal view) showing complete excision of the tumor

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