Case of the fortnight:

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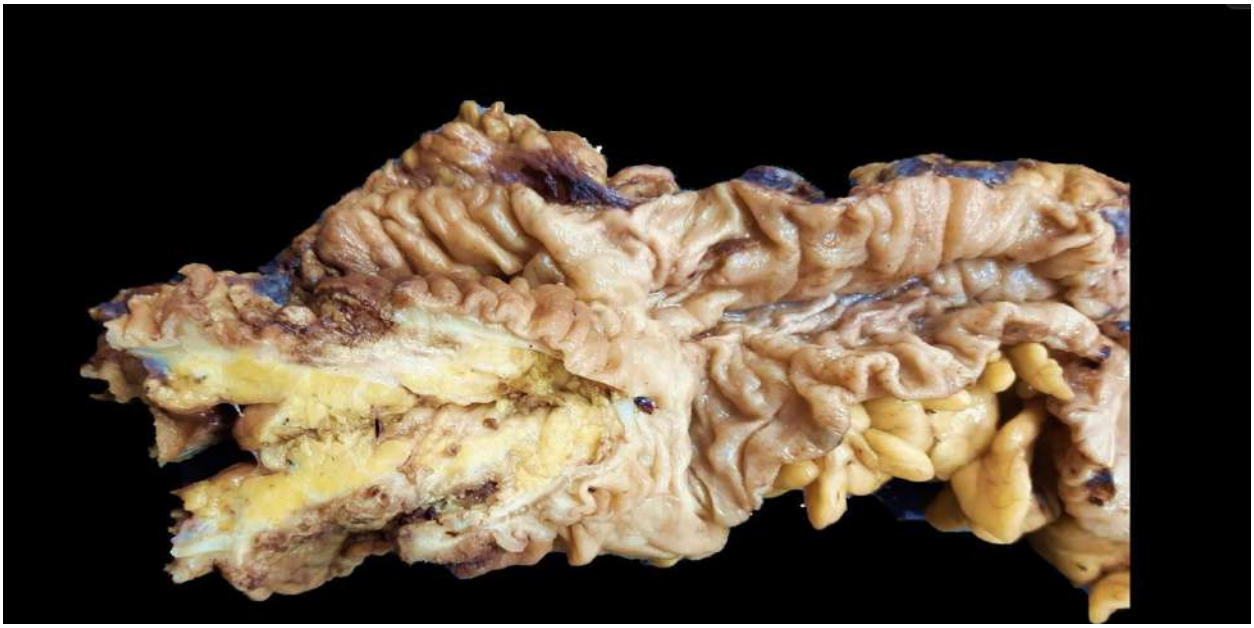
Case History:
38-year-old female presented to emergency room with spontaneous bleeding per rectum. Patient is a known case of rheumatoid arthritis and is on methotrexate since 2 years. Physical examination revealed that the patient is pale and is moderately built , vague abdominal tenderness in lower quadrant. Patient was managed conservatively initially in emergency department. Owing to the massive bleeding surgical intervention was sought for and patient underwent left hemicolectomy surgery and the specimen was sent for histopathological examination.

