Nonenhancing secondary central nervous system lymphoma

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Case report

Central nervous system (CNS) lymphoma is an aggressive neoplasm. Incidence is increasing, partly due to the use of immunosuppressive drugs and AIDS-related immunodeficiency. Nonenhancing lesions in CNS lymphoma are very rare, and often cause diagnostic delay. We report a case of nonenhancing secondary CNS lymphoma.

We present a 77-year old man with a medical history of nine attacks of transient pancytopenia during the last five years, associated with fever, elevated lactate dehydrogenase (LDH), and hepatosplenomegaly. Bone marrow aspiration and liver biopsy were normal. In order to treat pancytopenia of unknown origin, oral corticosteroid treatment was started during the last attack of pancytopenia, three month before admission.

The patient presented with progressive drowsiness and gait unsteadiness since one month. Brain MRI showed brainstem, bitemporal, periventricular, and meningeal abnormalities (Fig.). Lumbar puncture showed 150 lymphocytes/mm³. Anatomopathological examination of CSF revealed a large B-cell lymphoma. A diagnosis of secondary brain lymphoma was made after bone marrow aspiration showed the presence of a diffuse large B-cell lymphoma (centroblastic variant). Lack of typical gadolinium enhancement of brain lesions in our patient may be related to the intake of corticoids.

Discussion

Primary CNS lymphomas represent approximately 70-90% of all CNS lymphomas.

Approximately 10-30% of patients with systemic lymphoma may develop secondary CNS involvement. Almost all CNS lymphomas are non-Hodgkin B-cell tumors. Ninety percent of CNS lymphomas are supratentorial, commonly affecting frontal and parietal lobes, deep grey nuclei, corpus callosum and clustering around ventricles and gray matter/white matter junction. Lesions are often multiple. Spinal cord and leptomeninges are more frequently affected in secondary CNS lymphoma. The clinical presentation of CNS lymphoma is often non-specific and depends on the location of the neoplasm. Brain CT shows classically hyperdense lesion with uniform contrast enhancement. MRI shows most often homogeneous isointense/hypointense lesion on T1- and T2-weighted images with mild surrounding oedema (1, 2). Diffusion may be restricted on diffusion weighted images. Classically, gadolinium enhancement is strong and homogeneous. Ring-enhancing lesions with central necrosis are often seen in immunocompromised patients. Nonenhancing lesions in CNS lymphoma are very rare and often cause diagnostic delay. Repeated CSF examination, brain biopsy, 18F-fluorodeoxyglucose positron emission tomography (18FDG-PET) and thallium-201 single photon emission computed tomography (Tl-SPECT) can help to diagnose CNS lymphoma. Response to steroids and radiation therapy is often spectacular but short lasting. Long-term survival is poor. This is the first report of nonenhancing secondary CNS lymphoma. However, several cases of nonenhancing primary CNS lymphoma have been described, most often during or after corticoid treatment (2-8). It is unclear if findings of described cases can be extrapolated to secondary CNS lymphoma. Corticoid treatment is a well known cause of...
delayed or wrong diagnosis of CNS lymphoma. Therefore, if possible, the use of steroids has to be avoided before diagnostic procedures, when CNS lymphoma is suspected.

REFERENCES


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