

Hotel Senec, Senecké jazerá

25.05.2018



Slovenská spoločnosť patologov SLS
Slovenská divízia medzinárodnej akadémie patológie (SD IAP)
Cytopathos spol. s r.o. a Ústav patológie SZU Bratislava

usporiadajú v dňoch 25.- 26. mája 2018 v Senci

LETNÝ BIOPTICKÝ SEMINÁR SD IAP

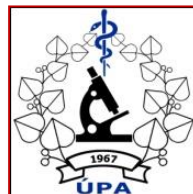
Prípad SD-IAP č. 682

Lukáš Plank

Národné konzultačné centrum bioptickej diagnostiky

lymfoidných a myeloidných nádorov

ÚPA JLF UK a UN v Martine





Prípád č. 682 = Konzultačná biopsia

Dáta dostupné pri konzultácii:

a) Údaje uvedené chirurgom na žiadanke k biopsii:

Muž, 66 r, resekcia tenkého čreva – jejuna,
Akútny chirurgický zákrok – pneumoperitoneum
Pri revízii zistená perforácia čreva, pribl. 2 m od lig. Treitzi.

Iné údaje neuvedené/neznáme.

b) Údaje uvedené odosielajúcim patológom:

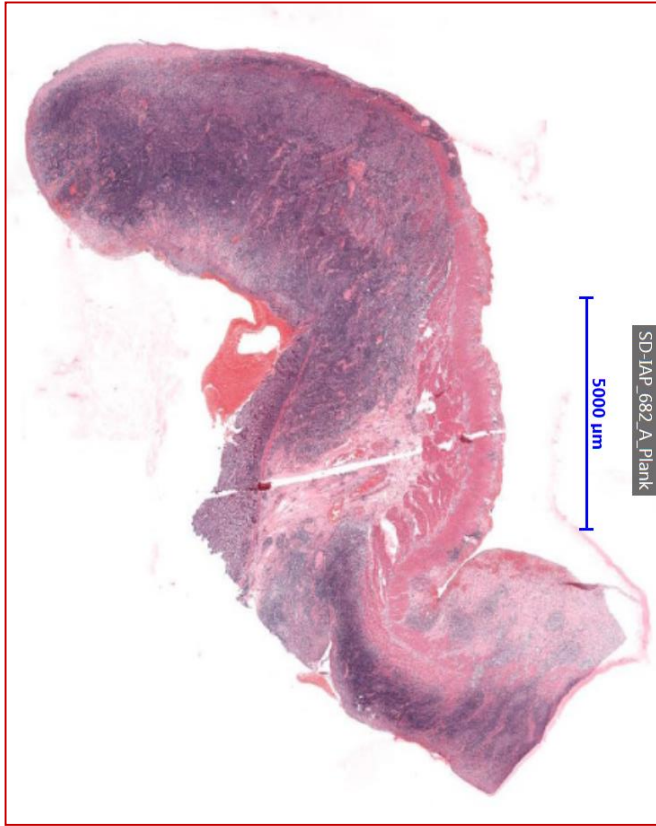
Naša pracovná dg.: T-NHL asociovaný s enteropatiou, v jejúne, s perforáciou čreva a fibrinózne-purulentnou peritonitídou.

„Materiál poslaný prof. Plankovi na konzultáciu“, 3 bloky + 3 HE preparáty.

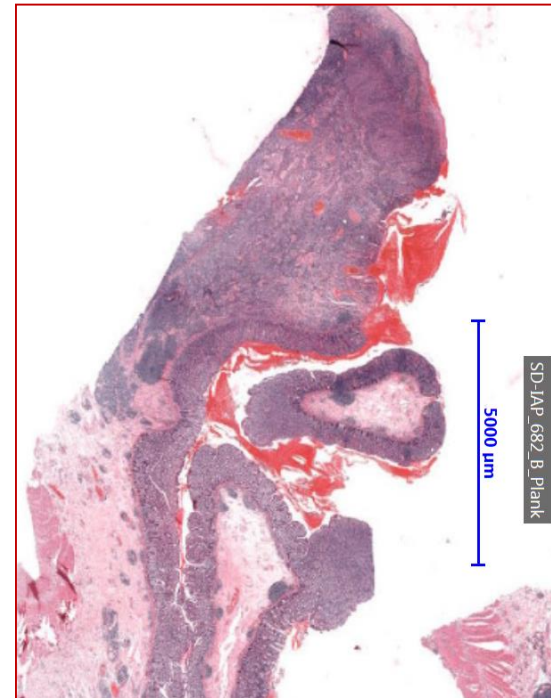


2 HE preparáty

A

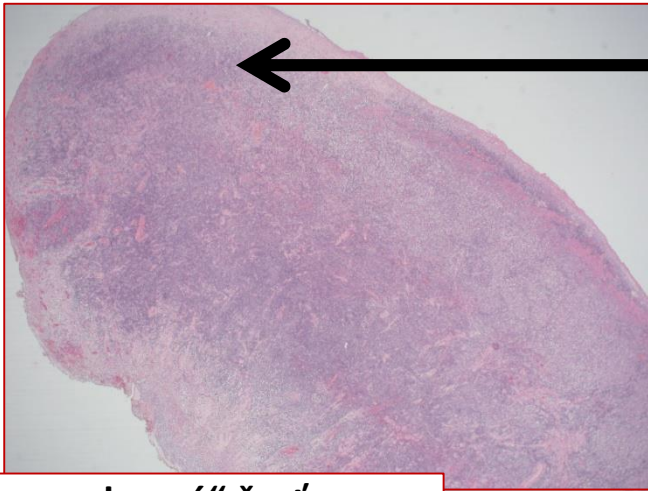


B

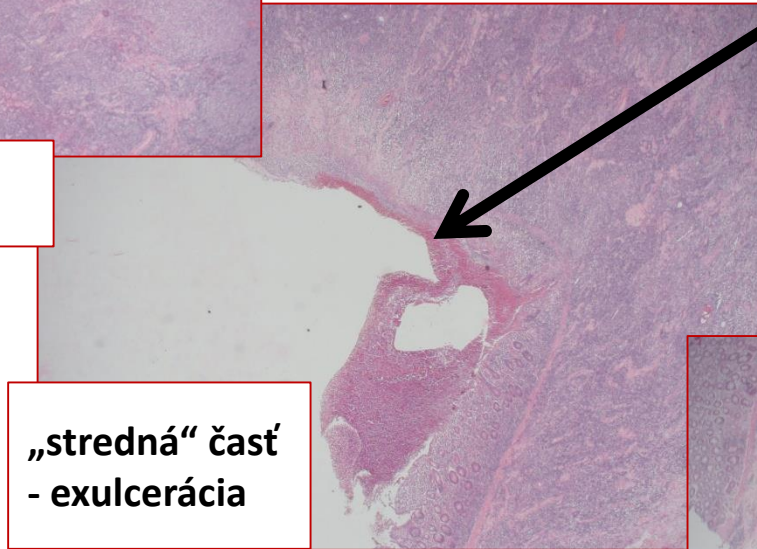




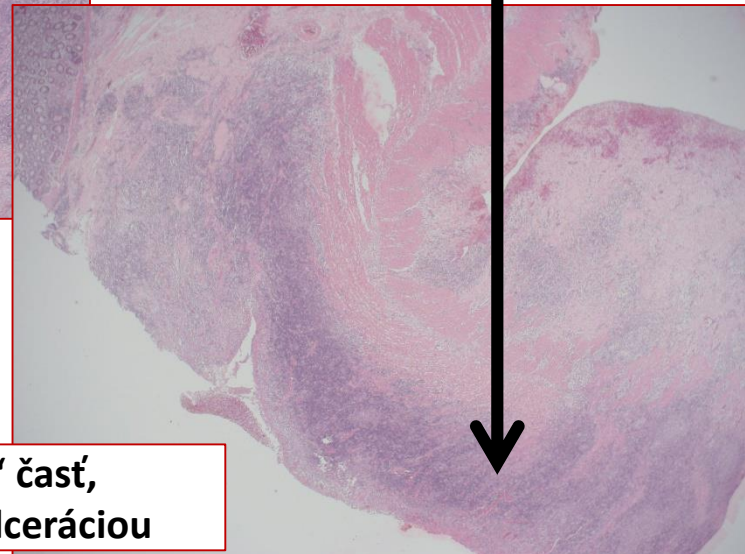
1. HE (A)



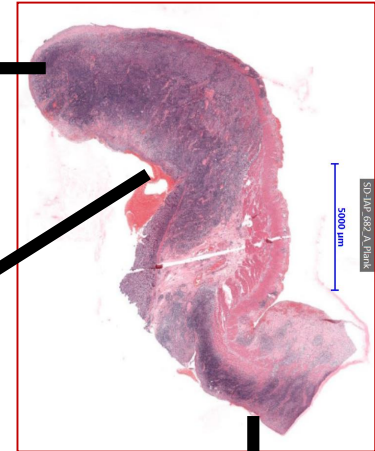
„horná“ časť,
„nad“ exulceráciou



„stredná“ časť
- exulcerácia



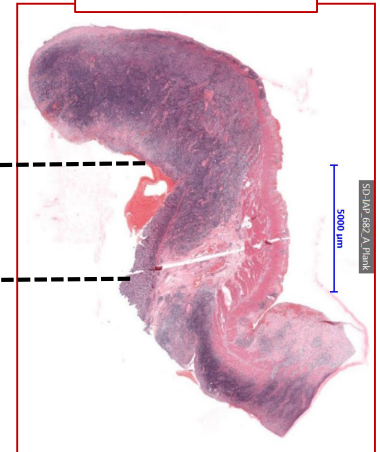
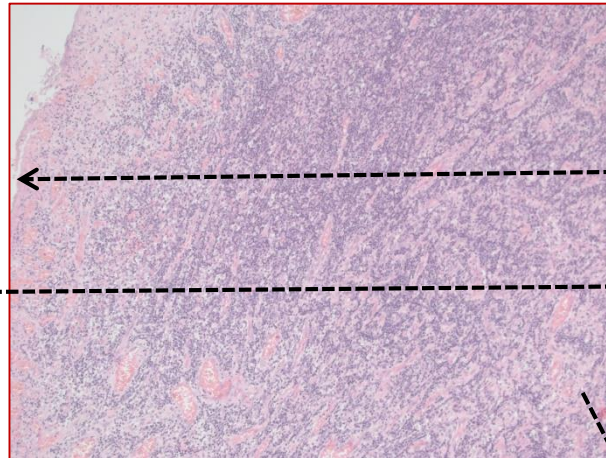
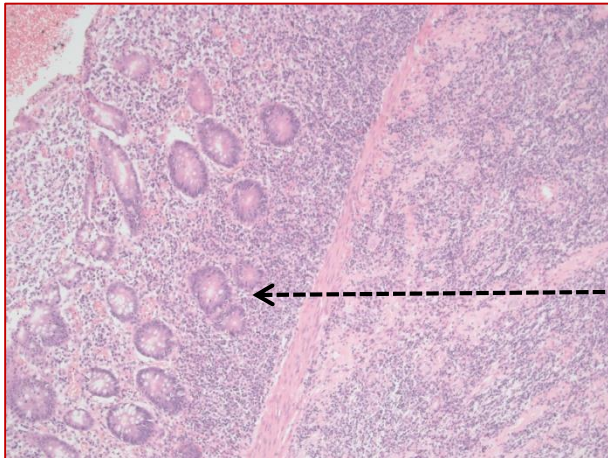
„dolná“ časť,
„pod“ exulceráciou



