Initial management of HHV-8-positive multicentric Castleman disease

Newly diagnosed HHV-8-positive MCD

Is there concurrent Kaposi sarcoma (KS)?

Yes

- Weekly rituximab for four weeks plus liposomal doxorubicin*
- Repeat cycles until response achieved
- Followed by surveillance and retreatment with this regimen at relapse

Is there life-threatening organ failure or poor performance status thought to be due to the MCD?Δ

Yes

- Weekly rituximab until response achieved*
- Followed by surveillance and retreatment at relapse

Progressive organ failure at any time

Consider clinical trial

No

Progressive organ failure at any time

- Weekly rituximab for four weeks plus either liposomal doxorubicin or etoposide*
- Followed by surveillance and retreatment with this regimen at relapse

HHV-8: human herpesvirus 8; MCD: multicentric Castleman disease; HIV: human immunodeficiency virus; IL: interleukin; VEGF: vascular endothelial growth factor; FDG PET/CT: fluorodeoxyglucose positron emission tomography/computed tomography.
* Antiretroviral therapy is started or continued for all patients with HIV infection. In addition, antiviral therapy directed at HHV-8 with ganciclovir is offered to patients with concurrent KS and/or uncontrolled concomitant HIV infection (eg, CD4 count <200 cells/mm³, high HIV viral load) since such patients are at risk for the development of or exacerbation of KS lesions when treated with rituximab.
Surveillance includes clinical evaluation, serum biomarkers, and imaging. Serum biomarkers include complete blood count, blood chemistries, IL-6, VEGF, C-reactive protein, serum free light chain assay, and quantitative immunoglobulins. Imaging with whole body FDG PET/CT is preferred. Examples of life-threatening organ failure that may be related to MCD include respiratory failure, renal failure, liver failure, and pancytopenia.