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Definitive Diagnosis of Intravascular Large B-Cell Lymphoma Originates in Extranodal Organs

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DESCRIPTION

Historically, most cases of intravascular large B-cell lymphoma were diagnosed postmortem. However, today it is possible to diagnose and treat these cases antemortem. Although hematogenous spread of non-lymphoma malignant cells is beyond the scope of current therapies, intravascular large B-cell lymphoma (hematogenous spread of lymphoma) can be treated with chemotherapy, making an accurate diagnosis. The onset and clinical course of intravascular large B-cell lymphoma are heterogeneous. Although many cases of intravascular large B-cell lymphoma present with rapid deterioration, others have a relatively gradual initial stage followed by rapid progression. Outbreaks of leukemia are not uncommon. It is difficult to distinguish between the intravascular large B-cell lymphoma and lymphoma that originates in and spreads systemically to extranodal organs. Accurate diagnosis requires minimally invasive and sensitive procedures.

Bone marrow biopsy and biopsy, and random skin biopsy are recommended. If intravascular large B-cell lymphoma is suspected, glucocorticoid therapy should be avoided before biopsy, even in severe cases, to obtain a correct diagnosis. Intravascular large B-cell lymphoma responds markedly to treatment with the rituximab-containing chemotherapy. Delaying rituximab administration and reducing chemotherapy doses in the first cycle may be the initial indications for older patients, poor performance, or high tumor burden. Aggressive combination therapy with the high-dose methotrexate is a novel idea because of the central nervous system involvement, but otherwise relapses are common and prognosis is poor.

Intravascular large B-cell lymphoma is a rare form of the non-Hodgkin's lymphoma characterized by the growth of tumor cells in the lumen of small to medium-sized blood vessels. Diagnosis of intravascular large B-cell lymphoma is usually not possible with computed tomography or magnetic resonance imaging because there is little or no accompanying solid disease. Bone marrow aspiration and biopsy revealed hemophagocytosis without involvement of the lymphoma cells. A random skin biopsy showed no lymphoma involvement. Fluorodeoxyglucose Positron emission tomography/computed tomography showed accumulation of fluorodeoxyglucose in the uterus. Magnetic resonance imaging showed only one uterine leiomyoma. Based on these findings, an endometrial biopsy was performed and a histologic diagnosis of the intravascular large B-cell lymphoma involving the uterus was made.

Abdominal computed tomography scan revealed bilateral renal hypertrophy, and general condition and renal function deteriorated rapidly. Soluble interleukin-2 receptor (sil-2R) levels rose to 5,928 U/ml and gallium scintigraphy showed poor uptake in both kidneys. Considering the possibility of malignant lymphoma, a renal biopsy was performed, and no glomerular abnormalities were observed. Accumulation was observed. Gallium scintigraphy showed no uptake in other organs, and examination of cerebrospinal fluid and bone marrow showed no tumor cells, so the patient was presumed to have kidney-confined intravascular large B-cell lymphoma. Chemotherapy was started immediately and general condition improved. Renal function deteriorated enough to require dialysis, but weaned from it. After chemotherapy, her enlarged kidneys returned to normal size. Then underwent intermittent chemotherapy and had no relapses. Intravascular large B-cell lymphoma, which primarily affects the kidneys, is generally difficult to diagnose antemortem and is sometimes found at autopsy.

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CONFLICT OF INTEREST

The author's declared that they have no conflict of interest.

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