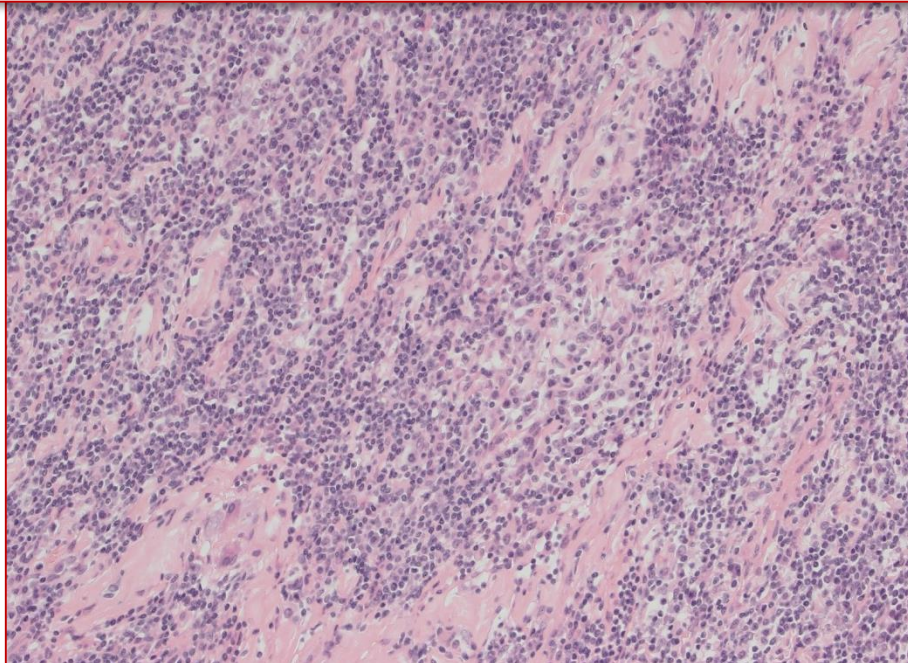


1. HE (A) - „stredná časť“

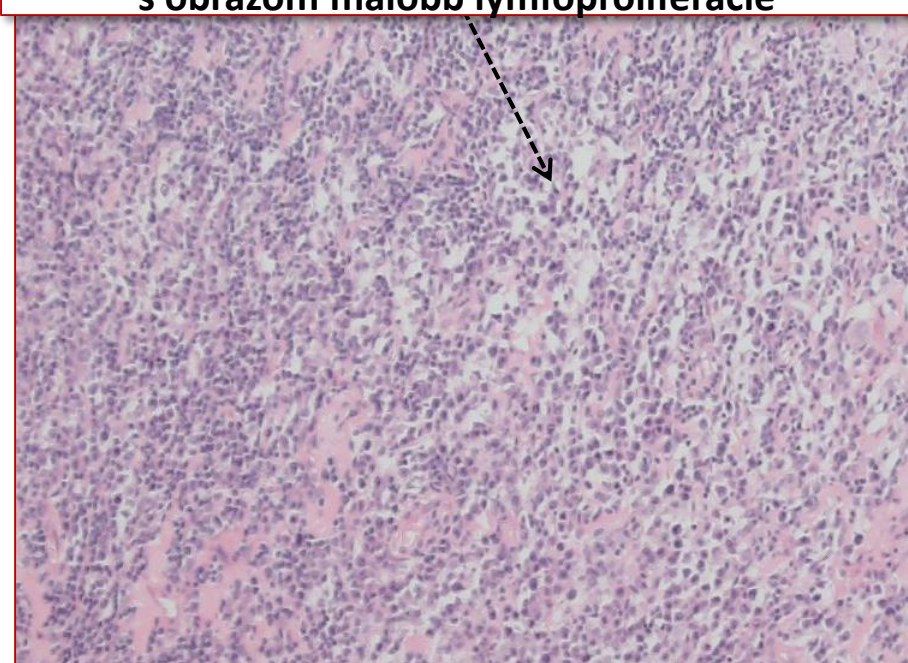
Stredná časť
- exulcerácia



„solídna“, predominantne malobb proliferácia

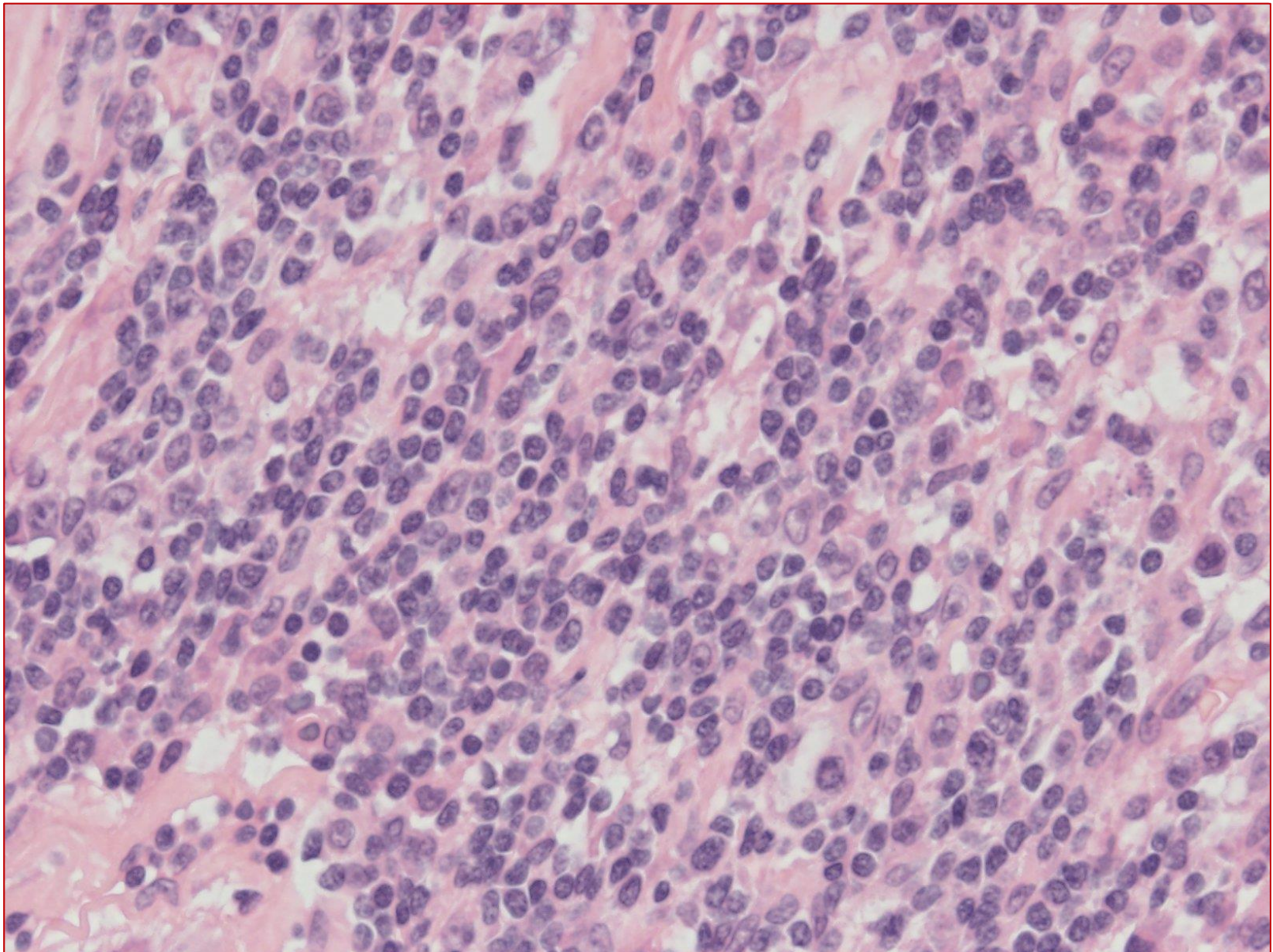


viac PM, „svetlobb“vzhľad, premiešané s obrazom malobb lymfoproliferácie



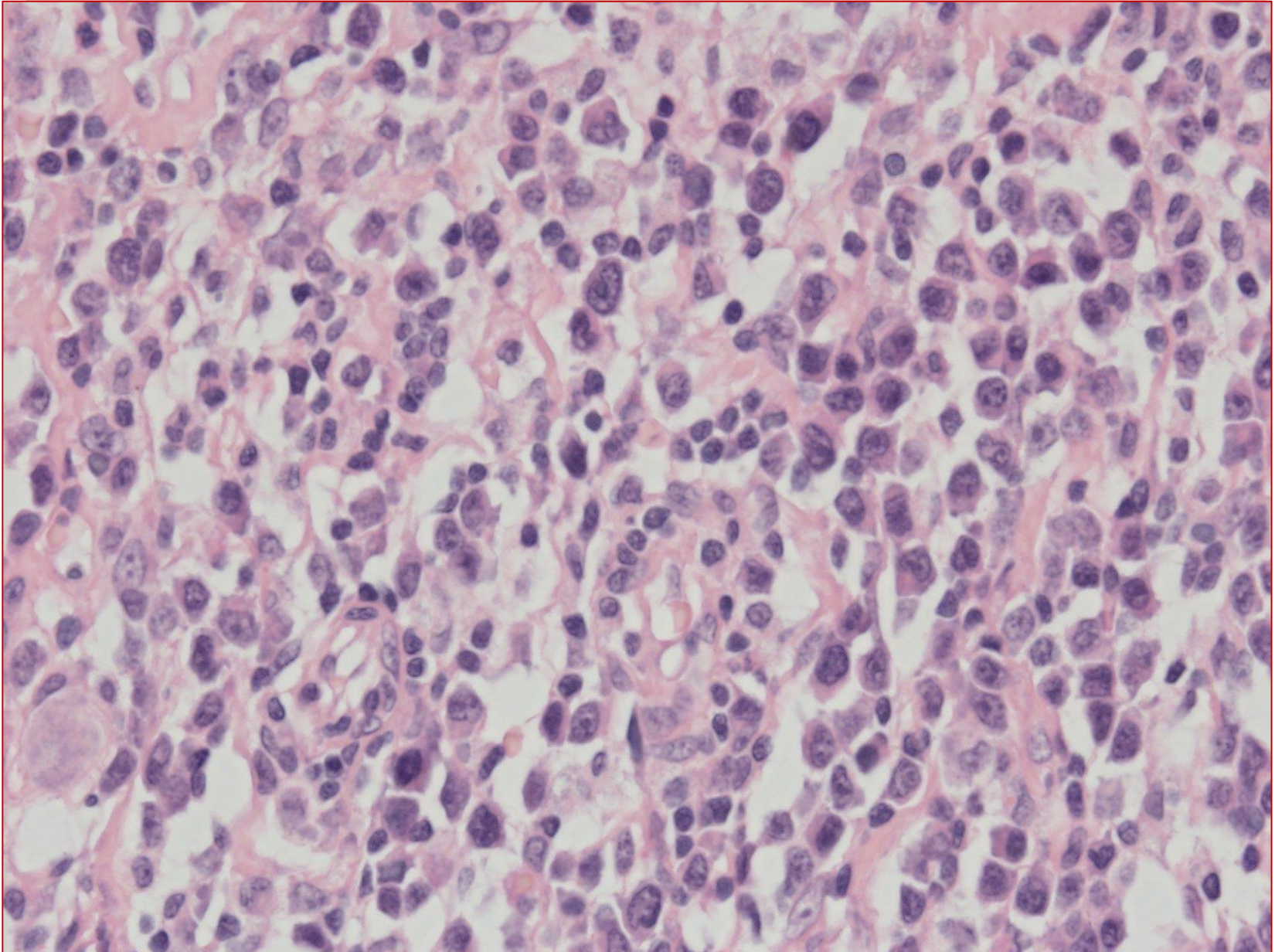


1. HE (A) - „stredná časť“





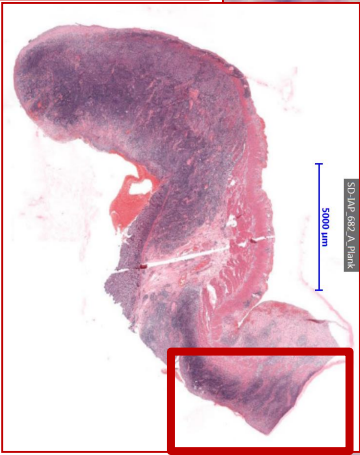
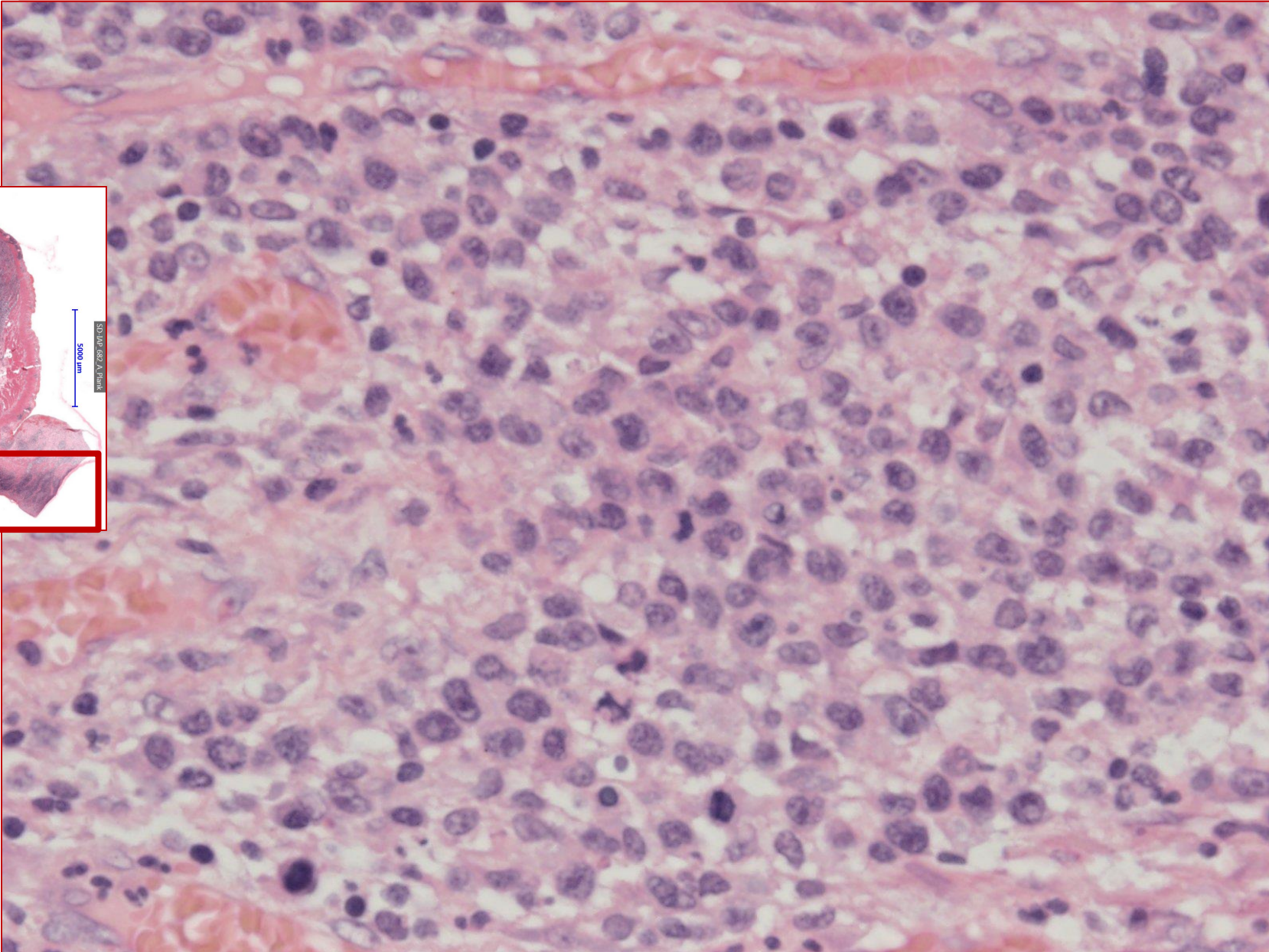
1. HE (A) - „stredná časť“





