Meningeal Involvement in Primary Plasma Cell Leukemia

Giovanni D'Arena¹ · Cinzia Bitetti¹ · Bruna Daraia² · Giuseppe Pietrantuono¹ · Pellegrino Musto³

Received: 31 January 2018 / Accepted: 27 February 2018 / Published online: 12 March 2018
© Indian Society of Hematology and Blood Transfusion 2018

Keywords Plasma cell leukemia · Central nervous system · Meningeal involvement

A 46-year old woman was diagnosed as having primary plasma cell leukemia (pPCL), with 4000/μl circulating plasma cells, massive bone marrow plasma cells infiltration and t(4;14),del 1p,1q gain. She was treated with bortezomib, liposomal doxorubicin and dexamethasone with immunofixation negative complete response. However, just before starting peripheral stem cell mobilization therapy, she experienced right facial nerve palsy. Column and cerebral magnetic resonance imaging (MRI) showed no apparent central nervous system (CNS) involvement. However, lumbar puncture and cerebro-spinal fluid (CSF)

cytospin documented meningeal infiltration of CD138+ CD56+ CD38+ plasma cells (Fig. 1). A very small IgG/λ paraprotein in serum and 10% of bone marrow plasma cells were also found. Intrathecal administrations of methotrexate, cytarabine and dexamethasone with cranial-lumbar radiotherapy were given, obtaining CSF plasma cells disappearance and complete resolution of neurological symptoms. The initial therapeutic program was restarted.

pPCL is a rare and very aggressive disease with poor prognosis. Meningeal localization is rare, but the exact prevalence is unknown because of reported cases are limited. A high number of circulating plasma cells seems to correlate with an increased incidence of CNS involvement.

✉ Giovanni D'Arena
giovannidarena@libero.it

¹ Hematology and Stem Cell Transplantation Unit, IRCCS “Referral Cancer Center of Basilicata”, Rionero in Vulture, PZ, Italy

² Hematology Unit, “Madonna delle Grazie” Hospital, Matera, Italy

³ Scientific Direction, IRCCS “Referral Cancer Center of Basilicata”, Rionero in Vulture, PZ, Italy

Fig. 1 Meningeal infiltrating plasma cells in CSF (a, b, and c) co-expressing CD138, CD56 and CD38 (d)

This case highlights on the need to consider CNS prophylaxis and investigation in pPCL patients with neurological symptoms or simply at risk.

Compliance with Ethical Standards

Conflict of interest All Authors have no conflict of interest to disclose.

Ethical Approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed Consent Informed consent was obtained from the patient.