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MALIGNANT PERIPHERAL NERVE SHEATH TUMOUR OF BUCCAL MUCOSA WITH ACUTE MYELOID LEUKEMIA:A CASE REPORT

Oncology

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ABSTRACT

The incidence of MPNST in buccal mucosa is rare, so is the association with acute myeloid leukemia. A 40 year old lady presented with lesion in buccal mucosa. Post-operative histopathology was suggestive of MPNST. During post-operative period, she was diagnosed with acute myeloid leukemia and is under treatment for the same.

KEYWORDS

MPNST, buccal mucosa MPNST, MPNST and leukemia

Introduction:
Malignant peripheral nerve sheath tumours are sarcomas arising from the peripheral nerve sheaths or associated cells. These most commonly arise from plexiform neurofibroma. 50% patients with NF1 develop plexiform neurofibroma of which 3-10% undergo malignant transformation. Also cases associated with NF1 have been reported to be associated with glioma,optic nerve glioma and leukaemia. Here we report one case of MPNST of buccal mucosa with leukaemia without clinical features corroborative of Nf1.

History:
A 40 years old lady presented with a gradually increasing swelling over right lower alveolus for 2-3 months. Clinically the lesion was extending to the floor of the mouth with doubtful involvement of the ipsilateral submandibular salivary gland and mandible.

CT scan showed a 3.5x6.2x2.5 cm lesion in right inferior gingivobuccal sulcus lesion abutting right submandibular salivary gland and mild erosion of mandible.

Punch biopsy from the lesion was suggestive of poorly differentiated malignant tumour. IHC was positive for Vimentin and LCA and weakly positive for S100. Chest X-ray and biochemical investigations were normal at the time of diagnosis.

Wide local excision was done. Perineural invasion and intense lymphostromal response was noted. Also submitted sections showed presence of submucosal tomour,mucosa was free. Post-op RT was planned conventionally with AP-Lateral fields to a dose of 60Gy in 30 fractions at 2Gy per fraction 5 days a week over 6 weeks.

After 40Gy 20 fractions she developed high grade fever (101-103 degrees Fahrenheit). Chest X-ray, urine analysis, viral markers for dengue,widal and malarial parasites were normal. Haemogram showed anemia with thrombocytopenia with 92% blast cells. This prompted bone marrow biopsy, which demonstrated presence of 85% blasts, RBCs, WBCs and platelets being adequate. IHC was suggestive of AML-M1.

During all these, RT was stopped at 27 fractions and Amphotericin B was started.

Discussion:

MPNSTs account for 5% of sarcomas. Two-third arise from pre-existing plexiform neurofibroma and rest arise denovo. Rarely, these may develop from malignant transformation of pre-existing schwannoma, ganglioneuroma or pheochromocytoma. The term MPNST has replaced previously existing terms like neurogenic sarcoma, malignant fibrosarcoma and malignant neurifibrilemmoma. Also, 50% is found in NF1; other malignancies like neurofibromas are seen. These patients may develop café au lait spots, freckles in unlikely places, Lisch nodules, and neurofibromas. Our patient did not show any of those features.

These are high grade sarcomas. The incidence is maximum in the extremities (40%), followed by retroperitoneum (38%). Head and neck occurrence is about 21%. Other sites in head and neck include neck, mandible and maxilla.

No gender predilection has been noted. Age range is wide with any age between 20-50 years being reported.

Reccurrence rate is high at approximately 40-80%. Hematogenous dissemination is common with lung being the most common site to be affected, followed by soft tissue and bone.

Microscopically,these cells sometimes show schwann call differenti ation, while sometimes they suggest clear cell peripheral nerve cell origin. These are highly cellular tumours that exhibit features of overt malignancy like anaplasia, necrosis, infiltrative growth pattern, polymorphism and high proliferative activity. Low power view often show areas of alternating high and low cellularity, thus the resultant “marble-like” name.

IHC plays an important role in diagnosis. Complete loss of SOX 10, p16, Neurofibromin has been noted. 80% MPNSTs and 31% schwannomas show loss of p75NTR. Ki67 labelling indices have shown 87% sensitivity and 96% specificity in MPNST. S100 is seen in 50-90% cases and is indicative of schwann cell differentiation. Collagen IV positivity, NFGR positivity, S100 positivity are suggestive of perineural differentiation. If CD34 is positive, maybe associated with perivascular fibroblast like cells. Some cases may show rhabdomyoblastic and cartilaginous differentiation. In our present case, Desmin, Actin, CD20, CD34, AE1 were found to be negative.

MRI is the imaging modality of choice. The lesions are generally oriented along the direction of the nerve of origin. Large lesions, especially those more than 5cm invade fat plane, are heterogenous, have ill-defined margins and perilesional oedema- all these features turn the diagnosis in favour of MPNST.

The role of FDG-PET in differential diagnosis of MPNST from its benign counterpart is unclear. However, recently, prognostic significance has been suggested.

Biopsy is integral for staging and grading and thus halps planning adjuvant treatment. FNAC is not often used in initial diagnosis as the minimum amount of sample aquired makes it difficult,if not impossible, to distinguish the architectural pattern. But FNA is often successfully used for recurrent disease.

Surgery forms the mainstay of treatment with the aims to achieve total resection with wide margins. This patient also was treated on the same line, followed by adjuvant radiotherapy.

Adjuvant RT has been found to increase both local control and overall survival. Pre-operative RT may be beneficial with accurate RT planning and tumour localisation, small treatment volumes, small doses required. Also it offers the advantage of oxygen enhancement effect. Tumour necrosis potentially decrease the chances of intra-operative tumour spillage. However, delay in surgery, delayed wound healing and less tissue for diagnosis are drawbacks. Post-operative RT, on the other hand, can avoid all the complications. But larger volume has to be treated and to a higher dose. Also the risk of spillage and seeding during surgery remains somewhat higher. Peri-operative brachytherapy may be a viable option, but it also poses problem in wound healing.

Chemotherapy,when used pre-operatively, may be used to decrease the tumour bulk and increase resectibility. Post-operative chemotherapy is to be tailored according to the patient to be treated.

Like elsewhere in the body, T-stage is the single most important prognostic factor. Ki67 is also an important prognostic index. Previously it was noted that NF1 positivity has worse prognosis than the sporadic variant. But recent reports fail to note any such correlation. 5-year survival rate is around 34-39%. Tumor size < 5 cm, lack of local recurrence, low histologic grade and extremity location are good prognostic factors. Low intensity p53 staining and positive S-100 staining is also found be associated with better outcomes.

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