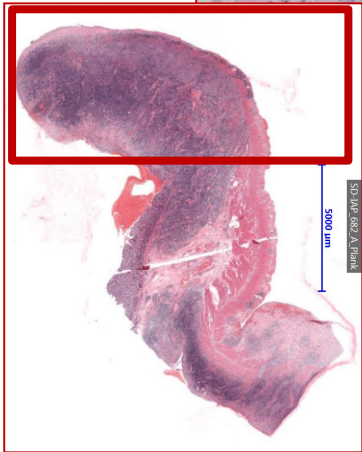
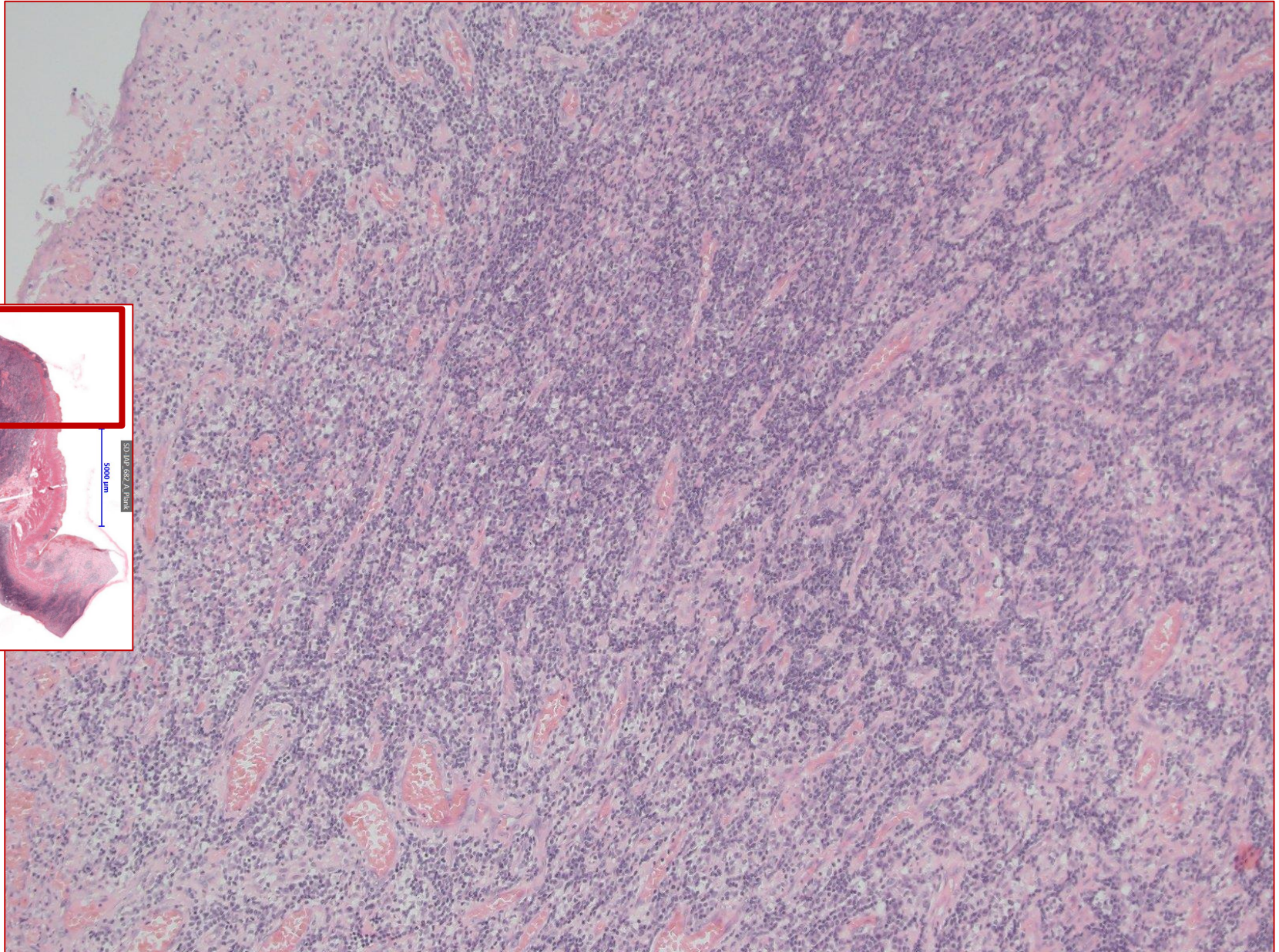
1. HE (A): „dolná“ časť – ešte viac PM

„dolná“ časť,
„pod“
exulceráciou





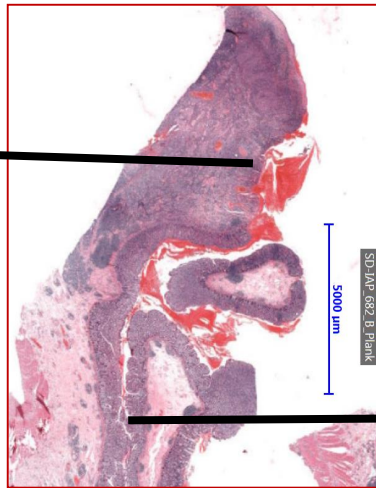
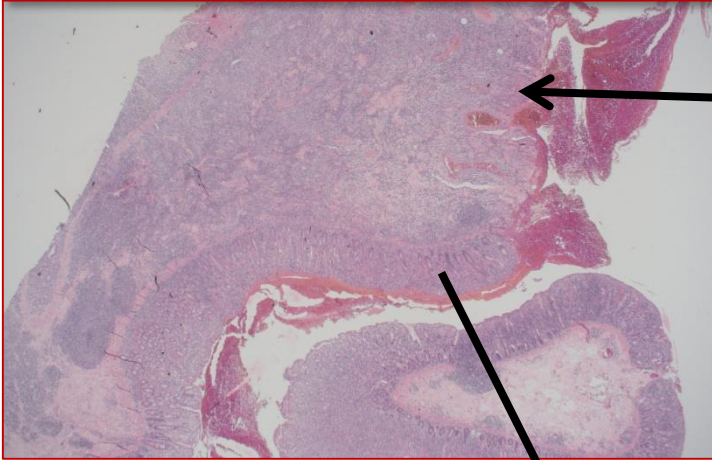
1. HE (A): „horná“ část



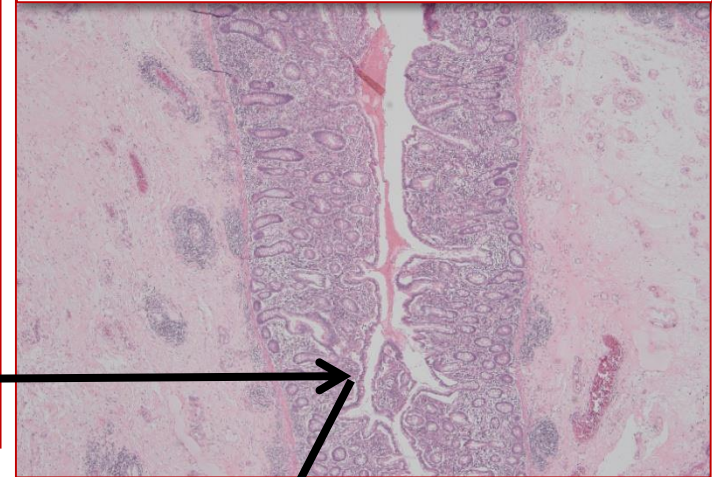


2. HE (B)

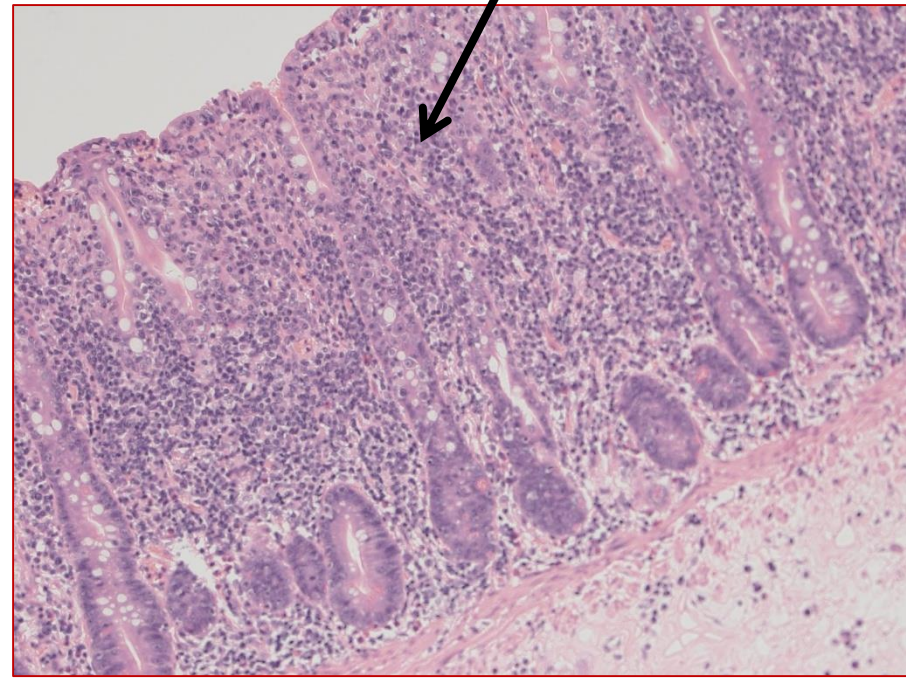
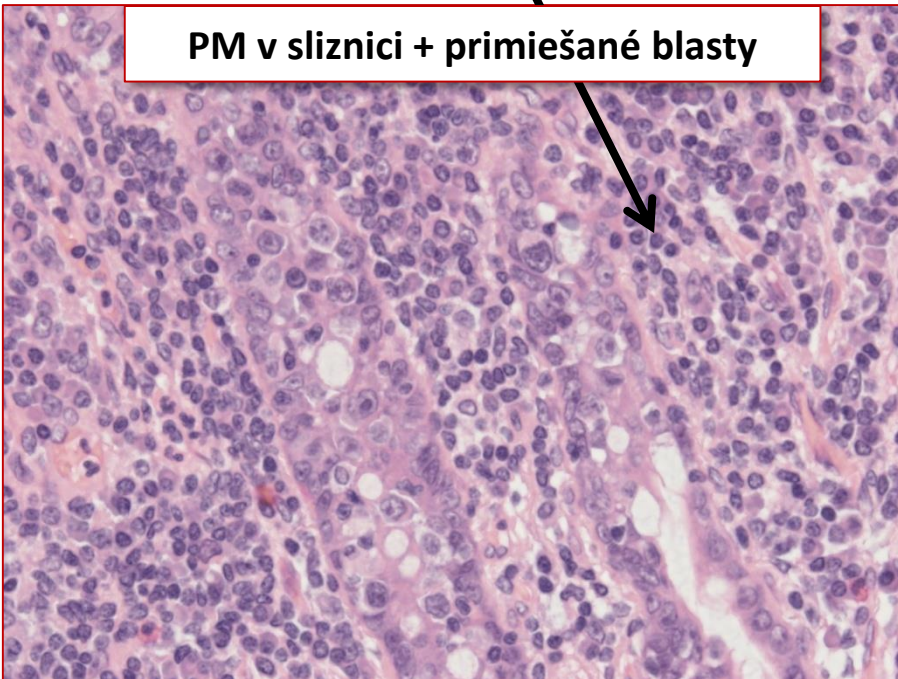
Predominantne malobb proliferácia



Sliznica bez ML infiltrácie



PM v sliznici + primiešané blasty





Takže čo ?

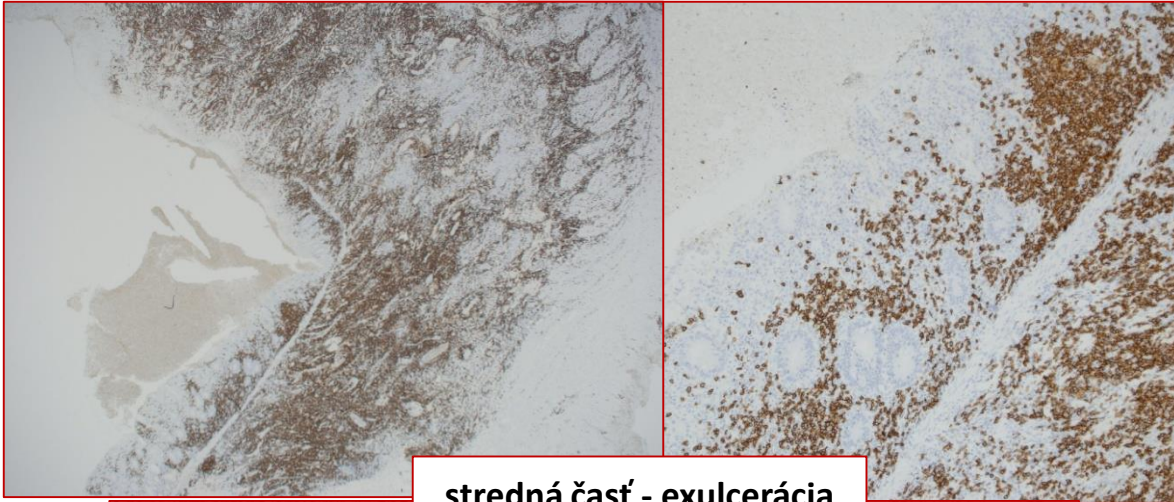
Máte návrhy na Dg. ?



