Ababou et al., J Clin Case Rep 2019, 9:2
DOI: 10.4172/2165-7920.10001215

Journal of Clinical Case Reports

Case Report Open Access

Multiple Cranial Nerve Palsies in Mantle Cell Lymphoma: A Case Report and Literature Review

Ababou M*, Jennane S, Mehtat EM, Elmaaroufi H and Doghmi K

Department of Clinical Hematology, Mohammed V Military Hospital, Rabat, Morocco

Abstract

Involvement of cranial nerves is unspecific and can occur in inflammation, different types of nerve tumours and in malignancies. Central Nervous System (CNS) dissemination during the Mantle Cell Lymphoma (MCL) is unusual. We report a rare case of a patient followed for mantle cell lymphoma, which brutally instituted multiple cranial nerve palsies, brain imaging showed a bilateral sphenoid sinusitis. Cytology later showed the presence of mantle cells in cerebrospinal fluid analysis. We review literature for the Central Nervous System involvement in Mantle cell lymphoma and we discuss possible change of therapeutic strategies. Although the cranial nerves involvement is extremely rare in Mantle cell Lymphoma, physicians should be aware of such patterns of Central Nervous System (CNS) involvement for the early diagnosis and adequate selection of treatment modality.

Keywords: Mantle cell lymphoma; Cranial nerves; Spinal fluid

Abbreviations: CNS: Central Nervous System; MCL: Mantle Cell Lymphoma; CT: Computed Tomography; MRI: Magnetic Resonance Imaging; ECOG: Eastern Cooperative Oncology Group; LP: Lumbar Puncture; CSF: Cerebrospinal Fluid; MNC: Mononucleated Cells; R-CHOP: Rituximab Cyclophosphamide, Hydroxydaunorubicin, Oncovin, Prednisone; R-DHAOx: Rituximab, High-Dose Aracytine, Oxaliplatin and Dexamethasone; HD-MTX: High-Dose Methotrexate; HD-AraC: High-Dose-Cytarabine; Hyper CVAD: Cyclophosphamide, Vincristine, Adriamycin, Dexamethasone; BTK: Bruton Tyrosine Kinase

Introduction

Multiple cranial neuropathy is uncommon but not rare; it has several causes with varying prognoses. Neoplastic processes are the most common cause by a wide margin [1]. CT, Magnetic Resonance Imaging, Radionuclide Scanning, and blood and spinal fluid studies are all important diagnostic tests in the workup of multiple cranial neuropathy [1].

Mantle Cell Lymphoma (MCL) is a rare lymphoma, accounting for 5% of non-Hodgkin lymphomas [2]. The Central Nervous System (CNS) involvement is a relatively rare event in the course of this type of lymphoma, it occurs in 4.1% [3].

We present a rare case of MCL with CNS involvement with multiple cranial nerve palsies and bilateral sphenoid sinusitis.

Case Report

62-year-old man was diagnosed with classical MCL in January 2018. At diagnosis, ECOG performance status was two, Ann Arbor stage was IV for pulmonary involvement, B symptoms were present, and MIPI score was six. The therapeutic decision was: 3 cycles of R-CHOP given every 21 days then reassessment for haematopoietic stem cell harvesting, 3 cycles of R-DHAOx and Autologous Stem Cell Transplantation. A few days after the 3rd cycle, our patient complained of acute onset of double vision and left eyelid droopiness, left facial deviation, swallowing difficulty with moderate headache without vomiting. He had not developed any fever in this period.

On neurologic examination, the patient was conscious, oriented and cooperated for the examination. He has symptoms and signs of involvements of the third, fourth, fifth, sixth, seventh, ninth, tenth, eleventh cranial nerves on both sides, without meningeal signs.

Computed Tomography (CT) scan and Magnetic Resonance Imaging (MRI) of the brain showed a bilateral sphenoid sinusitis; however, the Lumbar Puncture (LP) showed the presence of 1200 mononucleated cells (MNC)/mm^3 in the Cerebrospinal Fluid (CSF). The immunophenotype showed the typical pattern of mantle cell lymphoma. PET-SCAN 18-FDG showed progression of the disease with the appearance of a pontic hyper metabolic focus (Figure 1). Peripheral blood counts and biochemistry tests were normal.

