P11 ADULT T CELL LYMPHOMA (ATL) MASQUERADING AS FACIAL NERVE PARALYSIS

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Purpose of Study ATL is a rare and aggressive peripheral T-cell neoplasm characterized by clonal human T-cell lymphotropic virus type-1 (HTLV-1) proviral DNA integration with host T lymphocytes. These patients commonly present with lymphadenopathy, skin rash, fever, fatigue or altered mental status. The prevalence of CNS disease varies from 3 to 50% and is always in the presence of systemic disease. Isolated cranial neuropathy as a presenting symptom has not been described in literature.

Methods Used Retrospective chart review and review of literature.

Summary of Results 49 year old Caribbean male presented with 2 month history of left sided headache, 5 weeks of right sided jaw numbness and pain which progressed to contralateral side. He was now unable to smile and had food falling from the side of his mouth. He denied fever, fatigue, night sweats, rash, weakness or abnormal lumps. He had normal mental status and good motor strength. Facial exam reveal bilateral upper and lower facial paralysis, left lateral rectus palsy and horizontal gaze diplopia. Rest of the physical exam was unremarkable. Labs revealed WBC of 6 k/mm³ with normal differential, HB 16.5 gm/dl and platelets 238 k/mm³. Complete metabolic profile and peripheral smear was normal. MRI Brain showed irregular, fusiform enhancement of left trigeminal nerve, bilateral facial and abducent nerves. CSF flow cytometry showed clonal CD4+

CD25+ T cell population. HTLV-1 serology was reactive. Left infraorbital nerve biopsy confirmed involvement with ATL. CT Chest/abdomen/pelvis did not reveal enlarged lymphadenopathy. He was started on treatment with EPOCH and twice weekly intrathecal methotrexate for 4 months with clearance of CNS fluid. His jaw pain and vision improved but facial nerve paralysis persisted. He developed local relapse four months after treatment and was treated with high dose methotrexate for 5 cycles. Ultimately his performance status deteriorated and he succumbed to the disease progression.

Conclusions This case illustrates the unique presentation of this disease and gives an insight on one treatment approach. This patient achieved remission with our approach of aggressive chemotherapy with intrathecal methotrexate although the duration of remission was short lived.