Mantle cell lymphoma presenting with autoimmune hemolytic anemia

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BACKGROUND

- Autoimmune hemolytic anemia (AIHA) occurs most frequently with AITL and CLL but is much rarer in other types of NHL, such as mantle cell lymphoma (MCL).
- Secondary AIHA due to MCL has been reported particularly with the indolent form.
- Indolent MCL is usually characterized by leukemic disease, simple karyotypes, hypermutated immunoglobulin heavy chain variable region (IGHV), and a gene expression that does not include SOX11.

CASE PRESENTATION

- History:

A 73-year-old female with no significant past medical history was referred to the clinic for anemia.

- Labs:

CBC – WBC 3.8 x 10E3 uL, Hemoglobin 5.9 g/dL
Hemolytic markers: LDH 342 IU/L, total bilirubin 5.0 mg/dL, haptoglobin undetectable, absolute reticulocyte count 160,000 uL
Antibody screen positive. Direct coombs test positive for IgG-mediated warm antibody.

- Peripheral smear had moderate reticulocytes and occasional spherocytes.

- Imaging

PET-CT with diffuse mildly FDG avid lymphadenopathy above and below the diaphragm with spleen involvement.

- Pathology

Peripheral blood flow cytometry with CD5+ B-cell population.
Bone marrow biopsy: involvement by B-cell lymphoma with immunohistochemistry (IHC) positive for CD5 and cyclin D1. IHC was positive for SOX11. Karyotype was normal. Proliferative index with Ki-67 was low.

DIAGNOSIS AND TREATMENT

- MCL diagnosis was made with bone marrow biopsy demonstrating involvement by B-cell lymphoma with IHC positive for CD5 and cyclin D1.
- MIPI risk stratification was 6.5 points (high risk).
- Hemoglobin improved to 8.5 g/dL after 2-week prednisone taper.
- Patient has received 3 of the planned 6 cycles of Bendamustine + rituximab (BR) chemoimmunotherapy.

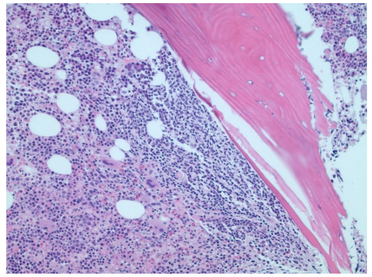

Figure 1:
20x magnification demonstrating involvement of B-cell lymphoproliferative disorder.

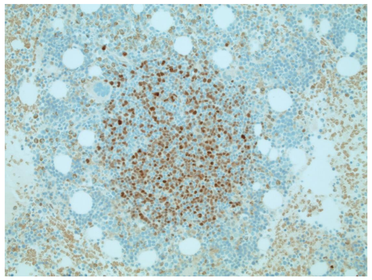

Figure 2:
Cyclin D1 IHC

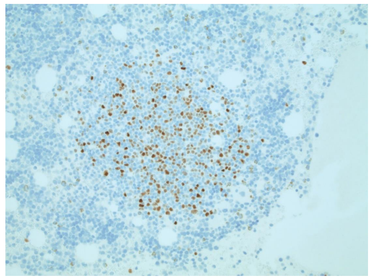

Figure 3:
SOX11 IHC

DISCUSSION

- Warm AIHA comprises 70-80% of AIHAs.
- Secondary warm AIHA is seen in 11% of all CLL cases but is only seen in 2-3% of other NHLs.
- Secondary warm AIHA has been reported with the indolent form of MCL.
- Although our patient did not have leukemic disease and had SOX11 expression, the mildly FDG avid lymphadenopathy, simple karyotype, and low proliferative index suggest an indolent course.
- Rituximab alone or in combination with chemotherapy is an active treatment for secondary AIHA.
- After 3 cycles of Bendamustine + rituximab chemoimmunotherapy, the patient's anemia resolved.
- Despite this being an infrequent occurrence, clinicians should be aware that AIHA is a potential complication of MCL.

REFERENCES

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