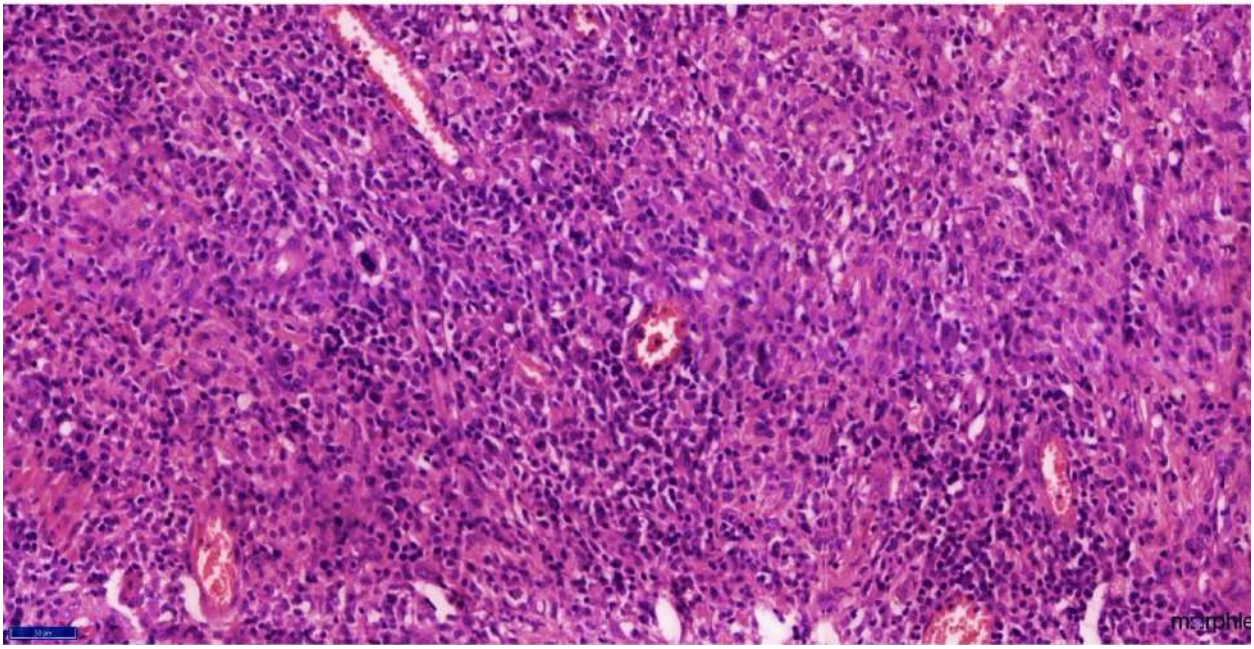
Gross:

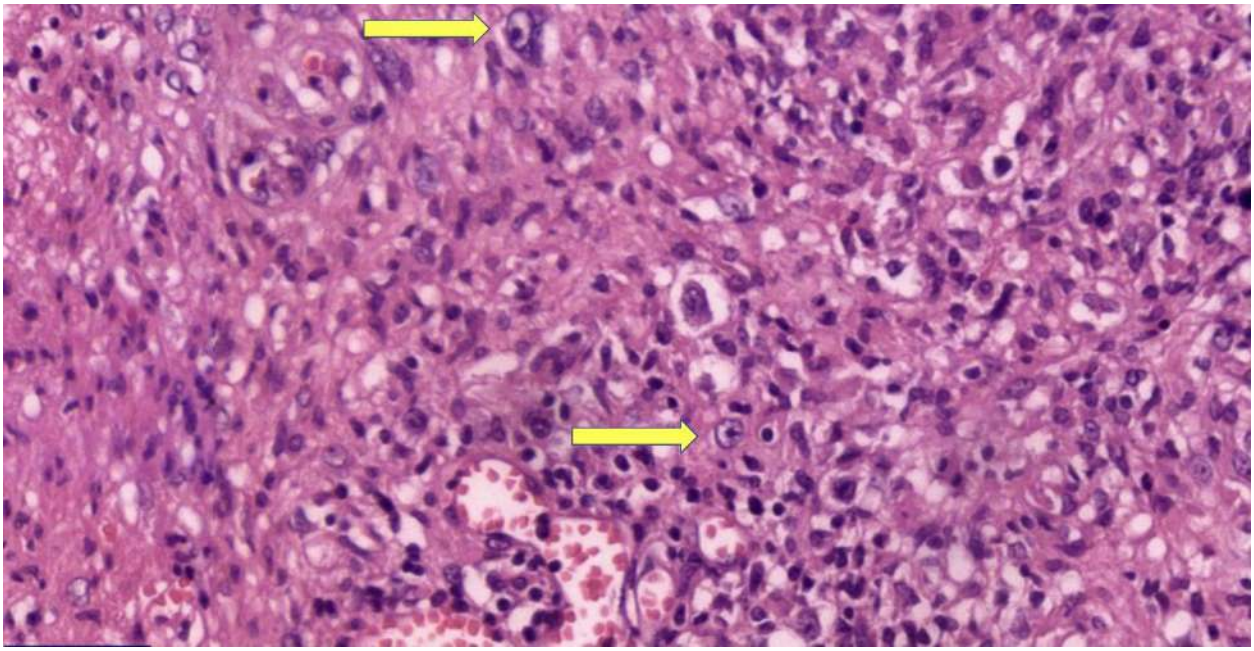
On gross examination of the specimen, there was a diffusely infiltrative grey-white thickening (measuring 3x1.5x1cm) at a distance of 28 cm from proximal dissected margin, firm in consistency. Serosal aspect was unremarkable. Adjacent bowl mucosa was normal.



