74-year-old man with h/o central spinal stenosis at L3-L5 and CKD who presents after a fall 3 days ago. CT revealed a large intrabdominal mass w/ associated right pelvic mass concerning for malignancy.
Mass, right lower quadrant, core needle biopsy

Contributed by Dr. Frank Zhao
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Immunohistochemistry

CD4

CD25

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Immunohistochemistry

CD30

CD43

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Immunohistochemistry

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PAX5  ALK1
Additional Studies

Paraffin immunoperoxidase stains show that the neoplastic cells are CD30+, CD4+, CD25+, CD43+, CD45+, focal EMA+, vimentin+, with a proliferation index of ~60-70% (by Ki-67 staining). They are negative for ALK1, CD3, CD5, CD7, CD10, CD15, CD20, CD34, CD56, MUM1, MART-1, NKX3.1, Pankeratin, PAX5, PAX8, S100, and Epstein-Barr virus (EBER).
Final Diagnosis

MASS, RIGHT LOWER QUADRANT, CORE NEEDLE BIOPSY:
- ANAPLASTIC LARGE CELL LYMPHOMA, ALK-NEGATIVE
Discussion

• ALK-negative (ALK-) anaplastic large cell lymphoma (ALCL) is a CD30+ T-cell neoplasm with a similar morphology to ALK-positive (ALK+) ALCL. It was included as a provisional entity in the 2008 edition of WHO classification but is now considered an accepted entity.

• Morphologic features include cohesive sheet-like sinus pattern of infiltration, pleomorphic and multinucleated cells, hallmark cells with eccentric horseshoe-shaped, or kidney-shaped nuclei, as seen in classical ALK+ ALCLs.

• Major differential diagnoses include CD30+ peripheral T-cell lymphoma, not otherwise specified (PTCL-NOS), classic Hodgkin lymphoma (CHL), and primary cutaneous ALCL (C-ALCL).
Discussion (cont’d)

• Differential diagnosis of the current case:
  • It is distinguished from the PTCL-NOS by the classical ALCL morphology and immunophenotype (except for the ALK negativity);
  • It is distinguished from CHL by the PAX5 negativity;
  • It is distinguished from C-ALCL by the lack of skin involvement.

• Genetic abnormalities include recurrent activating mutations of JAK1 and/or STAT3 pathway, DUSP22 rearrangements, rearrangements of TP63, and multiple copy number abnormalities, but none of these genetic abnormalities plays a role in distinguishing ALK- ALCL from other entities.

• So far there is no specific targeted therapy for ALK- ALCL. With the current regimen, its prognosis is better than PTCL-NOS and worse than ALK+ ALCL.