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ALK-positive large B-cell lymphoma, a rare B-cell lymphoma expressing ALK

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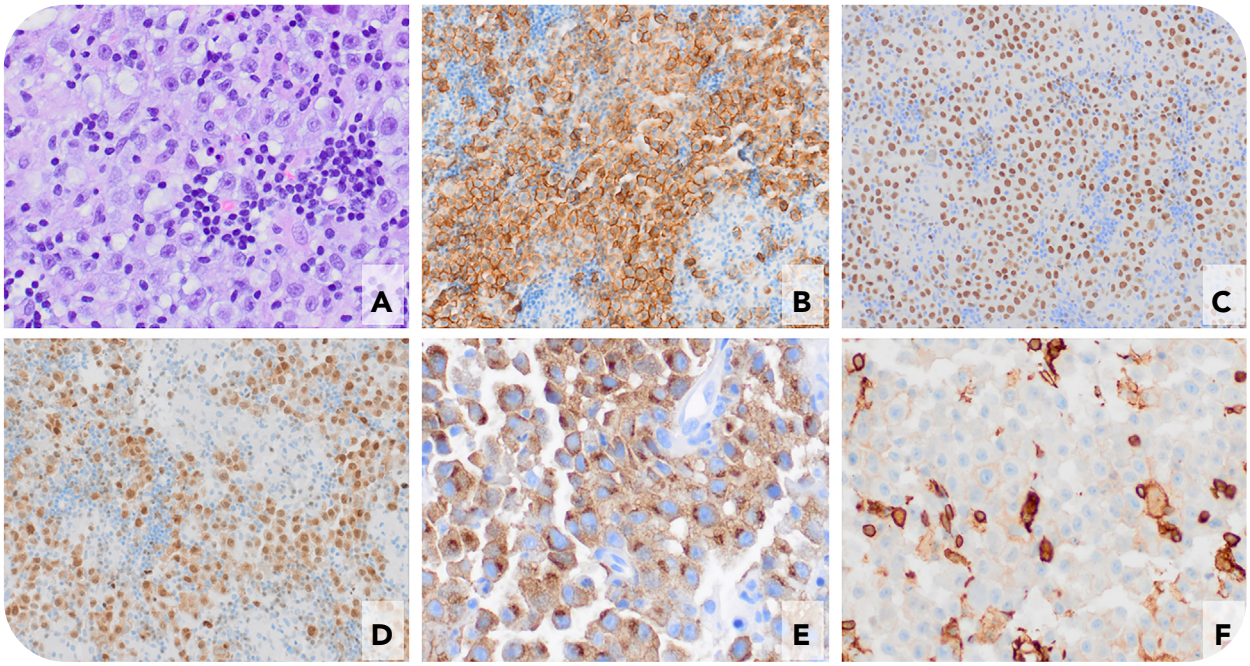

The patient is a 29-year-old male presenting with rapidly enlarging cervical adenopathy and no previous significant clinical history. Histologic sections show a diffuse lymphoma composed of large immunoblasts/plasmablasts (panel A; original magnification ×100, hematoxylin and eosin stain). The neoplastic cells are positive for CD138 (panel B; original magnification ×40), OCT2 (panel C; original magnification ×40), BOB1 (panel D; original magnification ×40), ALK (panel E; original magnification ×100), MUM-1, and EMA and negative for CD45 (panel F; original magnification ×100), CD30, HHV8, EBER, CD19, CD20, CD22, kappa, and lambda. The expression of ALK, with a granular cytoplasmic positivity, supported the diagnosis of ALK$^+$ large B-cell lymphoma (ALK$^+$ LBCL).

ALK$^+$ LBCL is a rare (<200 cases reported) and aggressive lymphoma with a plasmablastic immunophenotype and ALK expression. Other neoplasms showing ALK expression due to *ALK* gene rearrangements include ALK$^+$ histiocytosis, anaplastic large cell lymphoma (ALCL), inflammatory myofibroblastic tumors, and non–small cell lung cancers. The immunohistochemical staining pattern of ALK differs depending on the *ALK* fusion partner. A characteristic granular cytoplasmic pattern is associated with *CLTC::ALK* and *SEC31A::ALK*, whereas a nongranular pattern is associated with *NPM1::ALK*, *SQSTM1::ALK*, *RANBP2::ALK*, and *EML4::ALK* rearrangements. If the partner is *NPM1* (the most frequent partner in ALCL), the staining is both nuclear and cytoplasmic. In ALK$^+$ LBCL, some data suggest that a nongranular cytoplasmic pattern is associated with a more aggressive behavior.

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