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EBV$^+$ ALK$^+$ large B-cell lymphoma

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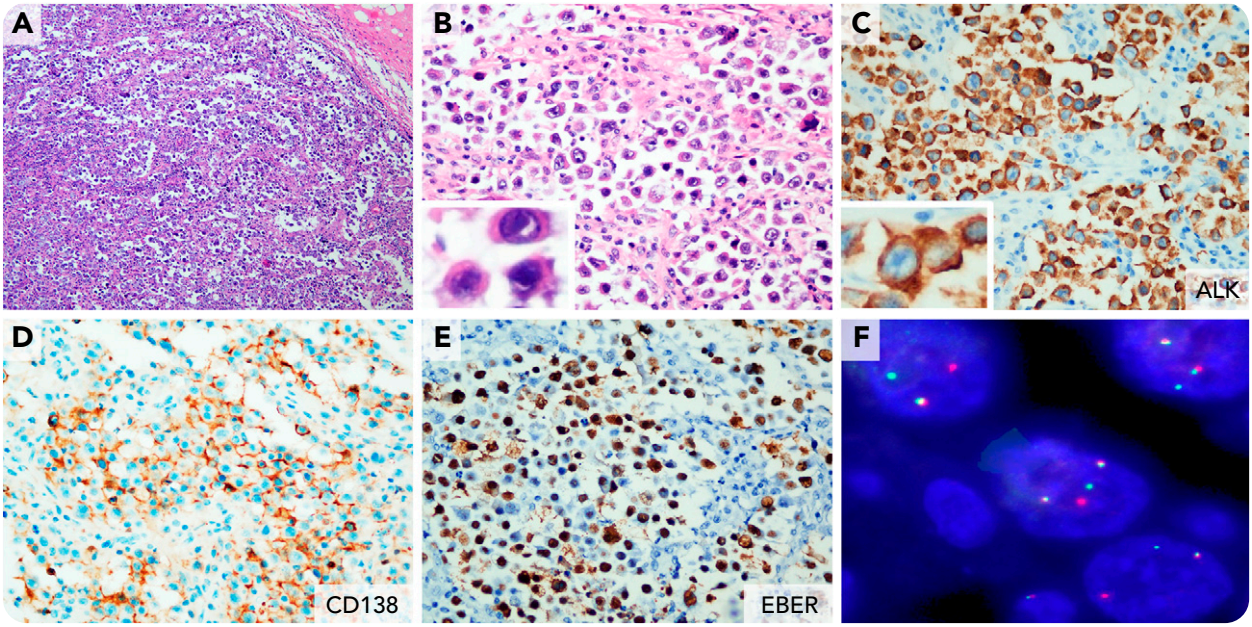

A 42-year-old man who had no known immunodeficiency presented with a 4-day history of a right cervical mass. A computed tomography scan revealed bilateral cervical, supraclavicular, mediastinal, abdominal, and retroperitoneal lymphadenopathy. A cervical lymph node biopsy showed a neoplasm with a striking sinusoidal growth pattern (panels A and B; hematoxylin and eosin stain; original magnification ×100 [A] and ×400 [B]). The tumor cells had a plasmablastic appearance with eccentric nucleus, distinct nucleoli, and abundant eosinophilic cytoplasm. They were positive for ALK, CD138, MUM1, and EBER (90%), weakly positive for CD22 and κ, and negative for PAX5, CD19, CD20, CD79a, λ, CD3, CD5, CD43, LMP1, and EBNA2 (panels C-E; original magnification ×400). The Ki-67 proliferation rate was ~70%. FISH studies revealed ALK gene rearrangement (panel F) and no *MYC* gene rearrangement. Serum Epstein-Barr virus (EBV) viral load was 4.12×10^4 IU/mL. The patient was diagnosed with EBV$^+$ ALK$^+$ large B-cell lymphoma (LBCL), EBV latency type I. He received EPOCH (etoposide, prednisone, vincristine, cyclophosphamide, and doxorubicin) for 6 cycles and achieved complete remission. He was free of disease 65 months after the initial diagnosis.

ALK$^+$ LBCL is a rare entity, with ~200 cases reported in literature. This is the first reported case of EBV$^+$ ALK$^+$ LBCL. This case raises the awareness of this rare disease among the differential diagnosis of EBV$^+$ LBCL and expands the spectrum of EBV$^+$ lymphoma currently recognized in the World Health Organization classification.

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DOI 10.1182/blood.2021013595

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