For lack of the ibrutinib that is becoming a promising targeted therapy approach for MCL patients with CNS involvement, our therapeutic decision was high-dose methotrexate (HD-MTX) in combination with High-dose of cytarabine (HD-AraC). The evolution

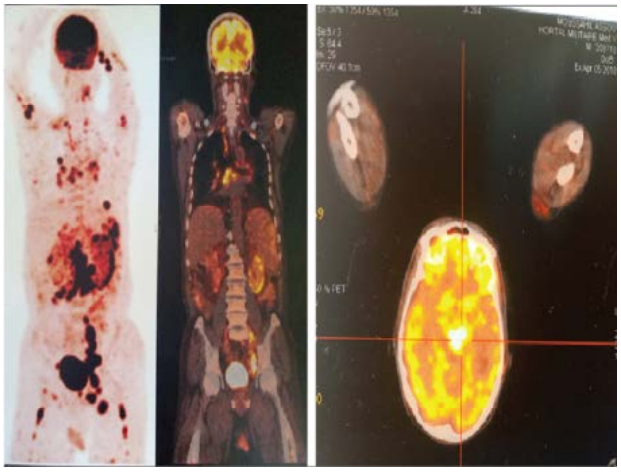

Figure 1: PET-SCAN 18-FDG showing the involvement of the pontic central nervous system.

***Corresponding author:** Ababou M, Department of Clinical Hematology, Mohammed V Military Hospital, Rabat, Morocco, Tel: +212634038175; E-mail: dr.ababoumounir@gmail.com

Received February 17, 2019; **Accepted** February 23, 2019; **Published** March 01, 2019

Citation: Ababou M, Jennane S, Mehtat EM, Elmaaroufi H, Doghmi K (2019) Multiple Cranial Nerve Palsies in Mantle Cell Lymphoma: A Case Report and Literature Review. J Clin Case Rep 9: 1215. doi: 10.4172/2165-7920.10001215

was marked by a progression of the disease with alteration of the general state. the patient died 5 months after the neurological manifestations of his lymphoma.

Discussion

Multiple cranial neuropathies affect a wide variety of cranial nerve combinations in diverse locations. It is uncommon but not rare. Several causes were reported, neurologic causes (e.g., multiple sclerosis, myasthenia gravis, Miller-Fisher syndrome, Wernicke encephalopathy, Ramsay Hunt syndrome, and heavy metal intoxication), autoimmune and vascular causes (e.g., CNS vasculitis, cavernous sinus thrombosis, Wegener's granulomatosis, systemic lupus erythematosus, polyarteritis nodosa, and sarcoidosis), infectious causes (e.g., viral [human immunodeficiency virus, progressive multifocal leukoencephalopathy, herpes simplex virus, cytomegalovirus, EBV], bacterial [Lyme, botulism, tuberculosis, nocardia, actinomyces, syphilis], and fungal [cryptococcus, aspergillosis, Mucor]), and malignant causes (e.g., leptomeningeal carcinomatosis or lymphomatosis, paraneoplastic encephalitis, lymphomas, and leukaemia's) [4,5]. The largest study was published by James R Keane about 979 unselected in patients with simultaneous or serial involvement of 2 or more different cranial nerves [6], tumour was the most common cause (30%) including lymphoma in 10 patients.

Cranial nerve lesions in lymphoma can appear at various stages of the disease and are caused by several mechanisms. Mantle Cell Lymphoma (MCL) is a rare and very aggressive subtype of non-Hodgkin lymphoma and is unique among lymphomas in its clinical, biologic, and genetic properties [7]. The Central Nervous System (CNS) involvement is a relatively rare event in the course of this type of lymphoma. After a review of the literature we find some reported cases and small series including 3-11 cases [8-15].

Cheah et al. [3] presented the largest series of 57 patients with MCL and Central Nervous System (CNS) involvement reported to date, the incidence of CNS involvement was 4.1%, the number of patients with multiple cranial nerve palsies has not been specified.

Then and Patel [16] presented a case close to our, a 65 year old white woman diagnosed with MCL, managed on Hyper CVAD (Cyclophosphamide, Vincristine, Adriamycin, Dexamethasone) who presented with acute onset of double vision, skew deviation of the eyes, left eye ptosis, right horizontal gaze palsy, right facial droop, dysarthria and dysphagia two months after the lymphoma diagnosis. Brain imaging was normal, Lumbar puncture showed an exaggerated lymphocytic pleocytosis (white cell count of 1681), Cytology later showed the presence of mantle cells on CSF analysis.