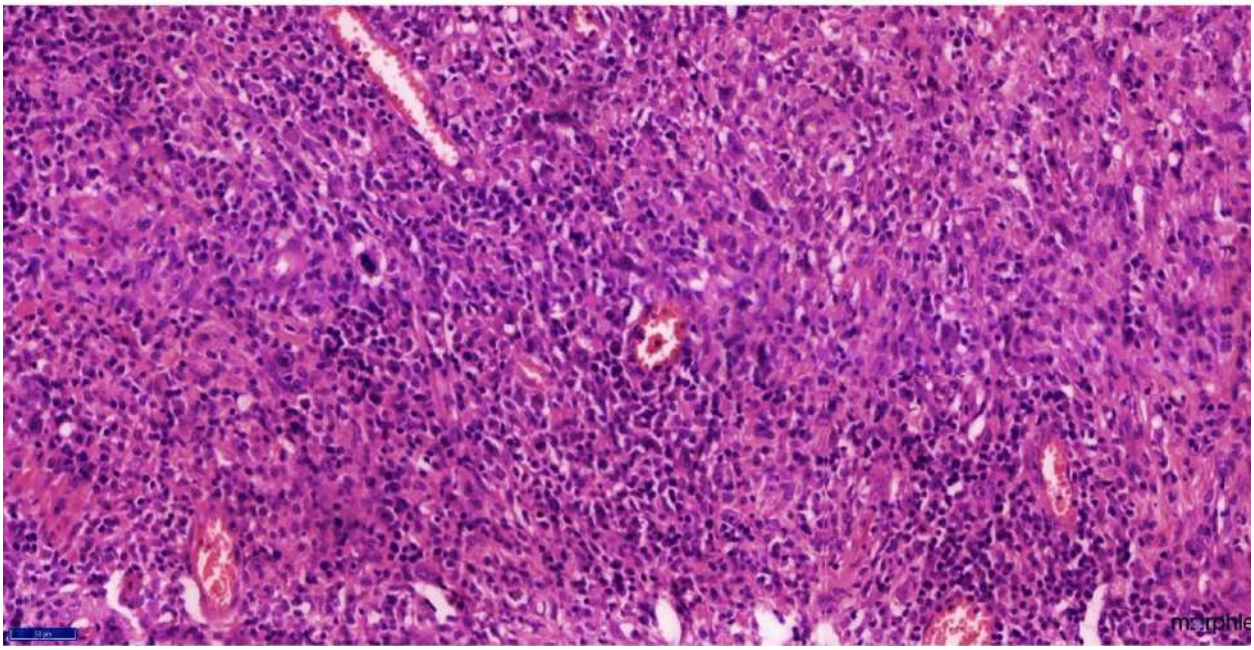
Gross picture showing diffuse mucosal thickening of bowel wall.

Microscopy:
Microscopic examination of multiple sections show ulceration with dense band of polymorphous infiltrate composed of plasma cells, lymphocytes, histiocytes, immuno blasts, large cells resembling Reed sternberg cells and apototic bodies. Areas of necrosis noted.

