H14 Where is the Neoplasm? The Postmortem Diagnosis of Intravascular Large B-Cell Lymphoma (IVLBCL)

Zoe Del Fante, MD, Rome, ITALY; Nicola Di Fazio, Università La Sapienza di Roma, Rome 00161, ITALY; Silvia Romano, Università La Sapienza, Rome 00161, ITALY; Alessandra De Matteis, MD, University Sapienza of Roma, Rome 00100, ITALY; Paola Santoro, MD, Rome, ITALY; Aniello Maiese, MD, Sapienza University of Rome, Rome 00185, ITALY*

Learning Overview: The goal of this presentation is to share the study results that demonstrate that the best instrument to make a diagnosis is represented by immunohistochemical staining of a tissue specimen (with a specific panel made of CD20 and CD45), while instrumental exams (Computed Tomography [CT], Magnetic Resonance Imaging [MRI], Positron Emission Tomography [PET], and biopsy) exhibit a high number of false negative results.

Impact on the Forensic Science Community: This presentation will impact the forensic science community by how this results in a valid aid for the pathologist when approaching an extremely rare disease, such as IVLBCL.

IVLBCL is a rare non-Hodgkin extranodal B-cell lymphoma that occurs with a prevalence lower than 1%. It has an aggressive clinical behavior and it is characterized by the presence of malignant lymphoid cells within the blood vessels, more specifically capillaries. Making the diagnosis can be challenging because of the heterogeneous and non-specific presentation of the disease due to the generalized organ dysfunction secondary to vascular obstruction; neurological signs and symptoms are the most common presentation of the disease. Clinical diagnosis is based on imaging (CT scan, MRI), peripheral blood smear, cerebrospinal fluid cytology, biochemical examination, and bone marrow biopsy results. Nevertheless, diagnosis may be difficult and literature suggests that very often the disease is recognized only after autopsy examination.

The case presented herein is that of a 50 year-old man with a history of myeloradiculitis hospitalized for paraparesis. The CT showed no evidence of lesions or other altered findings. Lumbar-spine MRI showed conus medullaris swelling between D11 and L1 and vasogenic edema of anterior and posterior cords. Although PET imaging showed an increase uptake in the adrenal glands and a focal area within the mesenteric adipose tissue, both serologic exams and bone marrow biopsy were negative. Spinal cord biopsy showed necrotic nervous tissue with low CD68+ macrophage infiltrates, reactive gliosis, and negative staining for MAP2. After two months, the patient died of respiratory failure.

Gross autopsy examination was negative, apart from findings suggestive of pulmonary infection and reduced consistency of the spinal cord. Examination of histologic samples from the brain, heart, lung, and liver tissue showed collections of atypical cells with enlarged, pleomorphic, hyperchromatic nuclei within blood vessels and interstitium as well as lymphocyte infiltration. Immumohistochemical examination was performed evaluating for the following markers: cytokeratin, CD3, CD20, CD45, PDL-1, CK20, CK7, MCK, and TTF1. A strong intravascular positivity for CD45 and CD20 was found. In sum, these findings would support a B-cell lineage for the neoplastic cells and the postmortem diagnosis of intravascular large B-cell lymphoma.

This case report demonstrates that the most reliable means of making the postmortem diagnosis of IVLBCL is via immunohistochemical staining of tissue specimens (with a specific panel including CD20 and CD45). Other techniques such as radiologic studies (CT, MRI, PET) or bone marrow biopsy show a high number of false negative results.

Intravascular Large B-Cell Lymphoma, Postmortem Diagnosis, Immunohistochemistry

*Presenting Author