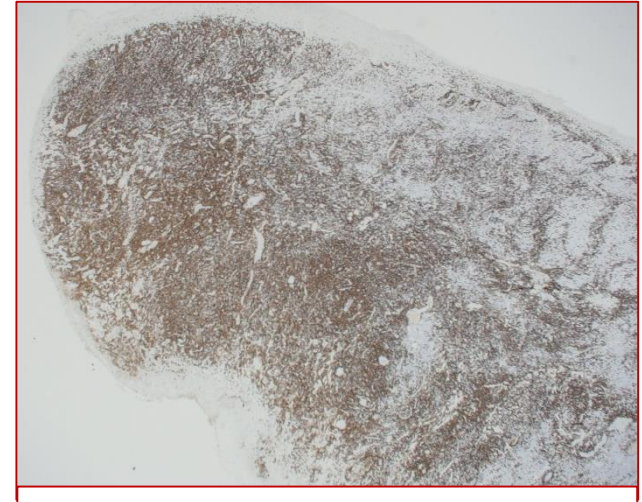
O.K., IHC ...



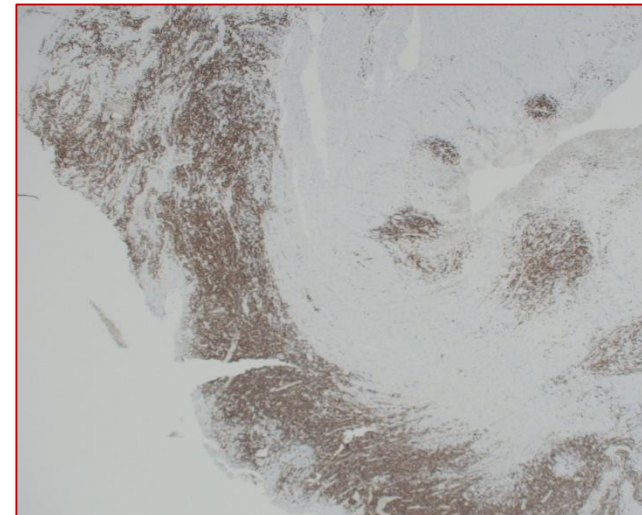
CD20 „masa“ v rôznych častiach



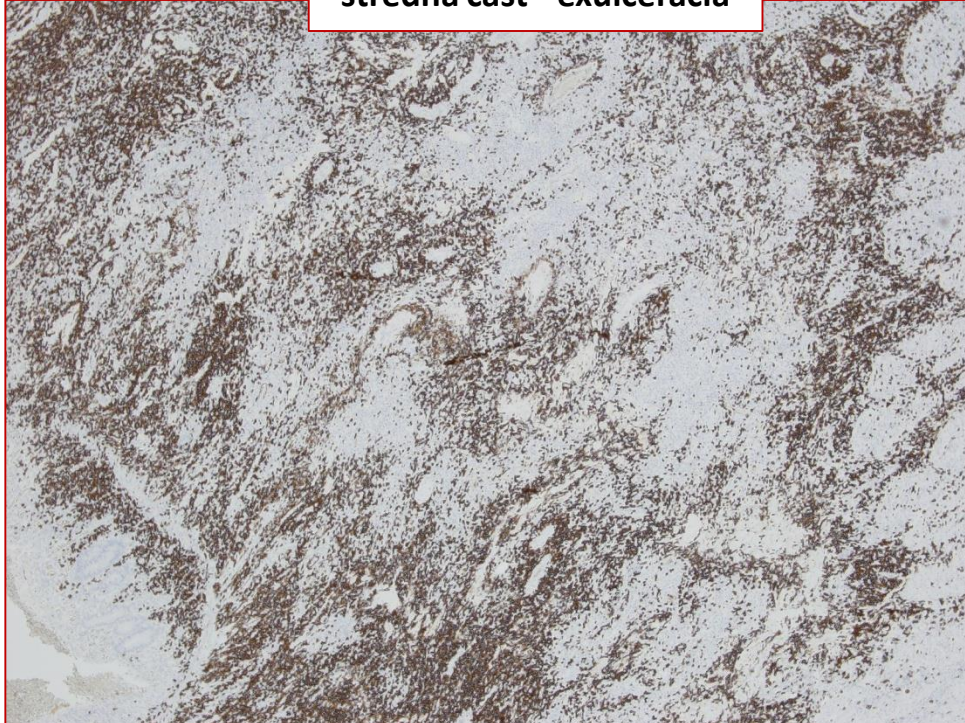
stredná časť - exulcerácia



„horná“ časť, „nad“ exulceráciou



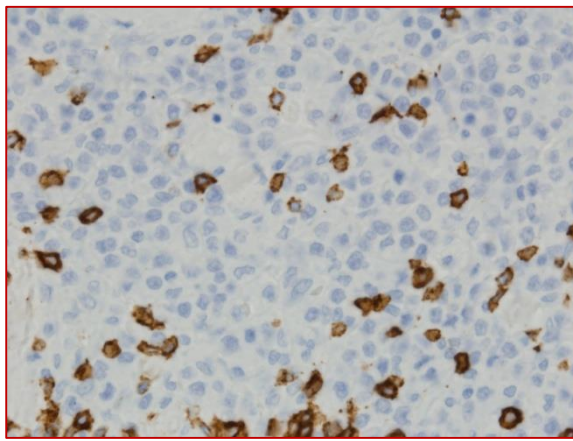
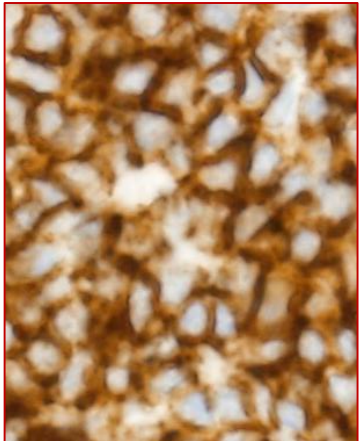
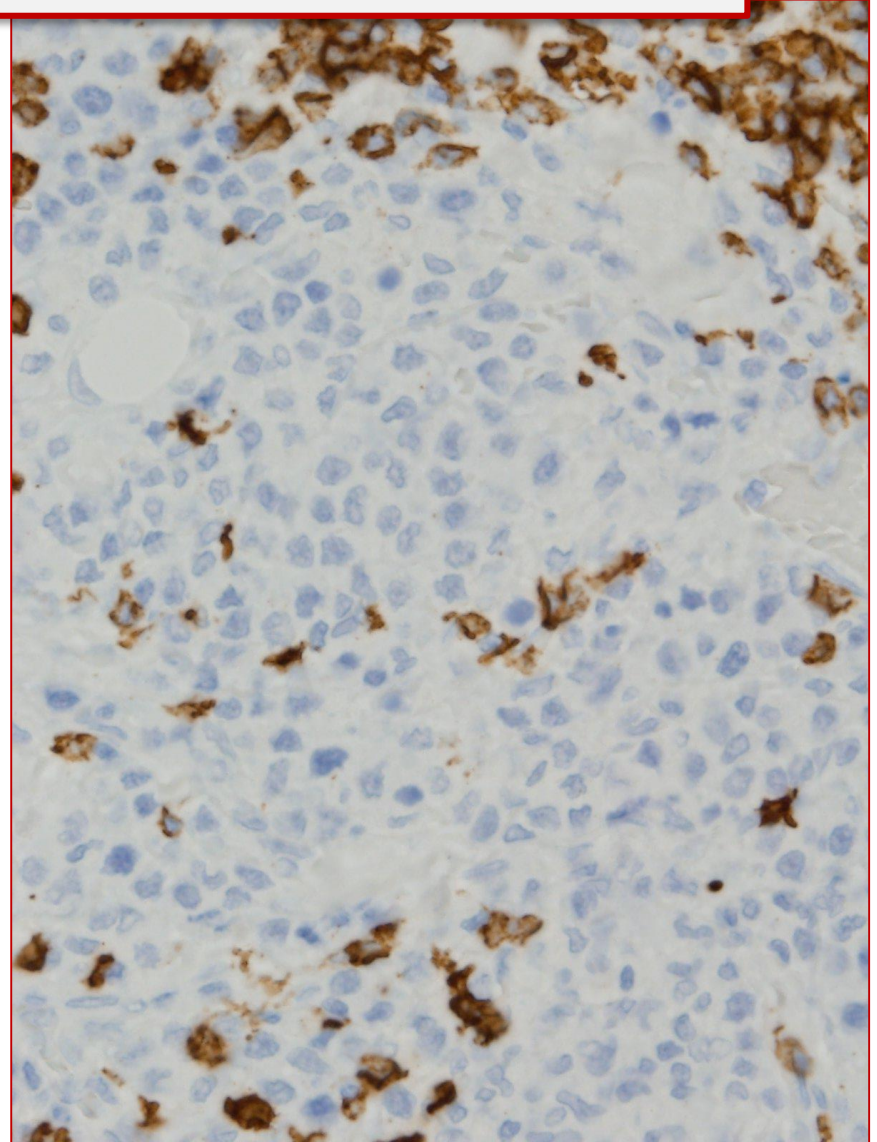
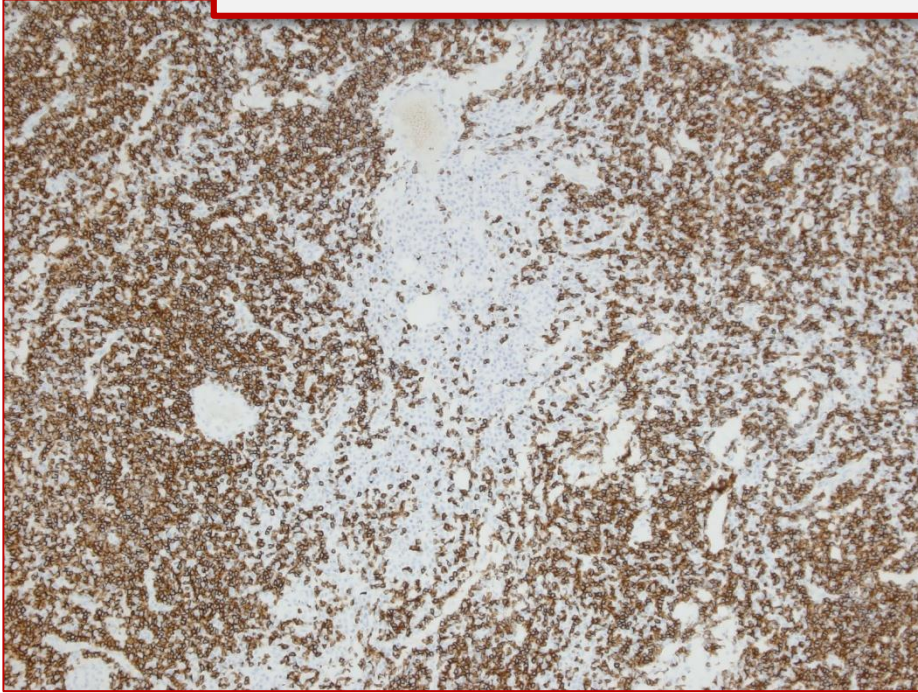
„dolná“ časť, „pod“ exulceráciou