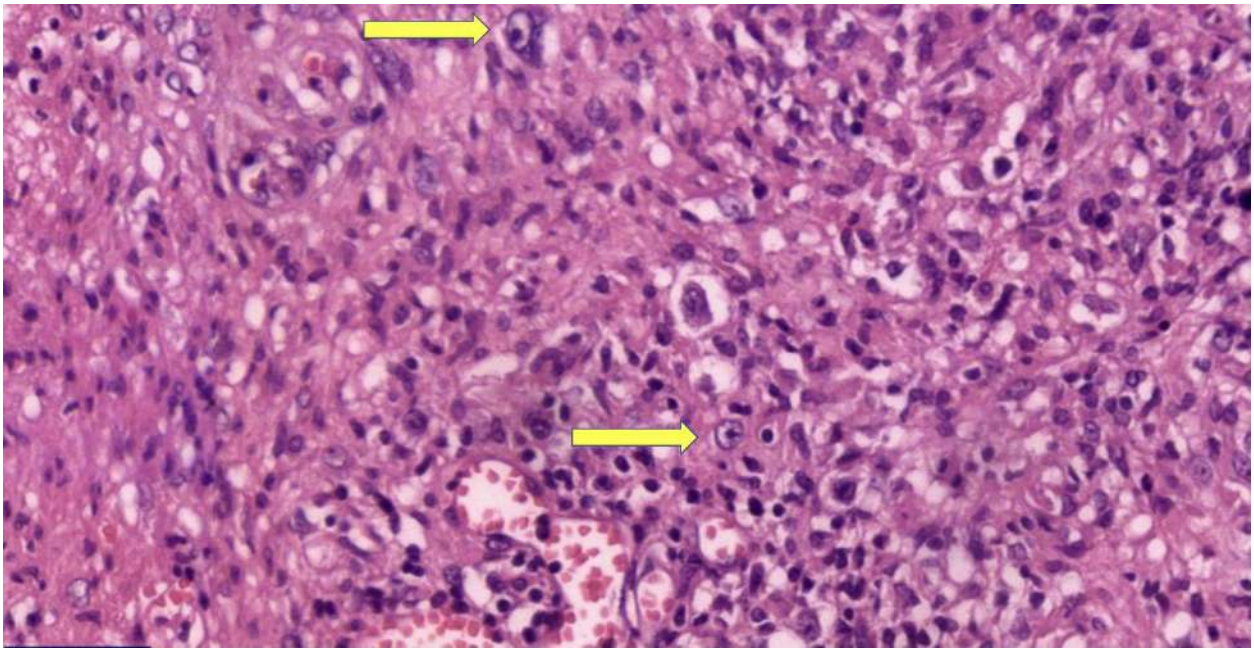




Microscopic examination of multiple sections show ulceration with dense band of polymorphous infiltrate composed of plasma cells, lymphocytes, histiocytes, immuno blasts, Hodgkin's like reed sternberg cells and apototic bodies. Areas of necrosis noted. For further diagnosis a panel of IHCs were suggested with the preliminary diagnosis of lymphoma.

