Isolated Medical Rectus Palsy as an Initial Presentation of Non Hodgkin Lymphoma

Elfrida Fernandes[1], Suresh B.V.[2], Balachandra A. Shetty[3]

ABSTRACT

Introduction: Cranial nerves, leptomeninges and cavernous sinus are often involved in lymphomas. Isolated oculomotor nerve palsy as the first manifestation of a lymphoma is rare, particularly when none of its other manifestations are initially identified.

Case report: A 31 year old man with no known co-morbidities, came with complaints of acute onset of blurring of vision and drooping of the right eyelid. Neurological examination revealed isolated right medial rectus palsy with dilated pupil and ptosis of right eyelid, suggesting complete oculomotor nerve palsy. CT Brain, CT Angiogram and CSF study were normal.Chest X Ray showed mediastinal widening and CECT Thorax showed anterior mediastinal mass. CT guided biopsy suggested lymphoproliferative disorder. IHC was diagnostic for Thymoma. The mass was resected. Ten days later, the patient returned, with worsening of his symptoms. Neurologic examination showed third, fourth and sixth cranial nerve palsy. IHC of the resected specimen revealed high grade B cell Lymphoblastic Lymphoma. Repeat CSF analysis showed leptomeningeal involvement. Patient was started on chemotherapy for high grade lymphoma and his extra ocular movements improved. One week later, he developed bilateral facial palsy and Left CN IX, X Palsy. Chemotherapy was continued until he was discharged at request. Patient was lost to follow up.

Conclusion: This atypical presentation of NHL can bring about a delay in the diagnosis due to the variability of its presenting symptoms and wide differential diagnosis. Thus early diagnosis and aggressive management is essential.

Keywords: Oculomotor nerve, leptomeninges, Non-Hodgkin Lymphoma, Cranial Nerves

INTRODUCTION

Cranial nerves, leptomeninges and cavernous sinus are often involved in lymphomas. However, isolated oculomotor palsy as the first presenting manifestation of a lymphoma is rare, particularly when no other manifestations of lymphoma are initially identified.[1] This report describes a case of undiagnosed Non Hodgkin lymphoma presenting with isolated medial rectus palsy and subsequently complete oculomotor nerve palsy that was detected during neurological examination.

CASE REPORT

A 31 year old man with no known co-morbidities, presented with complaints of diplopia in primary position that was insidious in onset, started 10 days ago and drooping of right eyelid since 5 days. The patient was evaluated in another center 5 days ago, where MRI Brain done, was normal. The patient was diagnosed to have Tolosa-Hunt syndrome (isolated medial rectus palsy with no pupillary involvement, and no ptosis) and was started on oral corticosteroids.

The patient presented to our center with no improvement in symptoms. He gave no history of fever, headache, or seizures. Neurologic evaluation showed isolated right medial rectus palsy with pupillary involvement and ptosis of right eye lid, suggestive of complete right oculomotor nerve palsy (cranial nerve III). Rest of examination was normal. General examination was unremarkable, no lymphadenopathy, pallor or skin lesions.

Hematological and Biochemical investigations were normal. CT angiogram showed no evidence of Posterior Communicating Artery aneurysm. CSF analysis was normal. Chest X ray was normal. The patient developed dry cough after 2 days hence repeat chest X ray was done that showed mediastinal widening (Figure 1). CECT Chest showed anterior mediastinal mass (Figure 2). CT Guided biopsy was suggestive of Lymphoproliferative disorder. Immunohistochemistry (IHC) suggested Thymoma. The mass was resected and specimen was sent for IHC at another center. Patient was discharged in a stable condition but with no improvement in symptoms. The patient returned after 7 days with puffiness of face, neck. Neurological exam revealed bilateral cranial nerve III, cranial nerve IV and cranial nerve VI palsy. IHC of resected specimen showed High grade B-lymphoblastic lymphoma. Patient was started on R-CHOP therapy (Rituximab, Cyclophosphamide, Doxorubicin, Vincristine, Predisolone). The next day the patient's Abducens nerve palsy (cranial nerve VI) had resolved but oculomotor nerve (cranial nerve III) palsy persisted. (Figure 3). Two days later he developed bilateral LMN facial nerve palsy (cranial nerve VII). Repeat MRI showed no cranial nerve enhancement. Repeat CSF analysis showed Cell count of 6800, all Lymphoblasts with

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convoluted nuclei suggestive of Lymphomatous involvement of leptomeninges. Bone marrow biopsy was normal. One week later the patient developed difficulty swallowing and this time the neurological exam revealed left CN IX, X palsy. Patient continued chemotherapy, and was on Ryle's tube feeding and consequently developed leukopenia. The patient was discharged at request of relatives and lost to follow up.

Figure 1: Chest X-ray of the patient (Left) taken on day one. (Right) taken on day 3 (after the patient developed dry cough) showing mediastinal widening.

Fig. 2, CECT THORAX: Anterior mediastinal mass pushing the Superior Vena Cava posteriorly.

