Challenges of Diagnosing Diffuse Large Cell Lymphoma vs. EBV Mucocutaneous Ulcer

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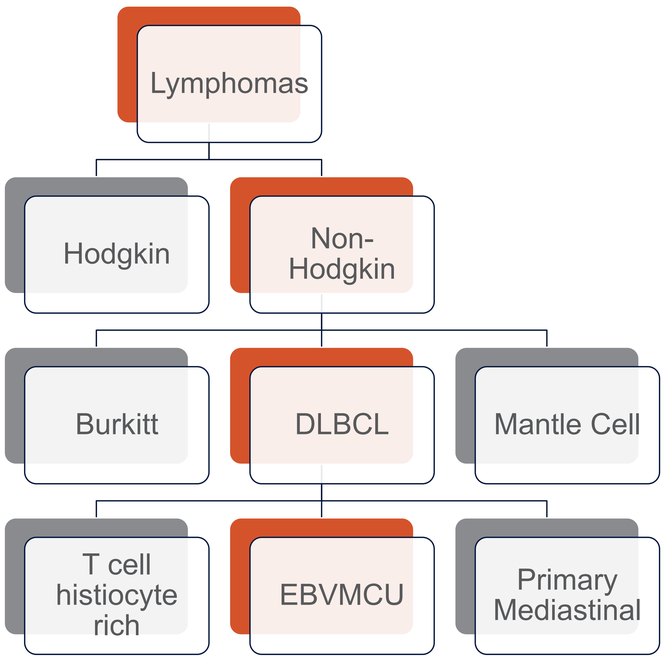
Background

- Lymphomas are broadly classified as Hodgkin's Lymphoma and non-Hodgkin's Lymphoma. Of the non-Hodgkin Lymphomas, Diffuse Large B cell Lymphoma (DLBCL) is the most common, making up about 25% of cases.
- DLBCL usually occurs de novo and is also an AIDS defining malignancy.
- DLBCL usually presents with an enlarging, symptomatic mass, most commonly found on the neck or the abdomen
- Epstein Barr Virus Mucocutaneous Ulcer (EBVMCU) is a subset of DLBCL
- It was first recognized as a lymphoproliferative disorder in 2010 and subsequently became a formally recognized clinicopathologic entity in the 2017 revision of WHO classification.
- Risk Factors:
  - Immunosuppressant drugs: methotrexate, tacrolimus, monoclonal antibodies
  - Age (Elderly)
  - Gender (Female)
- Pathology: EBV positive, atypical B-cells that may resemble Hodgkin and Reed Sternberg cells
- Pathophysiology: Though not fully established, it is thought to be reduced T cells in immunosuppressed patients that leads to inability to address all EBV associated antigens.
- Clinical Presentation:
  - Isolated, well demarcated ulcerations of the buccal mucosa
  - Ulcers on the skin or along the GI tract
  - No systemic symptoms (fever, night sweats)
- Treatment: Most cases have a benign self-limited course and respond to conservative and supportive management. Occasional persistent cases need more aggressive intervention (surgery, radiation, chemotherapy, immunotherapy)

Case Presentation

- 50-year-old Caucasian male with a long-standing history of Crohn's disease, on multiple therapies, most recently on ustekinumab for 4 years, presented to the emergency department with recurrent episodes of hematochezia and resultant severe anemia.
- CT Abdomen and Pelvis showed two separate areas of abnormal mass like thickening in the ascending colon. Findings were concerning for malignancy versus infectious/inflammatory colitis
- He underwent right hemicolectomy and incidentally had complications including perforation, resection of ileocolic anastomosis with end ileostomy & washout
- Formal Pathology Results:
  - EBV positive B Cell Lymphoproliferative Disorder, favor EBV positive Mucocutaneous Ulcer
  - Concurrent EBV lymphoproliferative disorder not be ruled out
- Because of the ambiguity between different pathologies, case was sent to an external pathology lab for a final review, they concurred with diagnosis
- EBV titers resulted as positive



Discussion

- We present a case of an individual with risk factors for both diseases: DLBCL and EBVMCU
- Important to note that the patient has a history of chronic immunosuppression, which has been documented as a risk factor for EBVMCU
- Next steps include staging for lymphoproliferative disorder once surgical recovery is complete
- The uniqueness of this case not only lies in its rare pathology, but also management going forward
- EBVMCU management is mostly conservative
- DLBCL management is primarily systemic therapy including chemotherapy

Conclusions

- This case highlights the significance of diagnostic accuracy in light of polar opposite treatment options
- It emphasizes the importance of recognizing less common, but important subtypes of DLBCL
- Further investigation should include more accurate methods of diagnosis and better understanding of risk factors and epidemiology
- It underscores value of understanding and recognizing the risks of chronic immunosuppression and EBV associated disorders

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