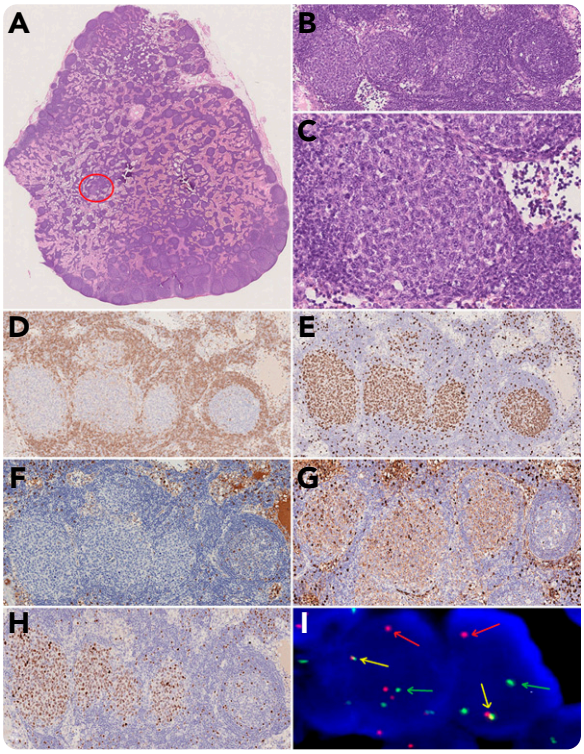
blood work
images in hematology

Early pattern of large B-cell lymphoma with *IRF4* rearrangement

Massimo Granai, University Hospital Tuebingen; and Stefano Lazzi, University of Siena



A slightly enlarged mesenteric lymph node included in an appendectomy specimen from a 12-year-old boy was examined. The lymph node architecture was preserved with florid germinal centers (GCs) and sinus histiocytosis. Three adjacent atypical GCs enriched in centroblasts lacked tingible body macrophages and showed absence of polarization (panels A-C; original magnification ×30 [A], ×100 [B], ×200 [C]; hematoxylin-and-eosin stain). Their immunophenotype was CD20$^+$, CD10$^+$, BCL6$^+$, BCL2$^-$ (panel D; original magnification ×100, BCL2 stain), with a high proliferation rate (panel E; original magnification ×100, Ki67 stain), λ light-chain restriction (panels F-G; original magnification ×100; K stain [F], λ stain [G]), and IRF4/MUM1$^+$ (panel H; original magnification ×100, IRF4/MUM1 stain). Fluorescence in situ hybridization (FISH) analyses for *BCL6*, *BCL2*, and *MYC* genes were negative. Notably, an *IRF4* rearrangement with a gain of the *IRF4* gene was detected (panel I; FISH analysis with break-apart probes showed split red/green signals in 85% to 90% of tumor nuclei with a gain of the *IRF4* gene, with an average of 3.5 and 3.4 red and green signals, respectively, per neoplastic cell; yellow arrows, normal alleles; red and green arrows, breakages). Laser-capture microdissection of the atypical GC confirmed identical VDJ rearrangements. Thus, our case might represent early or focal involvement of large B-cell lymphoma (LBCL) with *IRF4* rearrangement characterized by a follicular pattern without diffuse areas. The patient underwent complete clinical staging, which showed no evidence of disseminated disease. A watch-and-wait approach was adopted and, after 12 months, the patient is still healthy.

LBCL with *IRF4* rearrangement is a rare entity. An in situ variant of this neoplasm has never been reported; thus, its diagnostic criteria and clinical significance are yet to be defined.

ASH Image Bank

For additional images, visit the ASH Image Bank, a reference and teaching tool that is continually updated with new atlas and case study images. For more information, visit http://imagebank.hematology.org.

DOI 10.1182/blood.2020006406