CD20 v strednej časti

CD20+ „masa“ a CD20- PM oblasť v hlbšie intramurálne v stene

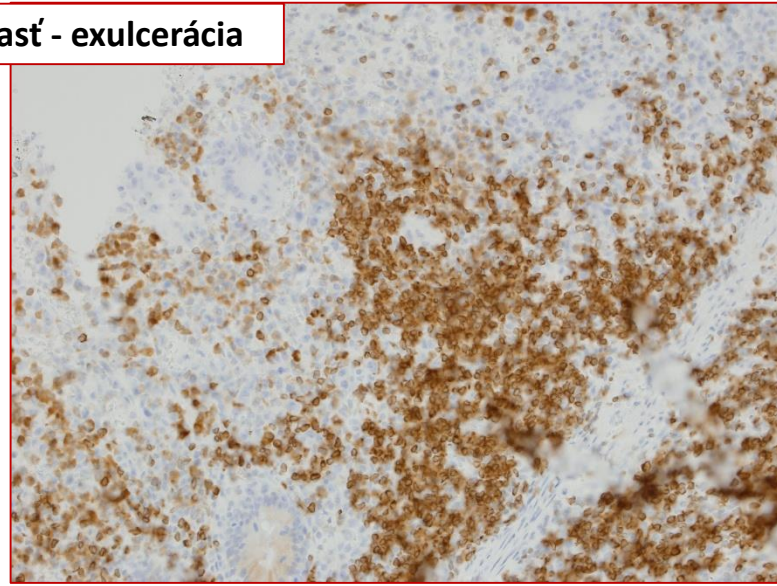




CD20+ oblasti sú bcl2 pozitívne



stredná časť - exulcerácia



„horná“ časť, „nad“ exulceráciou

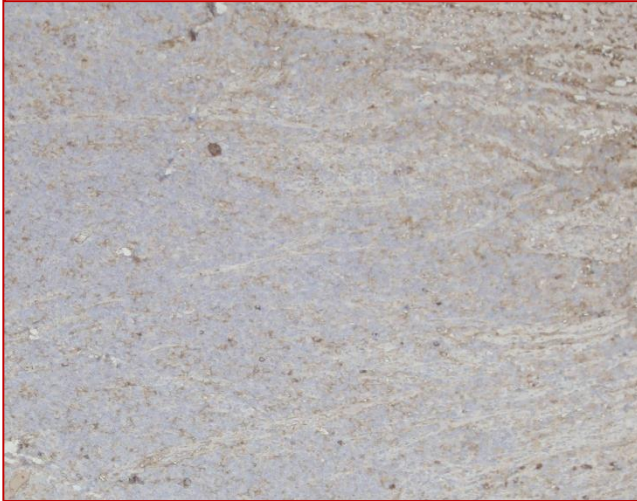


„dolná“ časť, „pod“ exulceráciou

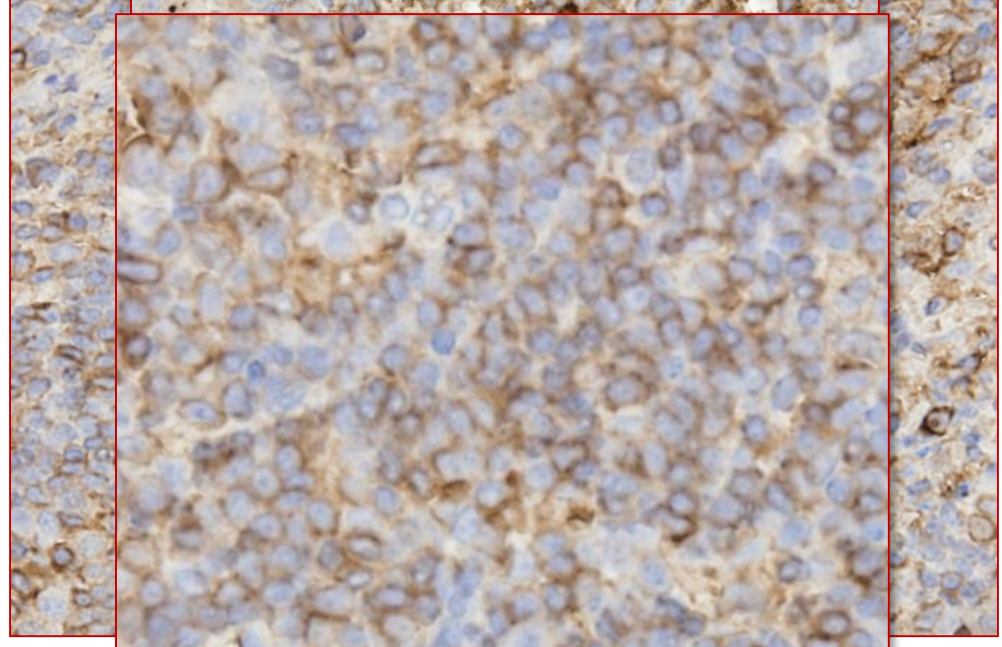


CD20+ oblasti: monoklon. plasmacytoidná diferenciácia

κ negativita



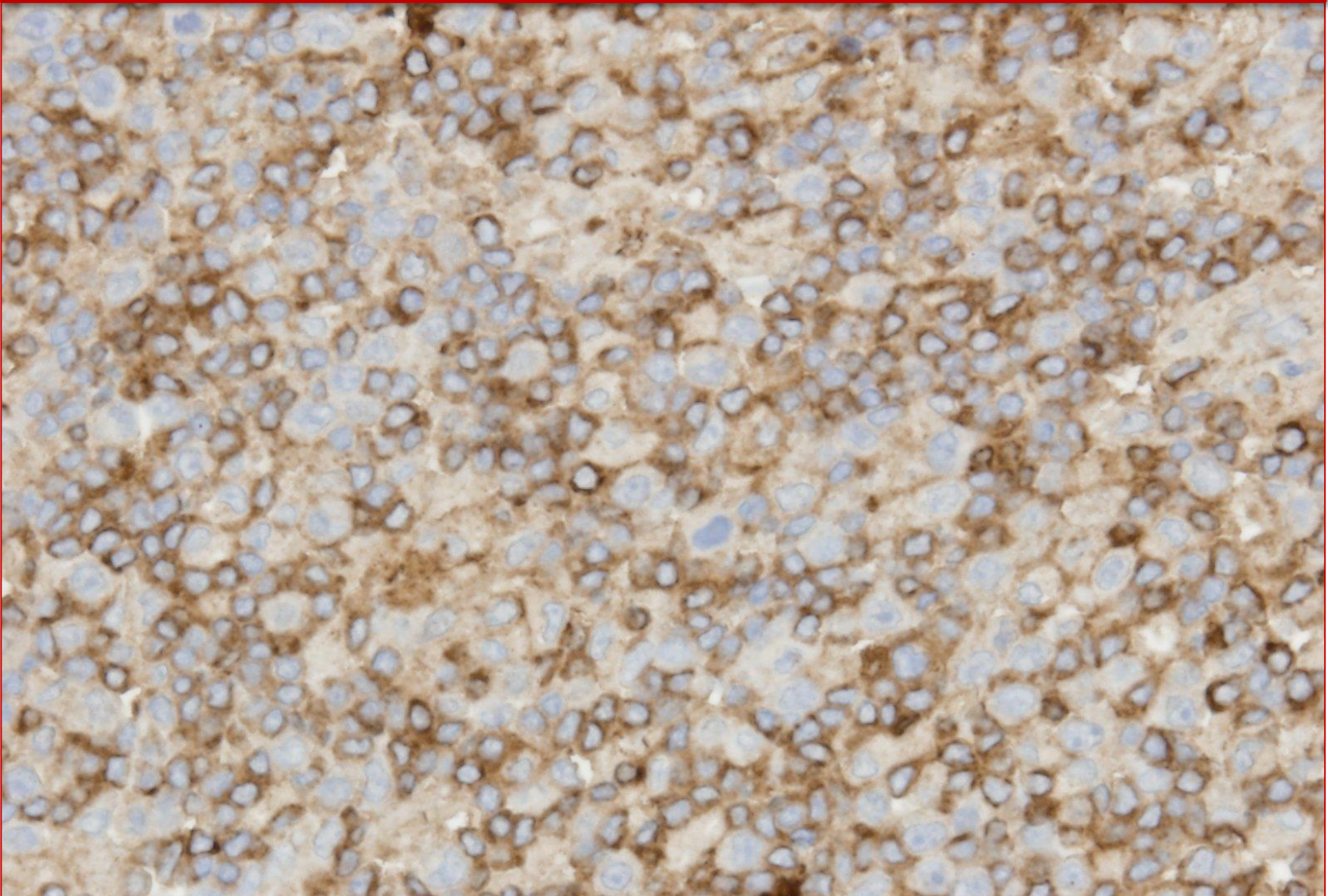
λ pozitivita





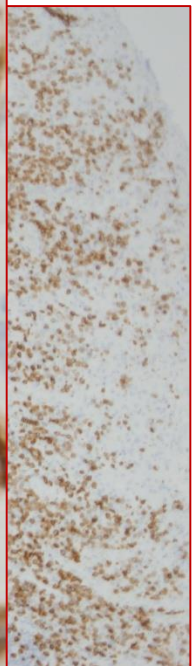
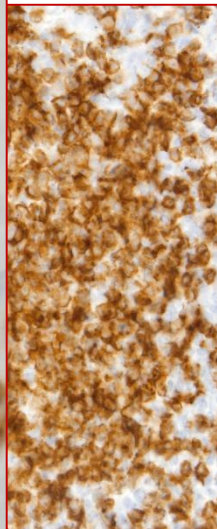
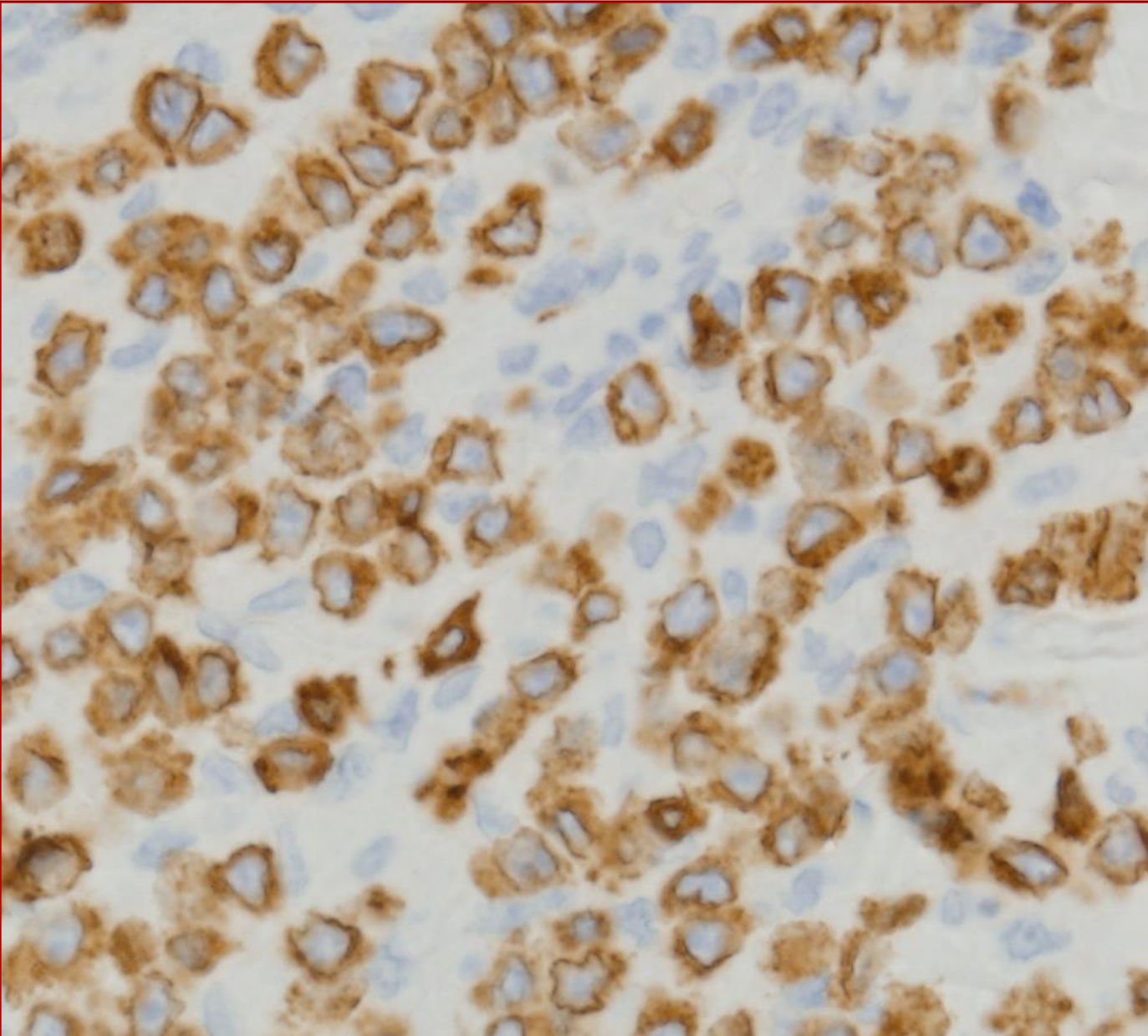
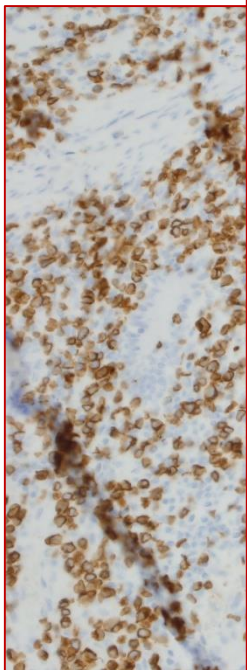
CD20+ oblasť a monoklon. pcoid diferenciácia

λ + malobb B-populácia premiešaná s PM CD20- veľkobunkovou populáciou



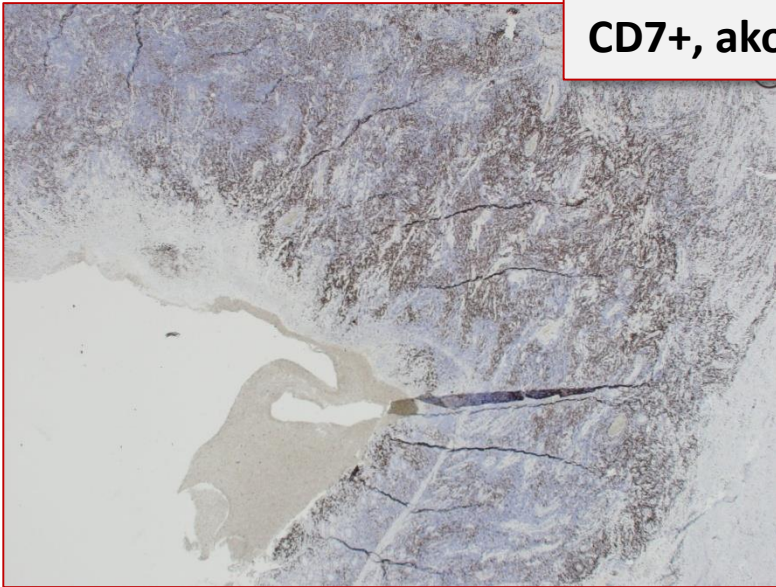


CD3+ v rôznych častiach, pozit. PM @ väčších bb („blastov“)

