SOHO ANNUAL MEETING 2024

Neurolymphomatosis: An Unusual Manifestation Of Relapsed/Refractory Lymphoma

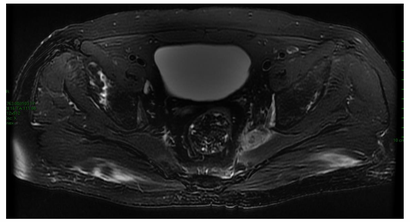
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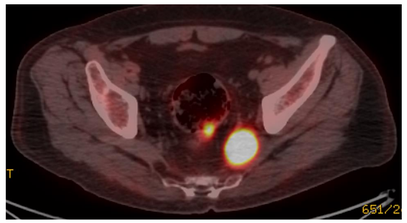
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INTRODUCTION

Secondary neurolymphomatosis is an extremely rare neurologic manifestation of non-Hodgkin lymphoma (NHL) and leukemia, representing a unique subtype of extra-nodal involvement. It refers specifically to lymphoid infiltration of the peripheral and cranial nerves, roots, plexuses and innervated structures that develops as progression or relapse of a previously diagnosed lymphoid malignancy.



MRI of the Pelvis showing enhancing sacral perineural soft tissue process, anterior to the left piriformis muscle, representing a secondary deposit of neurolymphomatosis



PET CT Scan FDG showing mass like well defined soft tissue density showing increased FDG avidity noted anterior to the left piriformis muscle

CASE PRESENTATION

A 58-year-old gentleman presented to the emergency department in distress, history of fatigue, and 10 kg weight loss in three weeks with drenching night sweats. On physical exam he had cervical and axillary lymphadenopathy without hepato-splenomegaly, and crackles on left lower lung. Laboratory results were significant for elevated levels of uric acid and LDH. A lymphoproliferative disorder was suspected, and an excisional biopsy of a prominent cervical lymph node revealed diffuse large B-cell lymphoma, germinal center type. CT findings showed disease staged as IIIB, possibly IV given the suspected lung involvement that could not be adequately diagnosed due to the patient's respiratory status. The patient initially received one cycle of dose-adjusted R-EPOCH, which was switched to R-CHOP with three cycles of intrathecal methotrexate once the FISH result revealed an isolated rearrangement of BCL2 gene. A follow-up PET/CT after four cycles of R-CHOP revealed complete remission, after which he received two more cycles of R-CHOP. Three months later, he returned to the clinic with lower back pain and hip pain radiating down his left leg. MRI of the spine and pelvis showed dumbbell shaped enhancing peri-neural soft tissue process at the level of the left neural foramen of L3-L4. A CT guided biopsy of the lesion showed proliferation of lymphoma cells confirming the diagnosis of secondary neurolymphomatosis. He then underwent two cycles of high dose methotrexate, rituximab and cytarabine followed by one cycle of methotrexate and rituximab with complete remission, then underwent ASCT and he is still in clinical and radiological remission with normal blood counts six months post-transplant.

CONCLUSION

Secondary neurolymphomatosis is an extremely rare condition, but a high degree of suspicion should be adapted for early recognition. Although there is no standard treatment, high dose methotrexate, rituximab and cytarabine has proven to give complete response in this case.

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