Cases of Primary Small cell variant Multiple Myeloma: A Single Institution Experience

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Disclosure

No Disclosure
Multiple Myeloma (MM) can exhibit a spectrum of morphologic features.

Rarely, we have encountered cases with predominantly small lymphocyte–like morphologic features mimicking mature small B-cell lymphoma.

**IgH translocation t(11;14)** has been described in up to 25-70% of primary plasma cell leukemia (pPCL) cases and is associated with **small cell variant MM**
Case 1

A 51 year male presented with back pain.

**Laboratory evaluation** showed a white blood cell count (WBC) 40 x 109/L, hemoglobin (Hb) of 4.4 g/dL. Serum protein electrophoresis (SPEP) revealed M spike of 0.4g/DL and immunofixation (IFE) consistent with Kappa chain myeloma.

**Peripheral flow cytometry** showed cells which are CD 38 +, CD 20+, CD 138 +, partial CD 56 + and Kappa +. Bone marrow (BM) demonstrated 92% plasma cells.

**Fluorescence In Situ Hybridization (FISH)** showed positivity for deletion 17P, 13Q and CCND1/IGH @ for translocation (11;14)(q13;q32). Skeletal survey revealed innumerable lytic lesions. He was treated as PCL, small cell variant.
Case 1

Treatment with Cyclophosphamide, Bortezomib and Dexamethasone (CYBORD) was initiated and he completed 5 treatments.

His WBC normalize initially but after his fifth cycle his WBC count began to rise.

Repeat Computed Tomography was consistent with progression of disease.

Further, he received bortezomib, dexamethasone, thalidomide, cisplatin, doxorubicin, cyclophosphamide and etoposide (VDT PACE) with poor response and hospice placement.
Case 2

A 74 year male presented with back pain.

**Laboratory evaluation** showed a serum calcium of 15 mg/dl and Hb of 10 g/dL. SPEP revealed M spike of 5 g/dL and IFE consistent with IgA Kappa myeloma.

**BM aspirate** showed lymphoid appearing plasma cell. Flow cytometry revealed +CD 38, partial CD20 and CD 56 cells.

**FISH** was positive for 1 q gain and CCND1/ IGH gene rearrangement with translocation (11;14)(q13;q32).

**Skeletal survey** demonstrated multiple lytic lesions.
He was treated as lymphoid small cell variant MM with CYBORD. He was not considered a B transplant candidate due to poor performance status.

He completed 11 cycles and had substantial peripheral neuropathy and Parkinsonism. Subsequently started on Revlimid and Dexamethasone.

He was ultimately switched to Pomalidomide, Carfilzomib and dexamethasone due to skin toxicity from Revlimid.

He completed 10 cycles and chose hospice care because of deteriorating status.
Discussion

The morphology of the plasma cells on bone marrow biopsy depicts a rare small cell variant which can often be confused with B cell neoplasm.

In addition, the translocation (11;14)(q13;q32) is associated with expression of CD 20 and cyclin D1 upregulation leading to a diagnostic dilemma.

In pCL, contrary to small cell variant MM, t(11;14)(q13;q32) may represent a more aggressive form of disease.
Discussion

*Treatment* with **proteasome inhibitors** remains the backbone of primary treatment however the prognosis remains poor. In trials, *Venetoclax* monotherapy has demonstrated activity in patients with Relapsed/Refractory MM positive for t(11;14).

As the genomic landscape of plasma cell dyscrasias continues to evolve it is important to focus on potential new therapies. There may be a role of incorporating novel agents including more recent **anti CD 20 therapies** in the primary treatment of small cell variant MM.