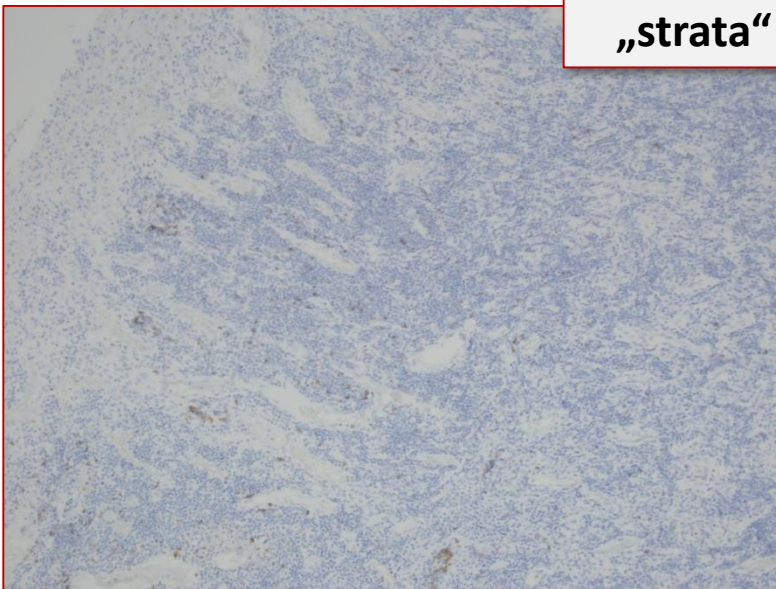
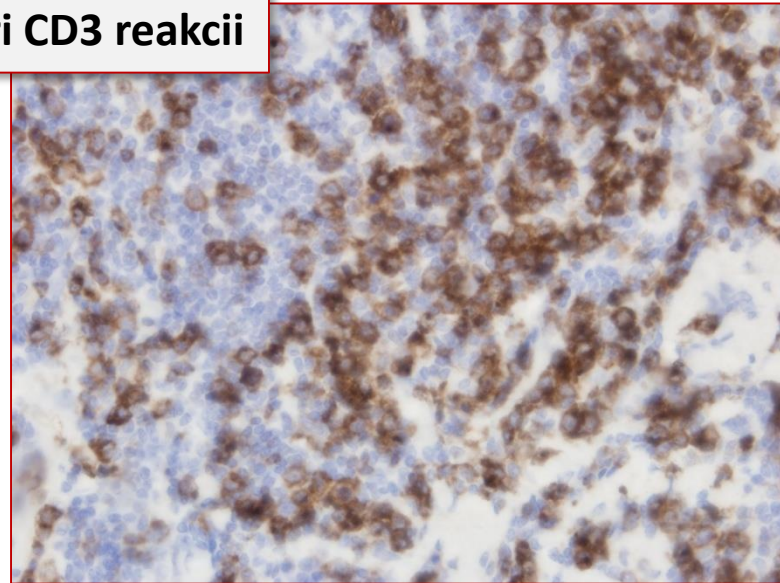