Once established, CNS involvement in MCL carries a poor prognosis with 3.7 months as median survival [3]. It has a very poor prognosis with current treatment options [17]. Our patient has evolved Recent limited studies tested Ibrutinib, an oral inhibitor of Bruton tyrosine kinase (BTK) and showed efficacy in mantle cell lymphoma with CNS involvement [13].

We think that the incidence of CNS involvement in MCL might be higher than previously recognized. So, we propose to look for CNS involvement in MCL when the clinical suspicion is high. we think also that the development of CNS prophylactic treatment strategies is urgently required, especially in high-risk patients for CNS involvement, as example patients with High Ki-67 [18].

Conclusion

Multiple cranial nerve palsies are a rare situation, it has several causes including tumours at the top of the list. The Central Nervous System involvement is a relatively rare event in the course of Mantle cell lymphoma, as we see in our case it can cause also multiple cranial palsies with normal brain imaging, a situation with a very poor prognosis. We think that studies should focus more and more on the CNS involvement in mantle lymphoma, with the establishment of new therapeutic strategies and criteria for possible Central Nervous System Prophylaxis at diagnosis.

Declarations

Acknowledgements

I would like to thank the entire medical and paramedical team of the clinical haematology department of the Military hospital Mohammed V in Rabat, for the enormous work they do.

Authors contributions

MA, SJ and EM collected data and drafted the manuscript. HE and KD reviewed the literature All authors read and approved the final manuscript.

Ethics approval and consent to participate

The authors declare no conflicts of interest between the authors and that this work was performed with all the due respect to the code of ethics.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

References

1. Beal MF (1990) Multiple cranial-nerve palsies: A diagnostic challenge. Mass Medical Soc 2: 1.
2. Jaffe ES (2009) The 2008 WHO classification of lymphomas: Implications for clinical practice and translational research. ASH Education Program Book 1: 523-531.
3. Cheah C (2013) Central nervous system involvement in mantle cell lymphoma: Clinical features, prognostic factors and outcomes from the European Mantle Cell Lymphoma Network. Ann Oncol 24: 2119-2123.
4. Habermann TM, Steensma DP (2000) Subspecialty clinics: Hematology. In Mayo Clin Proc 75: 723-732.
5. Weiss GA, Al-Ustwani MO, Makdissi MR (2012) Multiple cranial nerve palsies as the initial presentation of peripheral T-cell lymphoma. Clin Adv Hematol Oncol 10: 620-623.
6. Keane JR (2005) Multiple cranial nerve palsies: Analysis of 979 cases. Arch Neurol 62: 1714-1717.
7. Jares P, Campo E (2008) Advances in the understanding of mantle cell lymphoma. Brit J Hematol 142: 149-165.
8. Montserrat E (1996) CNS involvement in mantle-cell lymphoma. J Clin Oncol 14: 941-944.
9. Oinonen R, Franssila K, Elonen E (1999) Central nervous system involvement in patients with mantle cell lymphoma. Ann Hematol 78: 145-149.
10. Valdez R (2002) Cerebrospinal fluid involvement in mantle cell lymphoma. Modern Pathol 15: 1073.
11. Ferrer A (2007) Central nervous system involvement in mantle cell lymphoma. Ann Oncol 19: 135-141.
12. Gill S (2009) Mantle cell lymphoma with central nervous system involvement: Frequency and clinical features. Brit J Haematol 147: 83-88.
13. Bernard S (2015) Activity of ibrutinib in mantle cell lymphoma patients with central nervous system relapse. Blood 126: 1695-1698.
14. Koban Y (2017) Mantle cell lymphoma presenting with acute bilateral ophthalmoplegia. Turk J Ophthalmol 47: 235.
15. Mannina D, Loteta B (2017) Ibrutinib treatment of mantle cell lymphoma

relapsing at central nervous system: A case report and literature review. Case Rep Hematol pp: 1-4.

16. Then R, Patel M (2015) Rare case of central nervous system invasion of mantle cell lymphoma with serial negative magnetic resonances: Case report and Literature review. Neurol 84: 146.

17. Campo E, Rule S (2015) Mantle cell lymphoma: Evolving management strategies. Blood 125: 48-55.

18. Chihara D (2015) Ki-67 is a strong predictor of central nervous system relapse in patients with mantle cell lymphoma (MCL). Ann Oncol 26: 966-973.