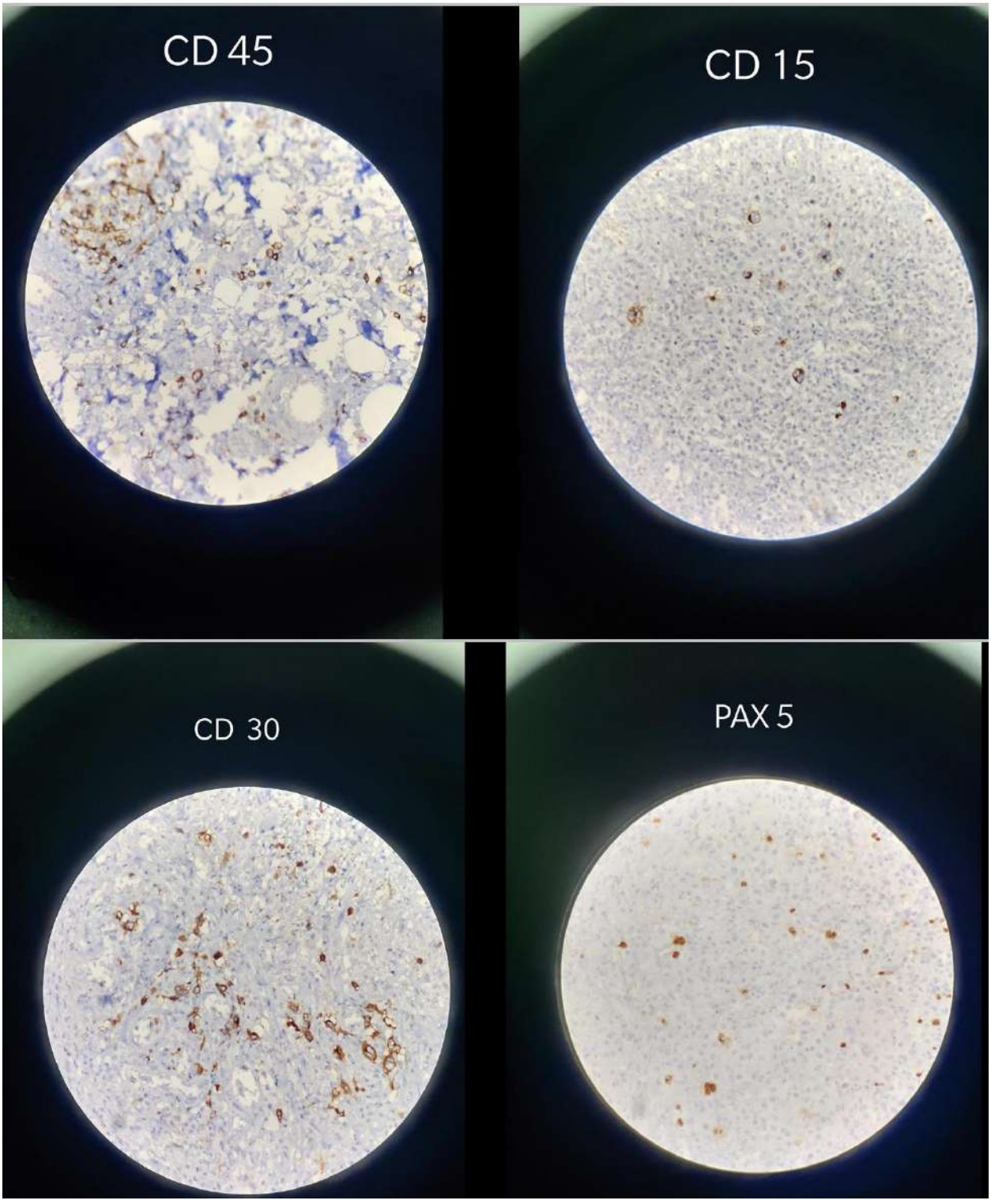


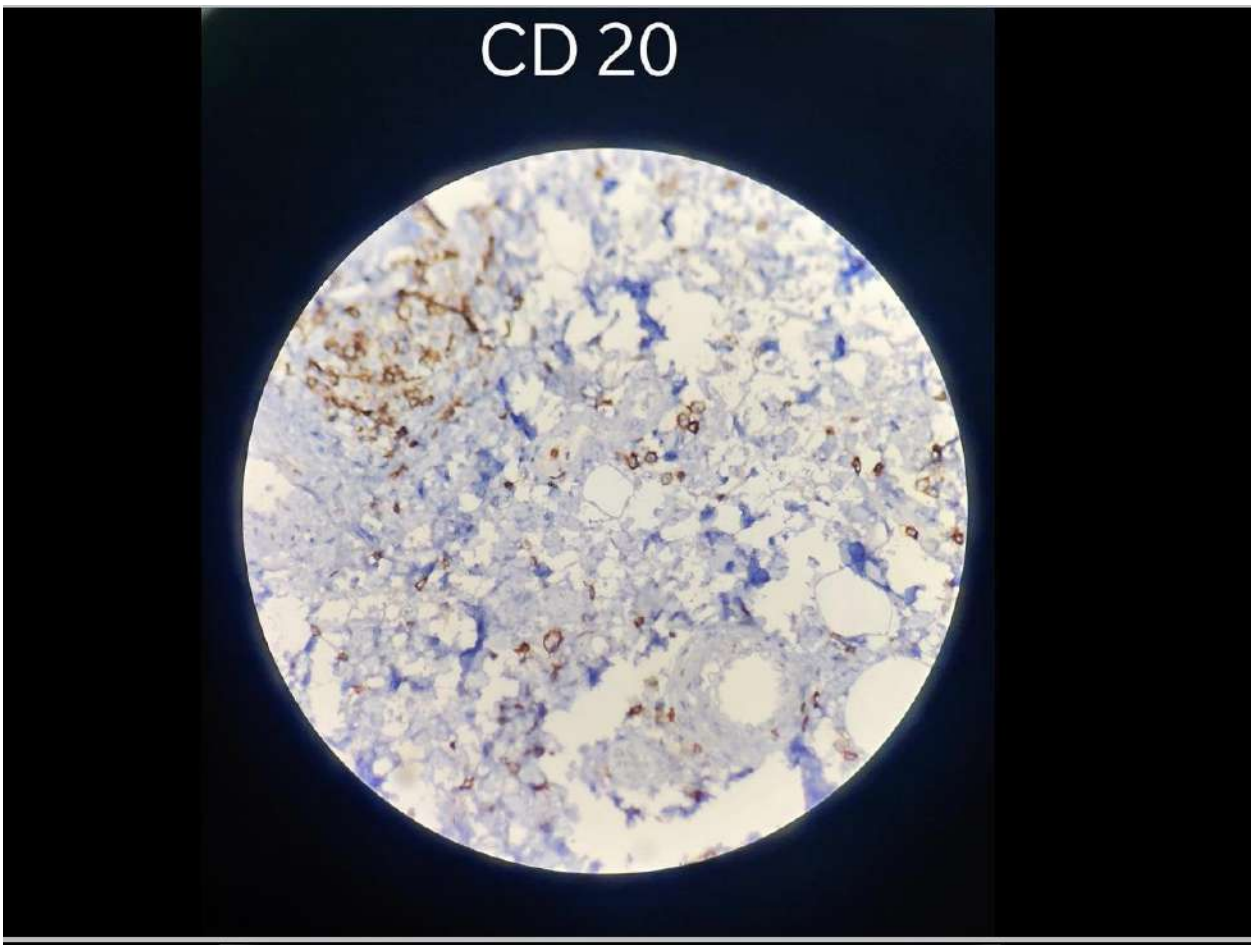
H& E shows: Atypical cells resembling Hodgkin Reed-Sternberg cells associated with polymorphous infiltrate of lymphocytes, histiocytes, plasma cells, and neutrophils (×40)