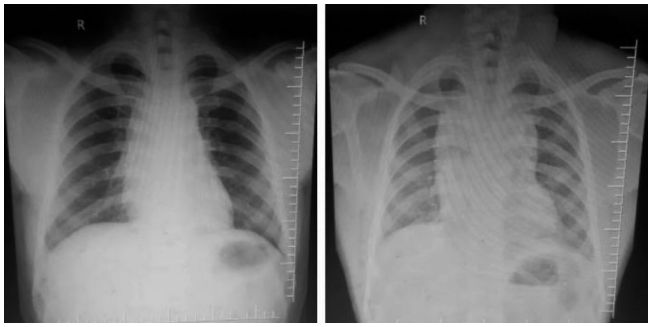

Figure-1: Chest X-ray: Left (day 1) and Right (day 3) shows mediastinal wedening

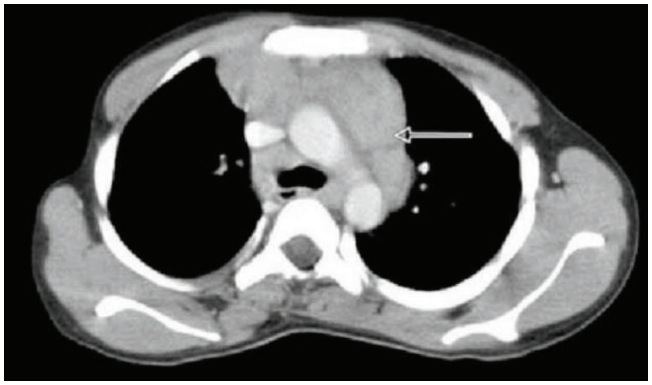

Figure-2: CECT Thorax showing anterior mediatinal mass.

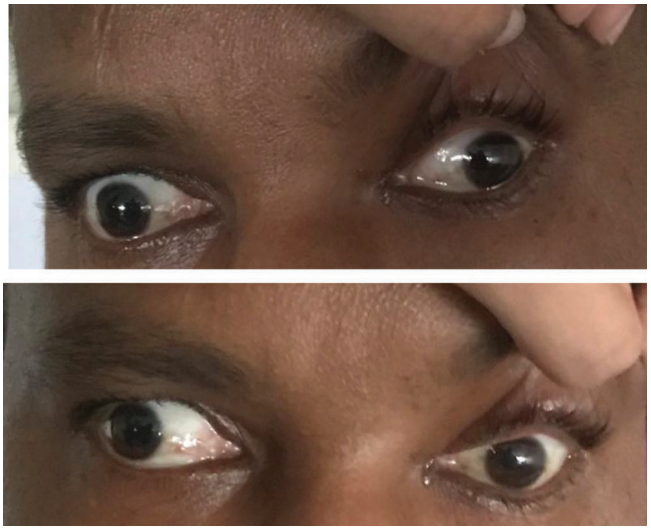

Figure-3: Persistent bilateral meial rectus palsy, with resolution of the bilateral abducens palsy

Figure 3 shows bilateral medial rectus palsy that persisted even after chemotherapy, bilateral abducens nerve palsy had resolved.

DISCUSSION

Non-Hodgkin Lymphoma (NHL) is a hematologic malignancy that originates in the lymphatic system and is characterized by clonal proliferation of the Lymphocytes.[2] An accumulation of errant lymphocytes, namely B-cells, T-cells, or natural killer cells, can disseminate throughout the body leading to both systemic and CNS involvement.[3] Since the central nervous system lacks a lymphatic system it is rare for NHL to affect the CNS, unless there is a breach in the blood-brain barrier. Typically, the lymphoma settles within 2 distinct areas of the CNS: the leptomeninges, the brain parenchyma, or both. However, the leptomeninges is most commonly affected and can present with multiple neurologic signs and symptoms, as the entire neur-oaxis is susceptible to infiltration by neoplastic lymphocytes.[4,5,6] The leptomeninges is a delicate layer of tissue that encases the brain and spinal cord. It is composed of 2 layers: the inner layer, the pia mater, and the outer layer, the arachnoid mater. In between these 2 layers is the subarachnoid space, which houses CSF. When NHL infiltrates the leptomeninges, it can spread diffusely throughout the neuro-axis by following CSF pathways within the subarachnoid space. Leptomeningeal involvement can be asymptomatic or may present with multiple cranial neuropathies, headache, spinal cord signs (i.e., focal limb numbness or weakness), mental status changes, or spinal root symptoms (i.e., radiculopathy).

Our patient presented with isolated Right Medial Rectus palsy with pupillary involvement, that suggested involvement of parasympathetic fibers of the oculomotor nucleus. Thus, a compressive lesion especially Posterior communicating artery aneurysm was ruled out by CT Angiogram. The incidental finding of mediastinal widening in the chest X-ray prompted us to consider a lymphoproliferative disorders especially lymphoma.

Cranial nerves are involved in lymphomas because their pathways intersect the subarachnoid space and pass through subarachnoid cisterns where infiltration or compression by neoplastic lymphocytes floating along the CSF pathways can occur. The cranial nerves most often affected are III, IV, V, VI, VII, VIII, and XII.[2]

However, neuro-imaging studies may be normal, as in the case of our patient. Therefore, a cytological evaluation of CSF is mandatory to look for leptomeningeal metastases in a patient who presents with cranial nerve palsies.

Diagnosis is often difficult and delayed because of variability of presenting symptoms and wide differential diagnosis including viral, inflammatory or paraneoplastic neuropathy, cranial neuritis multiplex, and nerve root compression.

CONCLUSION

Isolated Oculomotor nerve palsy as the initial presentation of NHL due to leptomeningeal lymphomatosis is rare. This atypical presentation in the absence of other supportive

signs of lymphoma brings about delay in the diagnosis and causes fatal and rapid progression. Thus, early diagnosis by effective use of imaging modalities such as MRI and PET-CT and aggressive therapies are important for prolonged survival.

ABBREVIATIONS

CN - cranial nerve, CSF- cerebrospinal fluid, CECT- contrast enhanced computed tomography, NHL - non-Hodgkin Lymphoma,

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