Dysmorphic Cells of Anaplastic Plasma Cell Leukemia

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CASE PRESENTATION

An 86-year-old man presented with anemia (Hgb 6.1g/dL) and renal failure (serum creatinine 2.7 mg/dL). Hematological evaluation revealed serum protein electrophoresis (SPEP) with IgG lambda 1.66 g/dL, urine protein electrophoresis (UPEP) positive for lambda light chains, serum free light chain assay with abnormal kappa/lambda ratio of 18.63/16007.27 (0.00116), erythrocyte sedimentation rate of 71, lactate dehydrogenase (LDH) of 277 Units/L (125–243), serum calcium and albumin were normal. Peripheral blood smear showed rouleaux formation, occasional tear drop cells, and ~15% circulating plasma cells (PC), some of which were 'multilobulated' (**Figure 1**). Peripheral blood flow cytometry reported 7.4% PC. CT imaging of chest, abdomen and pelvis without contrast displayed a right pleural effusion, pelvic lymphadenopathy, and diffuse osteopenia. Bone marrow evaluation showed hyper-cellularity (~70%), with an increase in PC (~75%), many multilobulated with peculiar nuclear segmentation (**Figures 2A, 2B**), expressing CD138 and lambda (**Figures 3A, 3B, 3C**) on immunohistochemistry. Flow cytometric analysis on bone marrow aspirate revealed CD38 positive PC neoplasm (84% of WBC, >99% lambda light chain. Cytogenetics showed 46XY, fluorescent in situ hybridization (FISH) revealed monosomy 13; deletion 1p +- duplication of 1q; Tp53 deletion; IgH rearrangement (14q32) with t(4;14); trisomy 15; gain 19; trisomy 7 and gain 3/3q. Based on the morphology and number of circulating PCs,

Figure 1. Peripheral blood smear showing rouleaux formation and circulating anaplastic plasma cell with multilobulated nucleus (x 1000 original magnification).

