Mantle Cell Lymphoma

In small cell lymphomas, central nervous system (CNS) involvement has been considered to be very rare. Mantle cell lymphoma (MCL) is a distinct subtype of non Hodgkin lymphomas consisting of small or intermediate lymphatic B-cells. It has a poorer prognosis than the other small cell lymphomas. Only a few MCL patients with CNS involvement have been reported in the literature to date.

Oinonen et al., analyzed retrospectively the incidence, clinical characteristics, and outcome of CNS involvement in 94 patients with confirmed MCL treated at one center from 1980 to 1997. Four of the 94 patients (4%) developed CNS lymphoma during the median follow-up of 51 months. The diagnosis was based on clinical, cytological and radiological findings. CNS involvement appeared at 4.6, 56, 66, or 86 months from the diagnosis of MCL. All patients had neurological symptoms and a leukemic disease; two cases were seen with a blastoid morphology. Malignant lymphatic cells were detected in spinal fluid in all cases and parenchymal infiltrations in brain in two. All patients were treated with intrathecal chemotherapy, without response. Survival time after diagnosis of CNS lymphoma ranged from 18 to 55 days. At diagnosis, no adverse prognostic factors predictive of CNS lymphoma were found. CNS involvement was associated with a progressive leukemic disease as a late event or a blastoid transformation. The prognosis of MCL patients with CNS involvement is poor [1].

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a 71-year-old-patient receiving antiplatelet therapy and being attended by emergency medical services for psychomotor retardation and gait disturbance. An emergency computed tomographic scan showed a bilateral subacute hematoma. The patient reported a fall 2 weeks earlier. We performed bilateral drills and saw a solid mass that was biopsied. The patient had a history of mantle cell lymphoma (MCL) in complete remission (results of bone marrow biopsy and whole-body positron emission tomography-computed tomography scans were normal 6 months earlier). We diagnosed an intracranial MCL by immunohistochemistry, flow cytometry, and fluorescence in situ hybridization. We performed magnetic resonance imaging. The results of a new bone marrow biopsy were positive for recurrence of MCL. MCL constitutes approximately 5%-6% of non-Hodgkin lymphoma. The incidence of central nervous system (CNS) involvement between MCLs is 4.1%. After a review of the literatures we found small series comprising 3-5 cases and a multicenter study with 57 cases. Until now, the median survival was 3.7 months. Ibrutinib, an oral Bruton tyrosine kinase inhibitor, has demonstrated efficacy and CNS penetration in relapsed or refractory MCL with rapid and complete response even after 1 year of follow-up. Our patient received ibrutinib and had a complete response at 3 months, which was maintained to the present (6 months). After a review of the literature, we found different pathologies that can mimic subdural hematomas. However, this is the first report of a lymphoma with CNS involvement mimicking bilateral subdural hematomas. This report contributes to the knowledge of lymphomas with CNS involvement. Its strange radiographic appearance and histologic type make it unique [2].

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Oinonen R, Franssila K, Elonen E. Central nervous system involvement in patients with mantle cell lymphoma. Ann Hematol. 1999 Mar;78(3):145-9. Review. PubMed PMID: 10211757.
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González-Bonet LG, García-Boyero R, Gaona-Morales J. Mantle Cell Lymphoma with Central Nervous System Involvement Simulating Bilateral Subdural Hematomas. World Neurosurg. 2017 Mar;99:808.e1-808.e2. doi: 10.1016/j.wneu.2016.12.122. Epub 2017 Jan 4. PubMed PMID: 28063897.