

"Double-expressor" intravascular large B-cell lymphoma involving the female genital tract

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A 43-year-old woman was admitted with chronic pelvic pain and unexplained fever. Her laboratory findings were notable for anemia (hemoglobin, 10.6 g/dL). No masses were detected by pelvic ultrasound. She underwent a total abdominal hysterectomy with bilateral salpingo-oophorectomy. Large atypical lymphocytes were noted within the vascular lumina of the cervix, endomyometrium, bilateral fallopian tubes, and ovaries (panels A-B; hematoxylin and eosin stain, original magnification $\times 40$ [A] and ×200 [B]). They were positive for CD20, CD5, CD10, BCL-2 (panel C; original magnification ×40), and MYC (panel D; original magnification ×200) immunohistochemical stains. MYC was expressed in 40% to 50% of the lymphoma cells. The KI-67 (MIB-1) proliferation rate was 90%. The patient was diagnosed with intravascular large B-cell lymphoma, with double expression of BCL-2 and MYC. Fluorescent in situ hybridization showed gain of 1 copy of the MYC gene. BCL-2 and MYC rearrangements were not detected. She had minimal bone marrow involvement (<5%). Lactate dehydrogenase was elevated at 1652 U/L (normal, 300-600 U/L). She received 6 cycles of rituximab, etoposide phosphate, prednisone, vincristine sulfate (Oncovin), cyclophosphamide, and doxorubicin hydrochloride (hydroxydaunorubicin) (R-EPOCH) and prophylactic intrathecal methotrexate followed by an autologous stem cell transplant and has achieved complete remission.

Intravascular lymphomas are uncommon and carry a poor prognosis. Uterine involvement is rare and is manifested as vaginal bleeding and/or fever. MYC and BCL-2 protein coexpression in diffuse large B-cell lymphomas are associated with inferior survival and central nervous system relapse; however, their prognostic relevance in intravascular lymphomas is unclear.



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