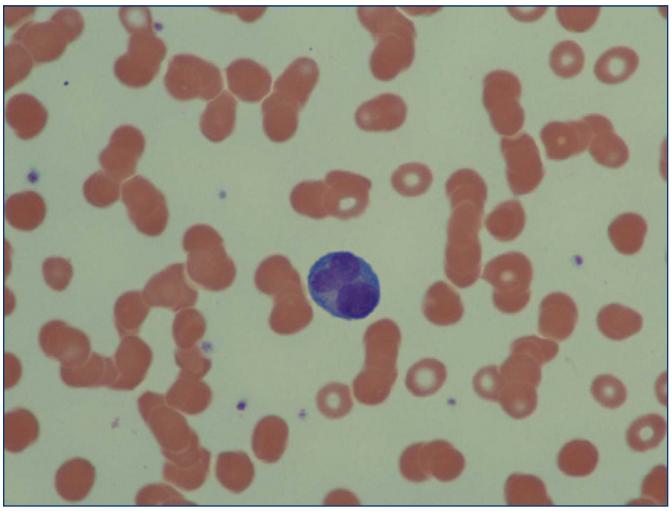
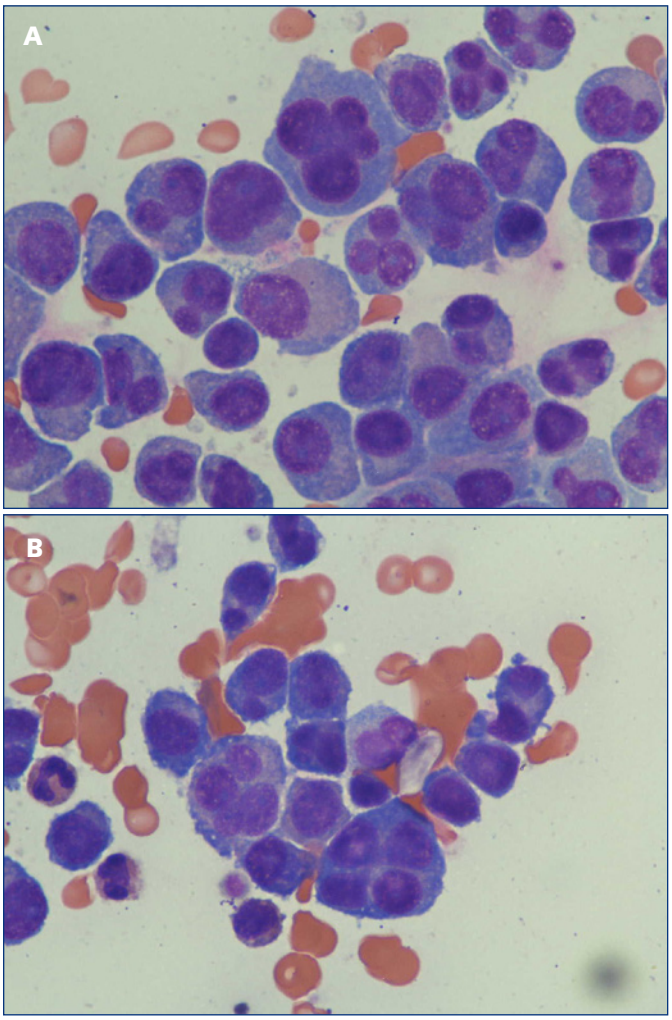
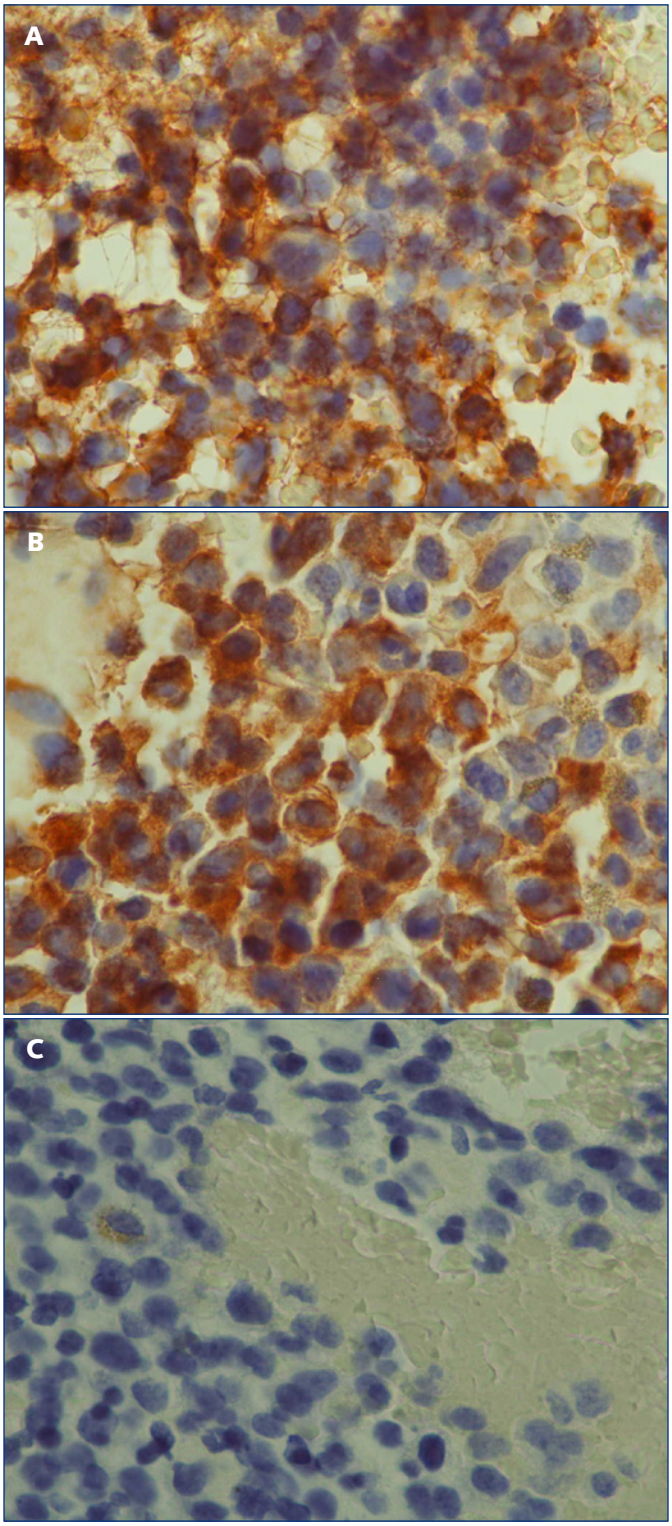


Figure 2A, 2B. Bone marrow aspirate revealing markedly pleomorphic plasma cells with multilobulated nucleus (x 1000 original magnification).



Figures 3A, 3B, 3C. Immunohistochemistry of the bone marrow aspirate (x 1000 original magnification) showing CD138 positivity [**A**], lambda positivity [**B**] and kappa negativity [**C**].



anaplastic plasma cell leukemia (APCL) was diagnosed; treatment was initiated with cyclophosphamide, bortezomib, and dexamethasone.

APCL (de novo or evolved) represents an extraordinarily rare variant of myeloma, with adverse cytogenetics, an aggressive course, poor responsivity to conventional chemotherapy, and poor survival. Lymphadenopathy and pleural effusions can be found. LDH can be elevated, and FISH (PCL) may express monosomy 13, del 17p, and abnormalities of chromosome 1. The morphology of the PCs is bizarre, and might be confused with carcinoma, an aggressive lymphoproliferative disorder, osteoclastic giant cells, or dysplastic megakaryocytes.

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