blood work
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γ Heavy chain disease

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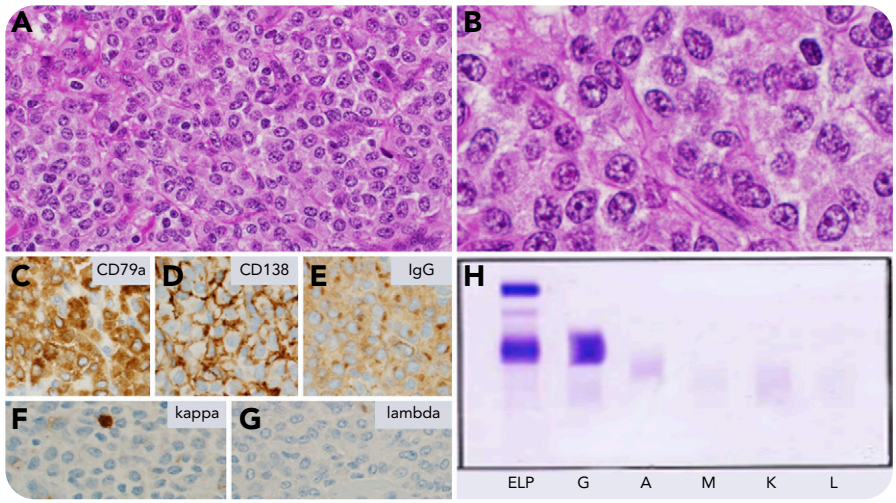

A 78-year-old woman from Sudan with presumed tuberculous lymphadenitis without preexisting autoimmune disease presented with fatigue, persistent fever, hepatosplenomegaly, and generalized lymphadenopathy. Inguinal lymph node biopsy showed a diffuse neoplasm with plasmacytic features, including eccentric nuclei, marginated chromatin, small nucleoli, and abundant pale cytoplasm (panels A-B; hematoxylin and eosin stain, original magnification ×400 [A] and ×1000 [B]). Immunohistochemical stains were CD19$^+$ and CD79a$^+$ (panel C; original magnification ×400); CD138$^+$ (panel D; original magnification ×400); CD38$^+$, IRF4/MUM1$^+$, and IgG$^+$ (panel E; original magnification ×400); and CD5$^-$, CD10$^-$, CD20$^-$, Pax-5$^-$, cyclin-D1$^-$, CD56$^-$, immunoglobulin A [IgA]$^-$, IgD$^-$, IgM$^-$, κ$^-$, and λ$^-$ (panels F-G; original magnification ×400). Suspected γ heavy chain disease was confirmed by serum (panel H) and urine immunofixation electrophoresis demonstrating IgG band and no light chains. Anemia (hemoglobin, 88 g/L) and elevated serum IgG 101.16 g/L (normal, 7.00-16.00 g/L) were notable. The patient deteriorated rapidly with complications of sepsis and died.

Interestingly, presence of only plasma cells is a feature of this case. γ Heavy chain disease is a B-cell neoplasm producing a truncated γ heavy chain with variable lymphocytes, plasmacytoid lymphocytes, and plasma cells. The diverse morphology and systemic symptoms can delay the diagnosis. Serum or urine immunofixation electrophoresis demonstrating IgG without light chains is diagnostic. Extent of disease, serum IgG levels, and associated autoimmune disorders determine the prognosis from indolent to aggressive.

DOI 10.1182/blood-2018-11-887133