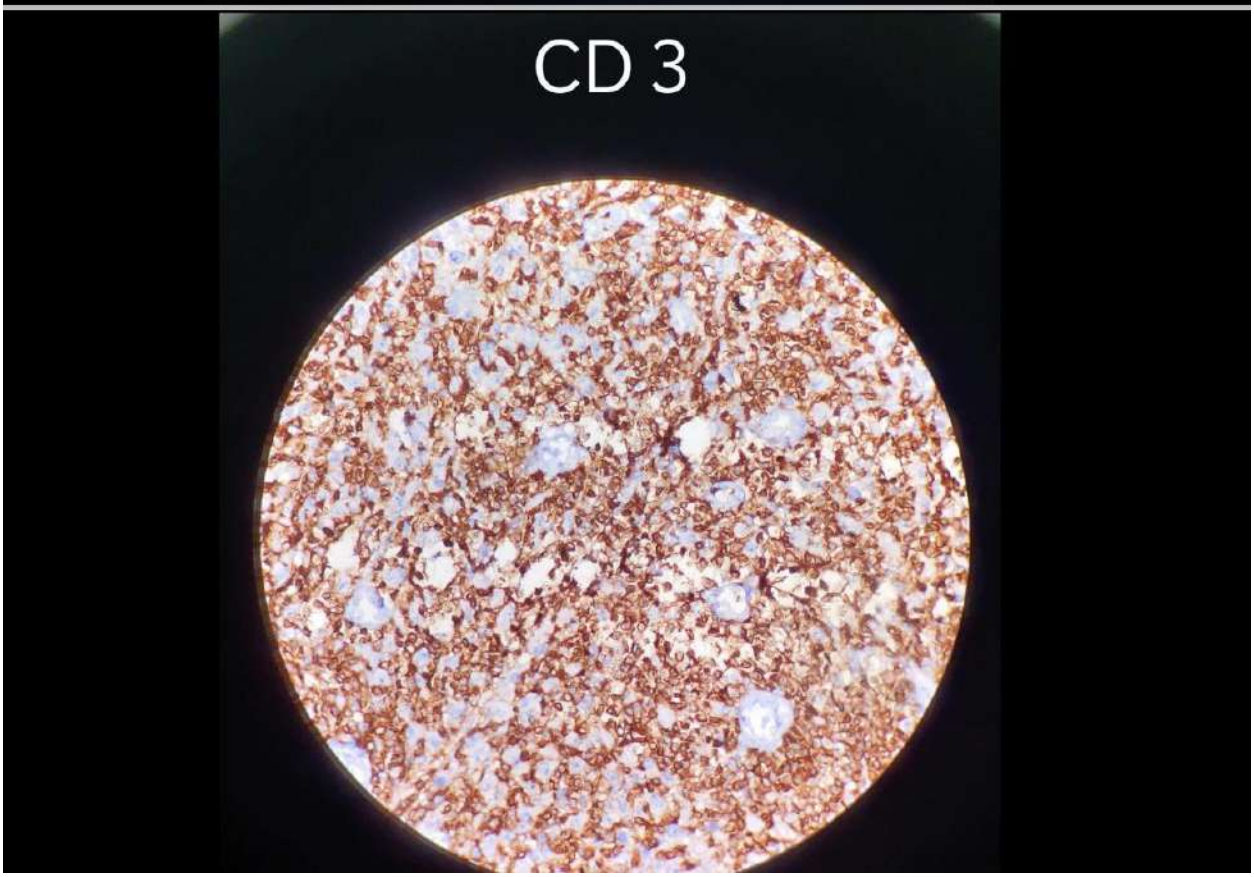


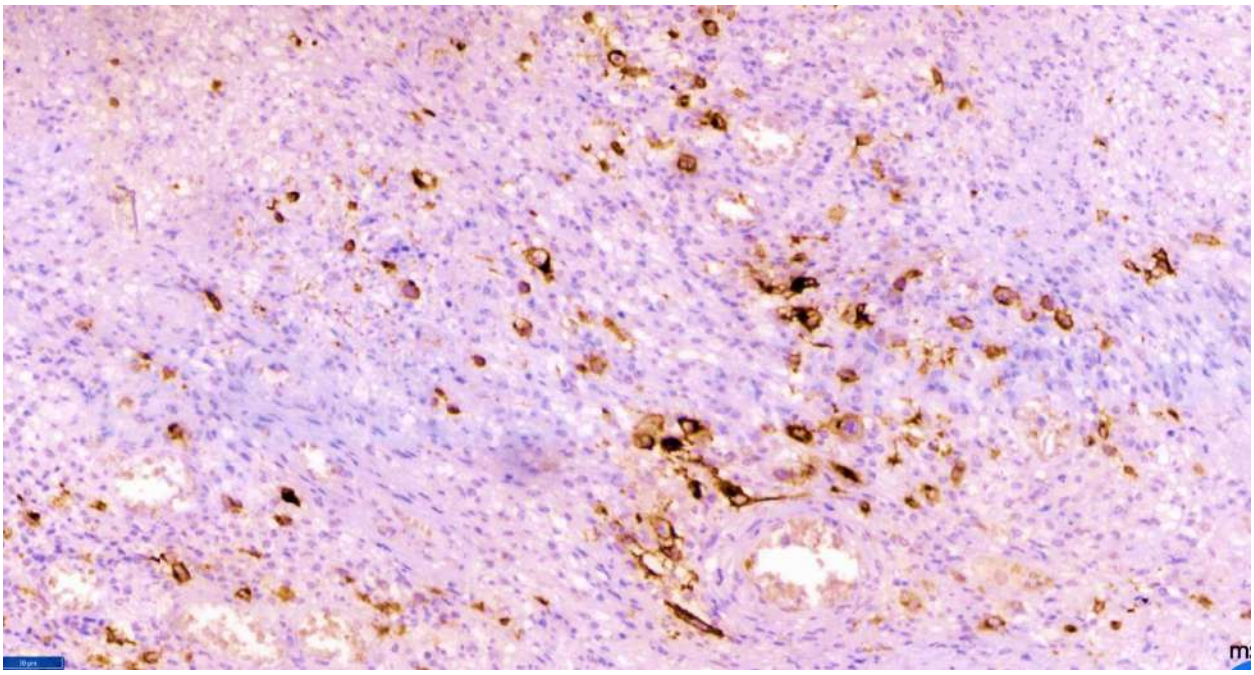
H&E : Atypical cells resembling Hodgkin Reed-Sternberg cells (yellow mark - x100)

Immunohistochemistry:




CD 20


CD 3



IHC for EBV showing positivity in atypical cells.

IHC showed that the large RS like cells showed positivity for EBV LMP, CD 15, CD30,CD45 , CD 20 and PAX 5. The surrounding T cells showed positivity for CD3. (40x)

Diagnosis:

H&E and IHC features favours a diagnosis of EBV positive Mucocutaneous ulcer.

Discussion:

Mucocutaneous ulcers positive for the Epstein Barr Virus (EBVMCU) are a newly recognized clinicopathological entity in the 2017 revision to the World Health Organization diagnostic criteria.This condition was described as a lymphoproliferative lesion associated with isolated skin or mucosal ulcers in elderly or immunosuppressed patients.EBV-MCU is associated with multiple etiologies, including primary immunodeficiency, HIV infection, post-transplantation , immunosenescence owing to aging and iatrogenic causes, such as methotrexate and TNF-alfa antagonists.Usually these present as localized sharply circumscribed ulcerative lesions, typically solitary (83%), that can occur in the oropharynx (52%), skin (29%) or gastrointestinal tract (19% - 40% colon, 30% esophagus, 20% rectum and 10% terminal ileum).

The differential diagnosis of EBVMCU includes secondary involvement by Classic Hodgkin Lymphoma (cHL), diffuse large B-cell lymphoma (DLBCL) associated or not with EBV, primary cutaneous anaplastic large cell lymphoma, lymphomatous granulomatosis (LyG) and aggressive Posttransplant Lymphoproliferative Disease (PTLD).

Typically, EBVMCU presents an indolent course. While no treatment guidelines exist, management tends to be conservative and, in cases related to the use of immunosuppressants, the withdrawal or decrease in immunosuppressant dosages is followed .

EBVMCU is a rare newly recognized entity. Clinical history, pathological examination, and immunohistochemistry are necessary for a definitive diagnosis .Multidisciplinary approach is mandatory.