Primary intracranial marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue a cerebral localization: case report

K.Tlohi, A.Benmoussa,M.Iamchahab, M.Rachid, N.Khoubila, S.cherkaoui, M.Qachouh, A.Madani
Department of Clinical Hematology and Pediatric Oncology, CHU IBN ROCHD, Casablanca
Faculty of Medicine and Pharmacy, HASSAN II University, Casablanca

INTRODUCTION

- Primary central nervous system lymphoma (PCNSL) is an aggressive extra nodal subtype of non-Hodgkin's lymphoma.
- In over 90% of cases, PCNSL is classified as diffuse large B-cell lymphoma. Rarely, PCNSL may be classified as marginal zone B-cell lymphoma (MZBCL) of mucosa-associated lymphoid tissue (MALT)..

AIM

- Describing a primary MALT-type MZBCL arising in a cerebral localization as an extremely rare presentation.

CASE DESCRIPTION

➢ *Clinical presentation :*

A 61-year-old woman, with a history of uterine fibroid surgery 20 years ago, presents a medical history dating back 4 months before the diagnosis. She experienced worsening headaches, along with loss of consciousness and generalized tonic-clonic seizures, which led her to seek consultation with a neurosurgeon. Radiological assessment and streotactic biopsy were performed, revealing a diagnosis of small B-cell lymphoma. The patient was referred to us for specialized management.

Upon admission, the clinical examination was normal, with no evidence of tumor-related symptoms, sensory deficits, or motor deficits

➢ *Paraclinics examinations:*

Examination	Results
Cerebral MRI	Bilateral frontal extra-axial masses measuring 69*35mm on the right and 20*14mm on the left.
Thoraco- abdnomino-pelvic CT scan	-Degenerative spine
Anatomopatho-logic study	Appearance suggestive of small cell lymphoma, raising the possibility of marginal zone lymphoma (BCL 2+, CD10-, CYC D1-, BCL 6-, CD138-,CD23-, low Ki-67)
Bone marrow biopsie	Morphological and immunohistochemical appearance consistent with nodular and interstitial medullary infiltration by a small B-cell lymphoma of the marginal zone.
CBC	WBC= 3400. NEUT= 1400 , LYMPH= 1200 HB=12,4 g/dl PLT=132 000 /mm3
Pre-therapeutic assessment	Echocardiographie was normal Viral serologies were negative. Renal and liver profil was normal.



Figure 1 : Cerebral MRI of the patient at diagnosis

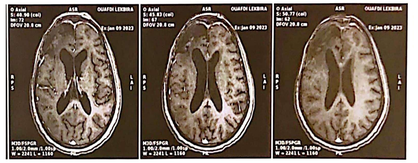


Figure 2 : Cerebral MRI of the patient after treatement

Treatement and Evolution:

The patient was treated with a Rituximab in day 1 of each course (375 mg/m2 in IV for fisrt course and 1400 mg in SC for 5 courses) – Bendamustine (100 mg total dose per day in day 1 and 2 of each course). She underwent 6 cycles with favorable clinical and radiological progress.

Eight months after treatment, she remained neurologically intact and free of disease under surveillance in consultation.

CONCLUSIONS

- Primary cerebral MZL is a rare, indolent low-grade CNS lymphoma, with a relatively good prognosis. Its treatment is multidisciplinary and often requires surgical intervention due to brain compression, along with low to moderate doses of radiotherapy and/or systemic chemotherapy.

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