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MALIGNANT PERIPHERAL NERVE SHEATH TUMOR OF TONGUE- CASE REPORT

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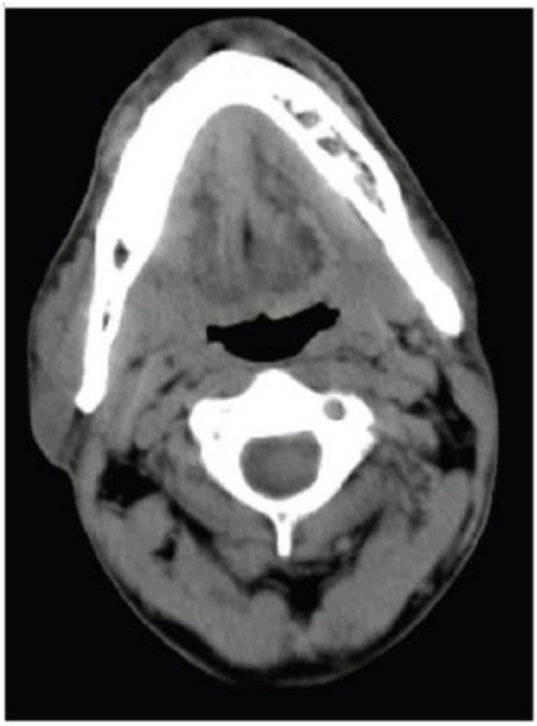
INTRODUCTION

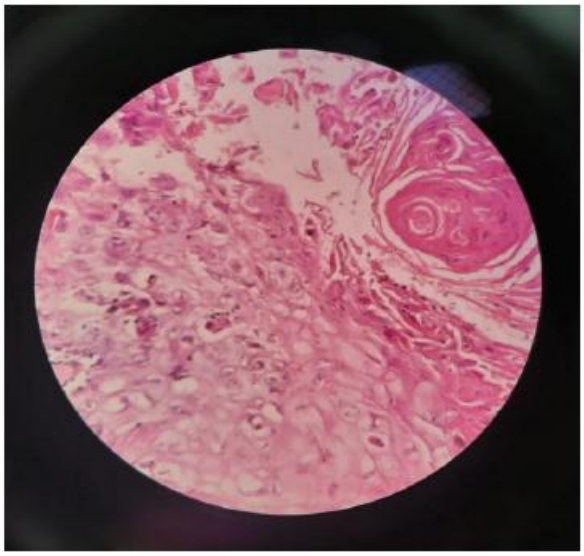
Malignant peripheral nerve sheath tumors (MPNST) are a rare variety of tumors which constitutes approximately 5-10% of soft tissue sarcomas. It accounts for around 6-16% of head and neck sarcomas. It has been observed that almost around 50% of MPNSTs are associated with NF1 and have a tendency to arise from pre-existing neurofibromas. Median age of presentation of these tumors is around 30-50years of age. There is no sex prediliction for this disease. Patients usually present with a slow growing swelling which may be associated with pain. These tumors usually tend to have a poor prognosis despite the use of multimodality treatment.

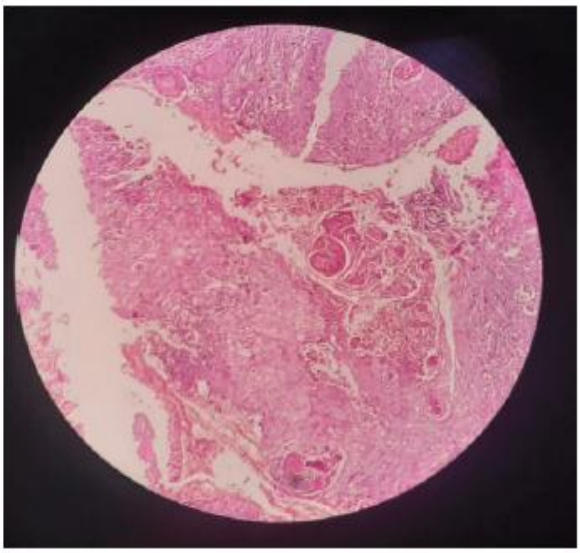
CASE REPORT

A 44 years old male patient, non hypertensive, non diabetic, known smoker and alcoholic for 10 years, painter by occupation noticed whitish and red patches over left side of tongue for which he consulted an oncologist and was diagnosed to have oral leukoplakia for which he was managed conservatively after which he remained asymptomatic for around 4 years, when he noticed redness over left side of tongue at the same site of leukoplakia which then progressed to form an ulcerative lesion which was gradually progressive in nature and was asssociated with localised pain around the lesion. No cafe au lait spots were seen on the body of the patient. Also,the patient had no family of NF1.CT scan of neck was done which showed heterogenous enhancing ill defined soft tissue mass lesion of 10*28*17 mm in left anterior 2/3rd of tongue with poor fat planes infiltrating into adjacentmusculature.



He underwent biopsy from the lesion which turned out to be malignant mesenchymal tumor .After this the patient underwent wide local excision of tongue with modified neck dissection. The surgical pathology report showed areas of sarcomatoid de-differentiation with highly atypical pleomorphic spindle cells and was staged as pT3N0.IHC was done which turned out to be positive for CK, Vimentin, S-100 and focally positive for SMA, CD57 and Ki67 was 60%.The patient was diagnosed as a case of malignant nerve sheath tumor of tongue, after which the patient presented to our department for further treatment . Proper metastatic workup was done and no evidence of metastasis was found.





The patient was started on EBRT to tongue using Cobalt-60 to a total dose of 60Gy in 30 fractions ,2Gy per fraction given 5 days a week along with concurrent CTRT with Inj cisplatin 40mg/m2 weekly. Further, the patient is planned to beta kenonadriamycin and if osfamide based adjuvant chemotherapy after the completion of radiotherapy.

DISCUSSION

Malignant peripheral nerve sheath tumours are rare, highly aggressive tumors. They can arise de novo or from pre-existing benign schwannomas or neurofibromas.[1,2] The incidence of tumours is about 4–10% of all soft tissue sarcomas. Most of these occur in the extremities and trunk, but the head and neck are involved in only about 8—14% of the cases. In a large series from the Mayo clinic, the calculated incidence of these tumours is 0.001% in the general patient population and 4.6% among patients with NF1.[3]

The mainstay of treatment of these tumors is mainly wide local excision. The involved nerve should also be resected proximal to the tumor site.[4,5] Prophylactic neck dissection is not advised as incidence of lymphatic invasion is very less. Post op radiotherapy is recommended to prevent local recurrences. Studies have shown improved disease free survival with the administration of adjuvant chemotherapy.

The prognosis for MPNST of the head and neck is poor. The survival rates are reported to be between 15 and 65%. Survival in patients with NF I is significantly lower (15% 5-year survival).Metastasis in MPNST occurs via a haematogenous route and about one-third of patients have pulmonary metastases at presentation.[6] High Ki-67 labelling index is associated with poor survival.

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