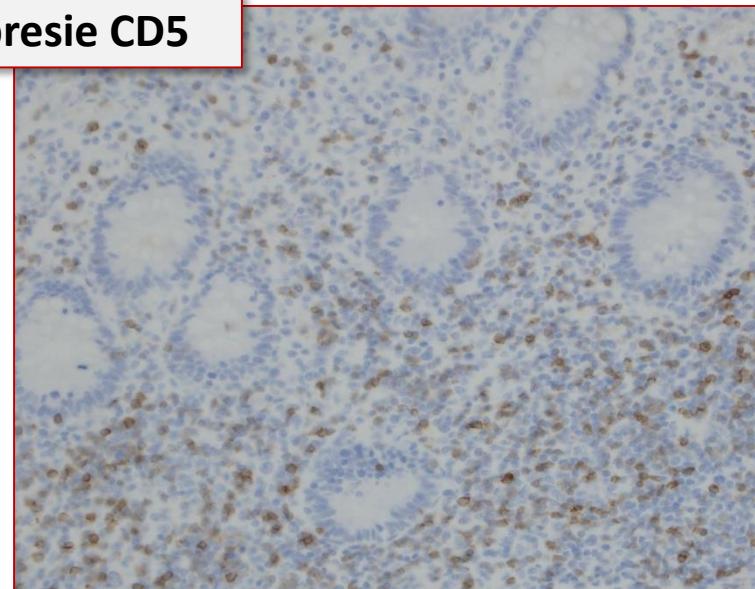
Iné T-bb znaky v rôznych častiach



CD7+, ako pri CD3 reakcii



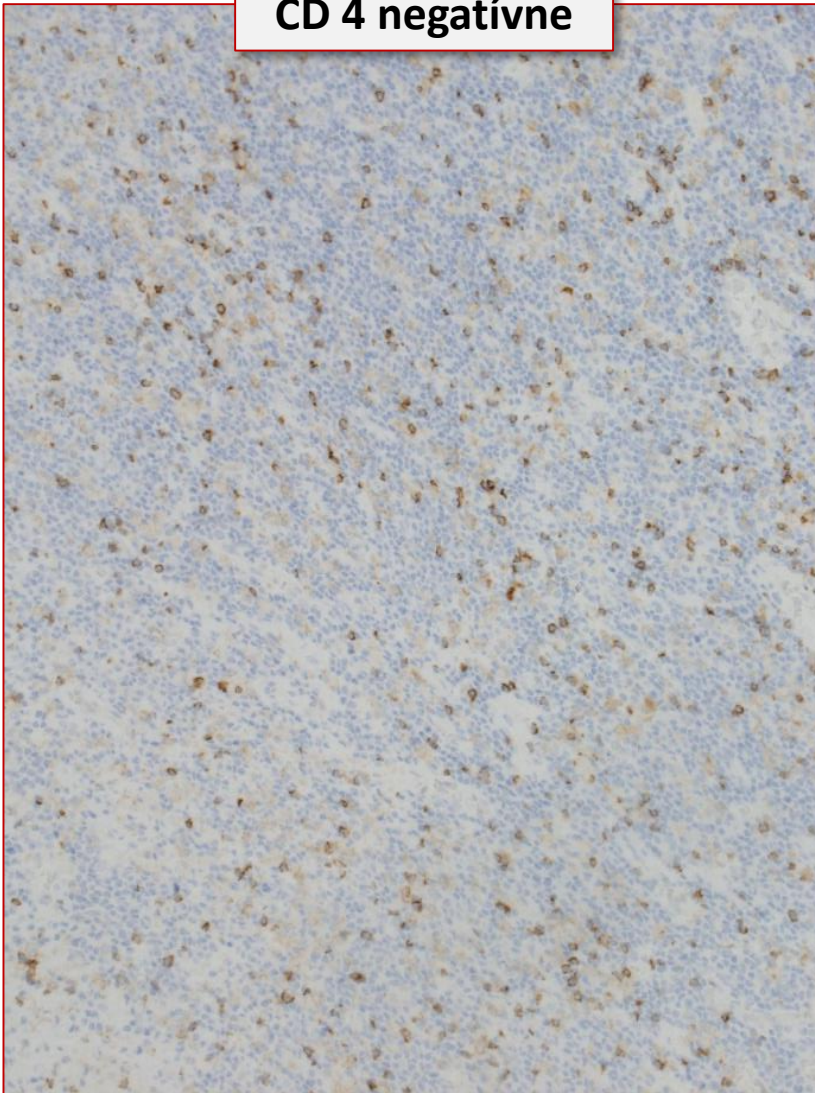
„strata“ expresie CD5



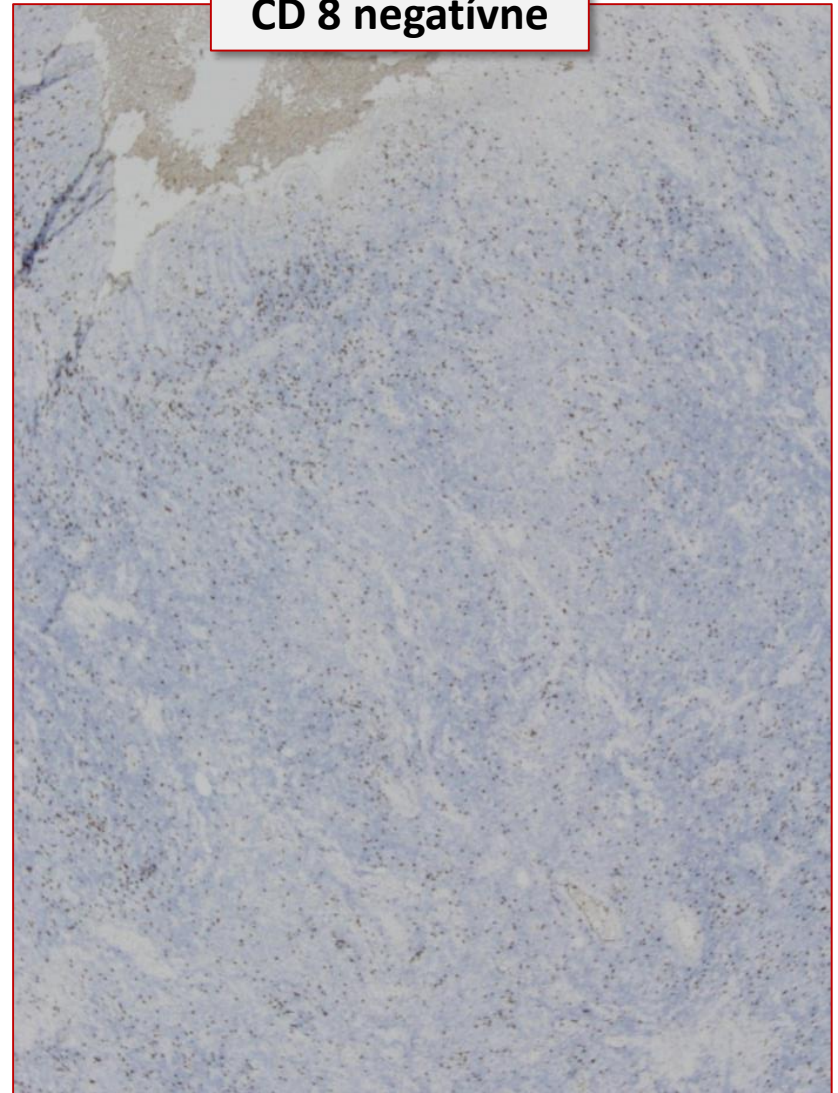


Iné T-bb znaky v rôznych častiach

CD 4 negatívne

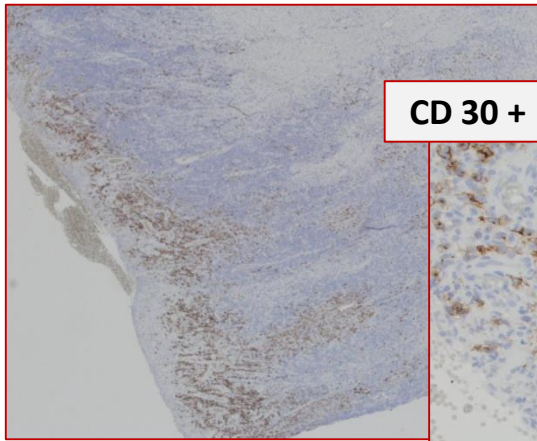


CD 8 negatívne

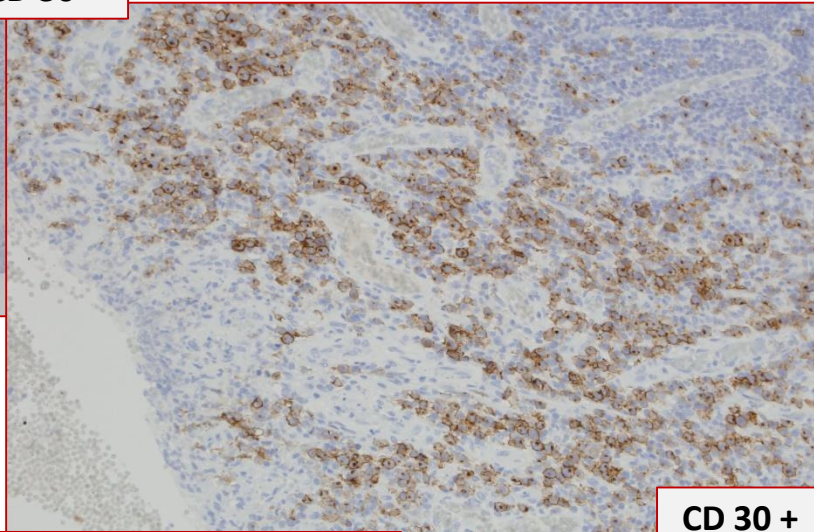




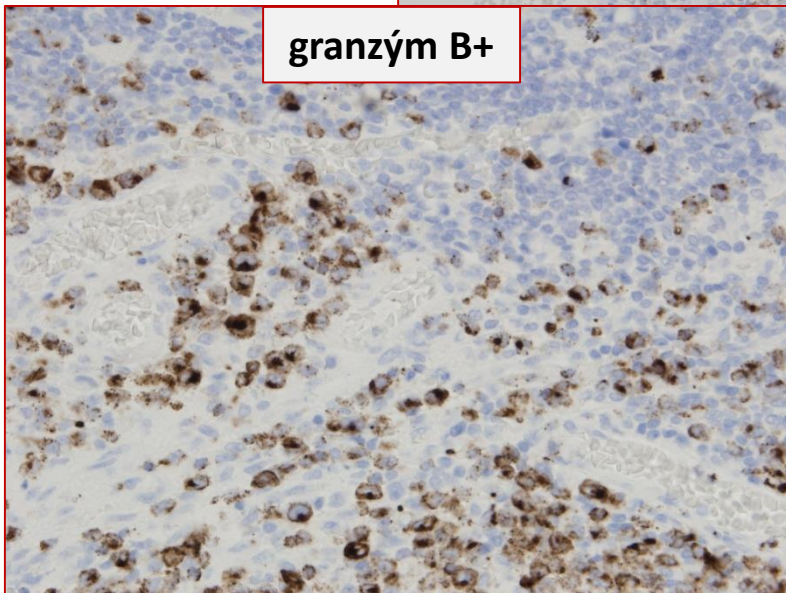
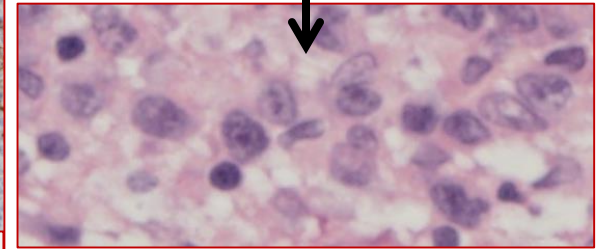
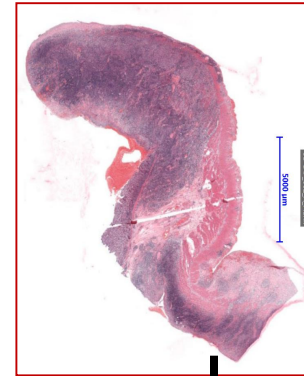
CD30+ vo veľkobunkovej T-populácii v „dolnej“ časti



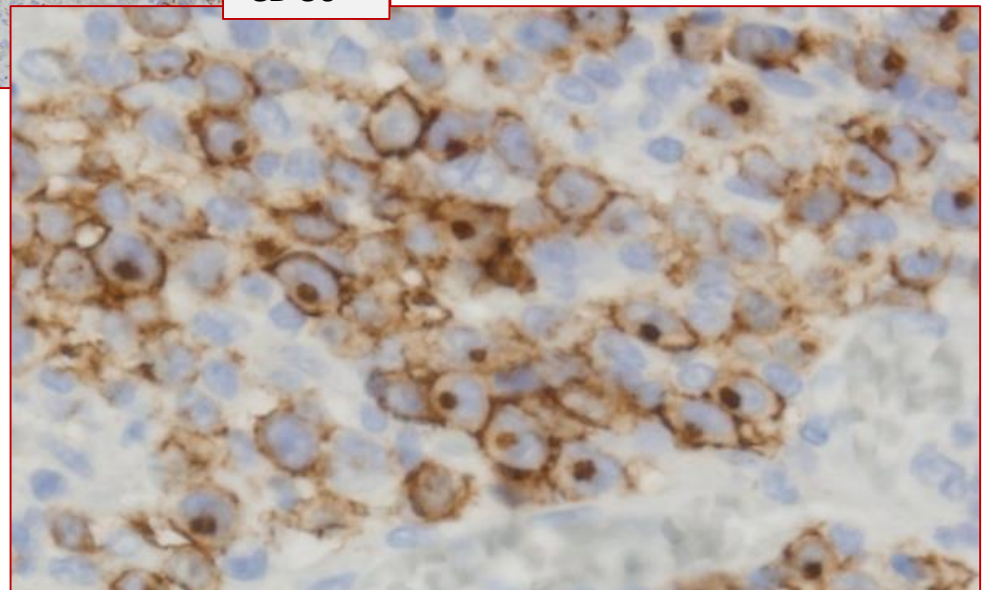
CD 30 +



CD 30 +



granzým B+

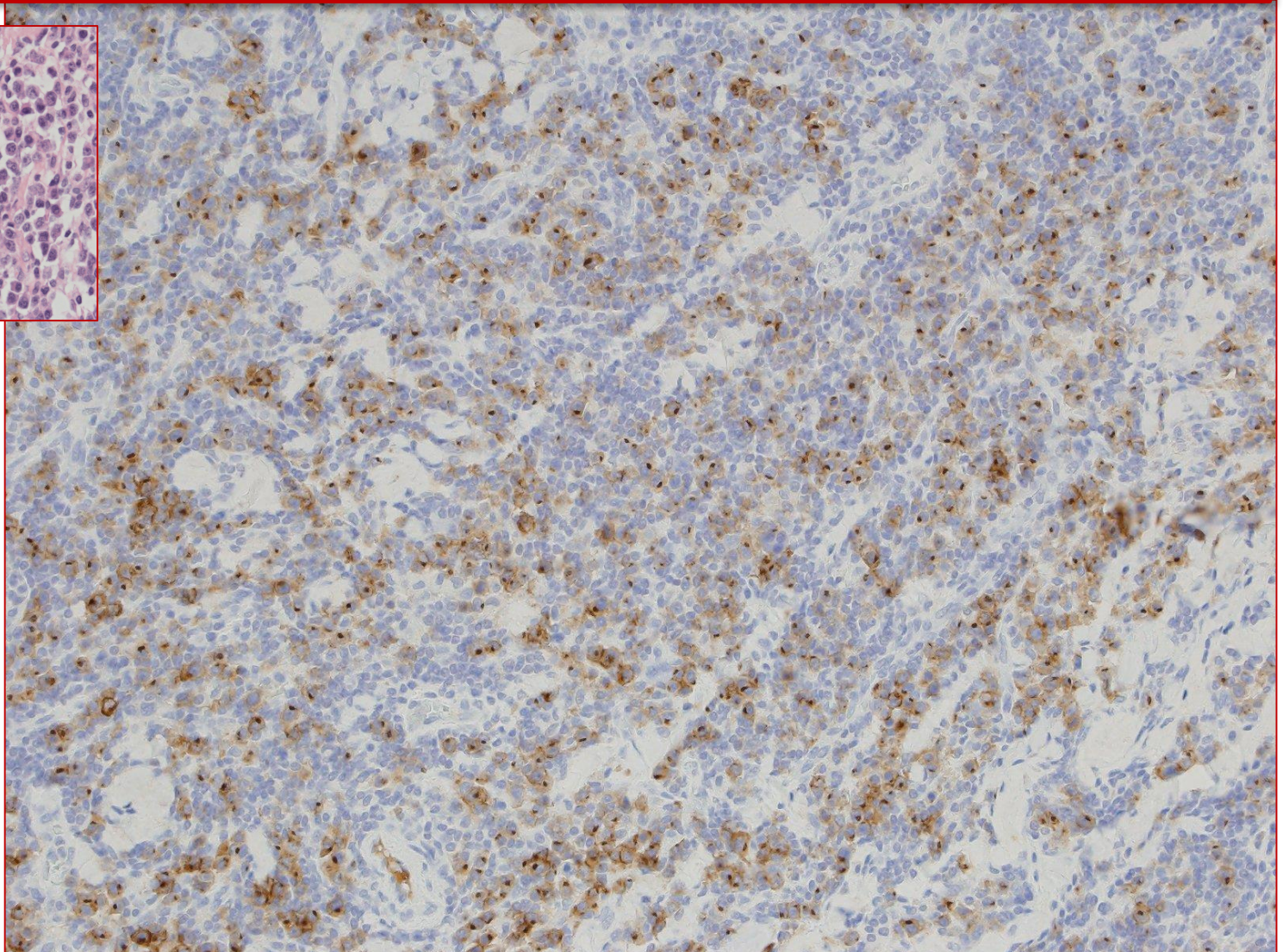
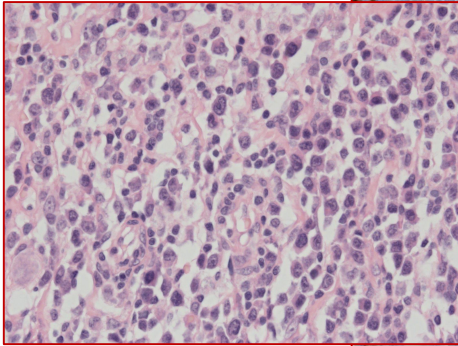


CD 30 +



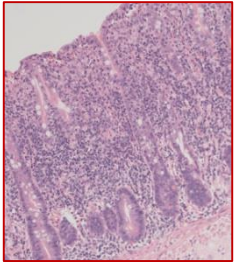
CD30 expresia v „strednej“ časti

CD 30+ veľké T-blasty premiešané s CD20+/ λ + B-populáciou

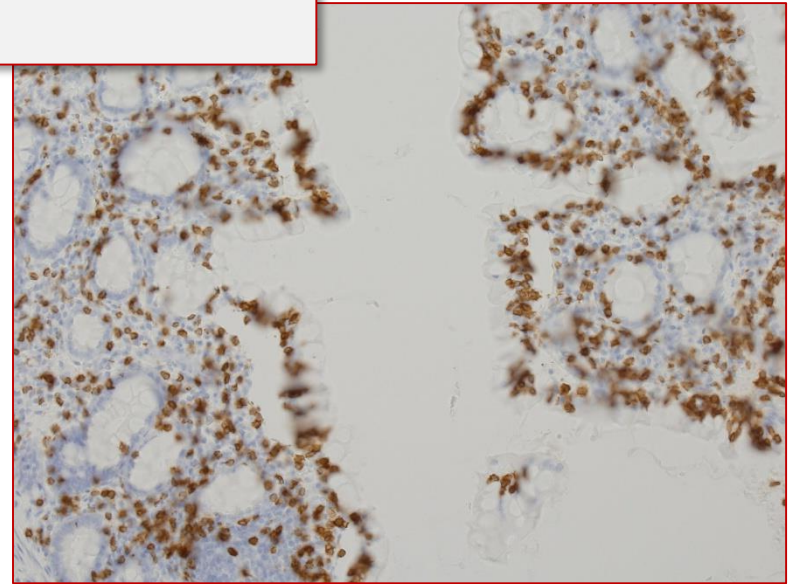
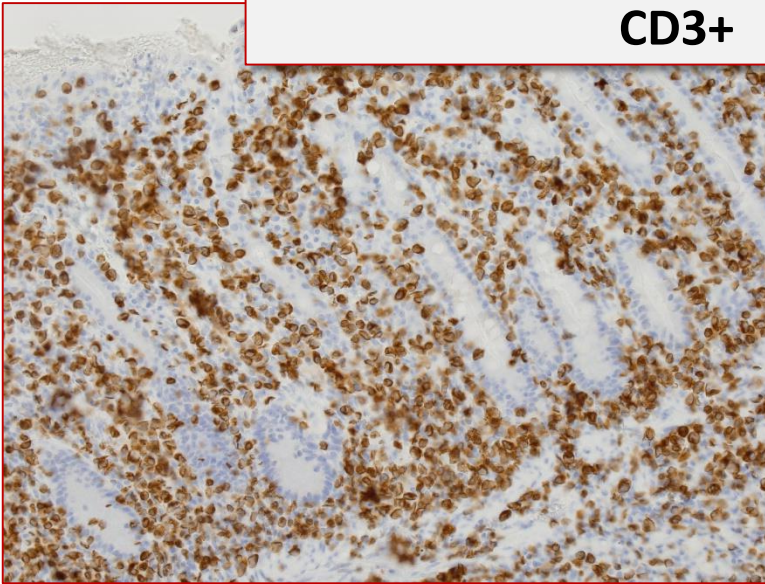




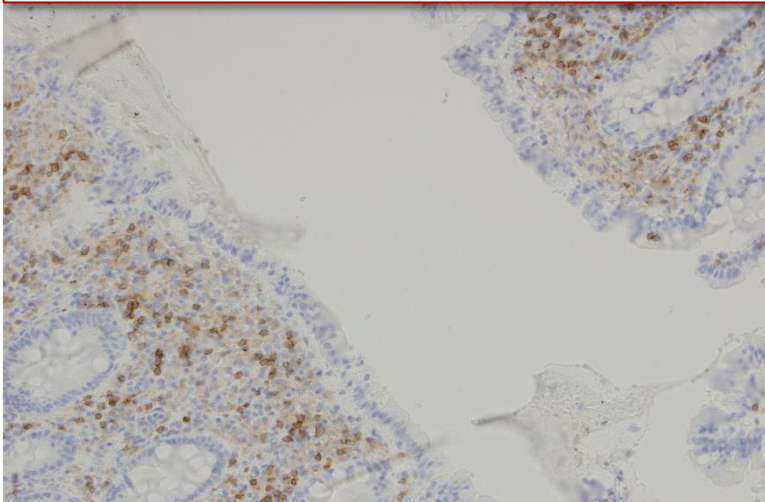
A čo enteropatia ? – 2. HE (blok B)



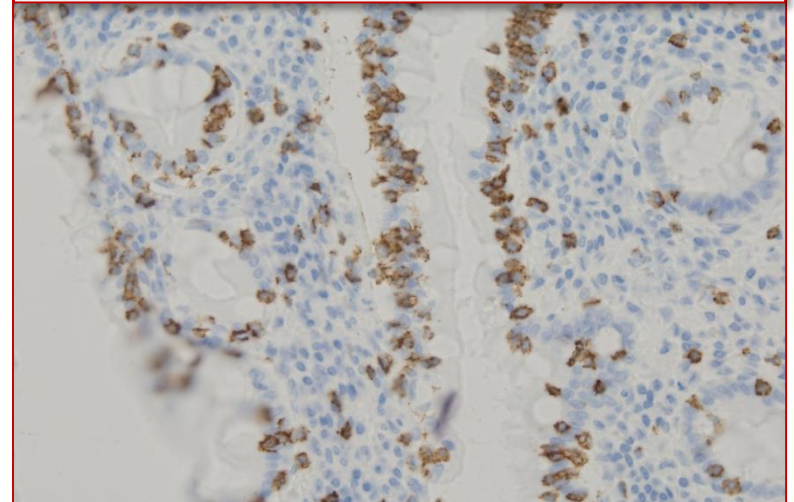
CD3+



Intraepitel. CD4 negat. T-bb

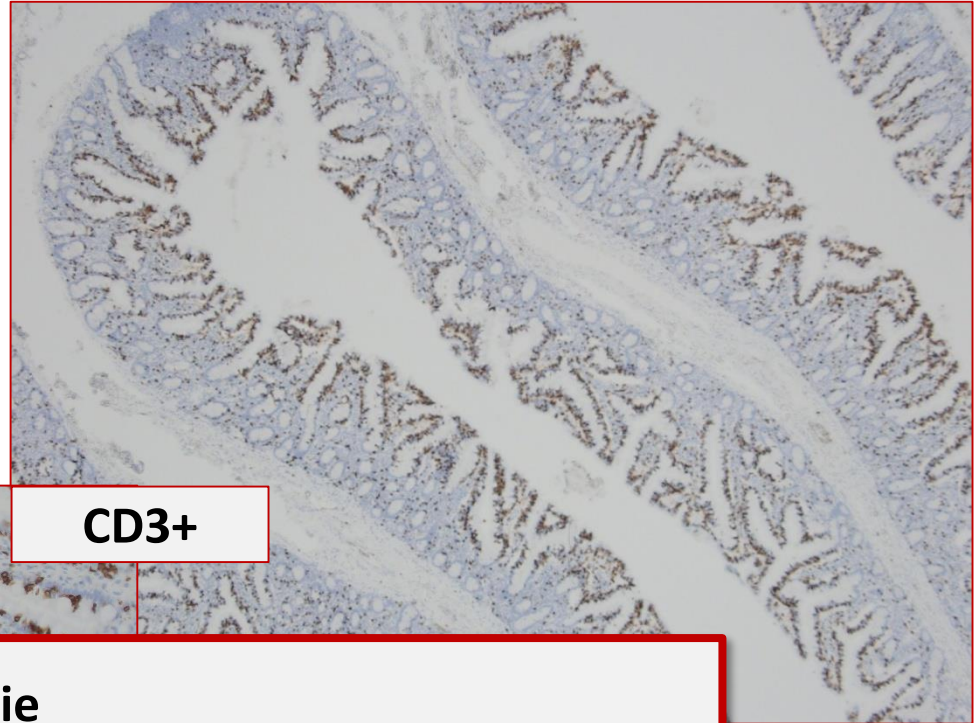
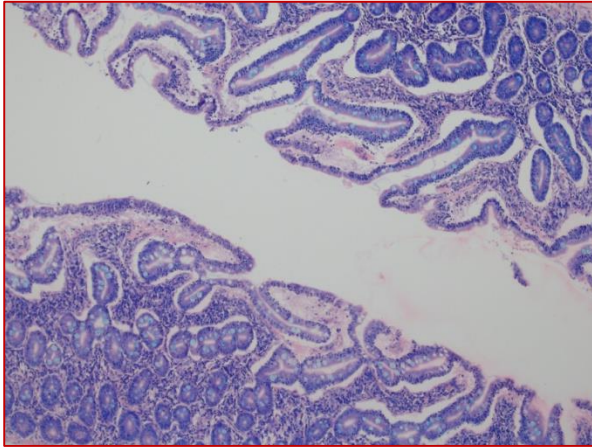


Intraepitel. CD8+/- pozit. T-bb





3. HE (blok)



CD3+

- MAS, typu celiakie
- modifik. Marshova klasifikácia:
deštruktívna lézia, typ 3c
- G2 sec. Corazza

(M. Kalman)



Diagnóza

- **simultánny / zmiešaný / kolízny**
T- a B-bb NHL u pac. s MAS typu celiakie:

= T-NHL: PTCL typu EATL
+
B-NHL: MZBL, MALT-typu

CAVEAT: riziko prehliadnutia B-ML



Zhubné nádory tenkého čreva a lymfómy: zriedkavosť

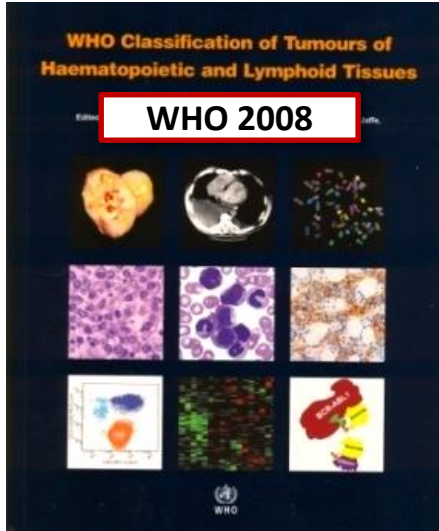
➤ „rakovina“ t. čreva – pribl. len 2% všetkých „rakovín“ GIT-u

- malígne lymfómy: 8% of **všetkých malígnych Tu t. čreva**
- **primárne tenkočrevné ML** tvoria len **9% všetkých primárnych GI ML**, zatiaľčo primárne gastrické ML tvoria až 75% týchto nádorov
- väčšina GI lymfómov: ML EN typu, B- alebo T-bb pôvodu
- **prevládajúci** primárny EN **B-ML tenkého čreva** je: imunoproliferatívne ochorenie t. čreva (**IPSID**) seu „alpha chain disease“
- **predominant** primárny EN **T-ML t. čreva** je: s enteropatiou asociovaný T-lymfóm (**EATL**)



ML tenkého čreva asociované s enteropatiou I.

Enteropathy associated T-cell lymphoma podľa WHO 2008: type I and type II



EATL type I. M9717/35

- PM, histomorphological spectrum
- **preceded by** refractory celiac dis. (**RCD**), with or without ulceration, probably IEL representing EATL in situ
- CD3+, CD5-, CD7+, CD8-/+, CD4-, CD30+/-

EATL type II. M9717/35

- monomorphic, medium-sized cells
- adjacent mucosa: may show villous atrophy and crypt hyperplasia + IELs
- CD3+, CD4-, CD8+, CD56+



ML tenkého čreva asociované s enteropatiou I.

Enteropathy associated T-cell lymphoma podľa WHO 2008: type I and type II

International Journal of Celiac Disease, 2015, Vol. 3, No. 2, 59-68
Available online at <http://pubs.sciepub.com/ijcd/3/2/1>
© Science and Education Publishing
DOI:10.12691/ijcd-3-2-1

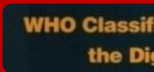
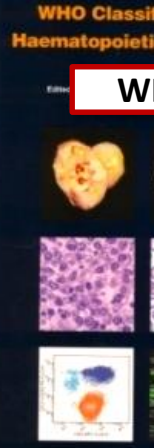


Primary Intestinal Malignant Lymphomas Associated With Celiakia – A Pathologist's Review

Lukáš Plank*

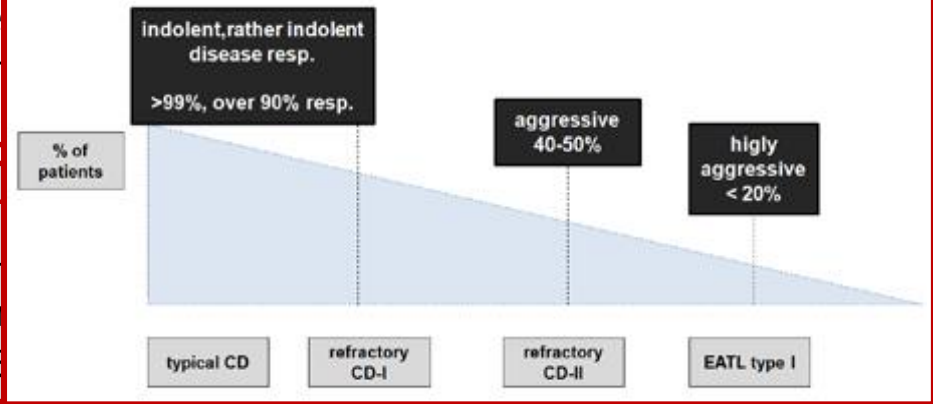
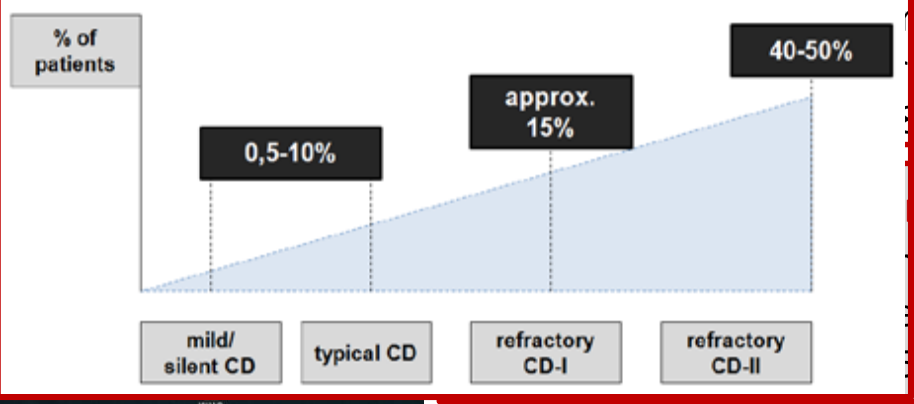
Department of Pathology, Comenius University in Bratislava Jessenius Medical Faculty and University Hospital in Martin and Martin's Biopsy Center, Ltd., Slovakia
*Corresponding author: plank@jfmcd.uniba.sk

IELs



WHO 2010

- PM, histomorphological spectrum





ML tenkého čreva asociované s enteropatiou I.

Enteropathy associated T-cell lymphoma podľa WHO 2016/2017

WHO 2017

WHO Classification of Tumours of

bjh correspondence

CD4-positive small T-cell lymphoma of the intestine presenting with severe bile-acid malabsorption: a supportive symptom control approach

The only primary intestinal T-cell lymphomas (ITCL) recognized as distinct entities by the World Health Organization (WHO) classification are enteropathy-associated T-cell lymphoma types I and II (EATL I and II). Other T-cell lymphomas may involve or arise in the intestine but tend to occur without enteropathy (Isaacson *et al*, 2008). More recently, several indolent T-cell/Natural killer (NK) cell proliferations have been described but have yet to be acknowledged as distinct entities.

Twenty cases of a distinct ITCL associated with sprue-like symptoms have been reported, showing a lymphomatous infiltrate of small, mildly atypical CD4⁺/CD8⁻ lymphoid cells occupying the lamina propria, rarely infiltrating the epithelium and having a strikingly benign clinical course (Carbonnel *et al*, 1994, 1999; Zivny *et al*, 2004; Svrcek *et al*, 2007; Margolskee *et al*, 2013; Perry *et al*, 2013 and Malamut *et al*, 2014).

We report an additional case of a 59-year-old Afro-Caribbean female patient evaluated for a 1-year history of diarrhoea and moderate weight loss. Duodenal biopsies were

symptomatic improvement and an unchanged neoplastic infiltrate in the duodenum throughout the five sequential samples analysed.

Further investigations revealed severe bile acid (BA) malabsorption [Tauroselcholic (75 selenium) acid retention of 4%] and mild pancreatic insufficiency. The patient was successfully managed with dietary changes (reduced fat intake), Colesevelam hydrochloride 625 mg and Creon 10 000 lipase units, with major improvement in previously reported GI symptoms and stable weight.

The most recent WHO Classification recognizes EATL as the only distinct type of primary ITCL showing a poor response to chemotherapy, severely debilitated patients and dismal prognosis (Isaacson *et al*, 2008; Perry *et al*, 2013). Carbonnel *et al* (1994) described a low-grade ITCL with peculiar clinical, morphological and immunophenotypic features. Nineteen further cases with remarkably homogeneous characteristics have been described. They shared similar symptoms and most were initially diagnosed as coeliac disease but failed to improve with gluten-free diet.

malabsorption, or adult

intestinal obstruction,

ic intestinal T-cell

inal T-cell lymphoma

d cells

d yet





ML tenkého čreva II.

Gastric MALT lymphoma, an EN lymphoma primarily described in the stomach,

- can also present in the small bowel
- and is **not** associated with malabsorption
- lymphoma of the elderly

(in contrast to EATL typically diagnosed in **young** adults)

Case Reports in
Gastroenterology

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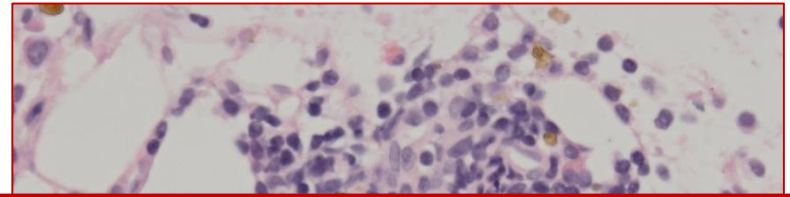
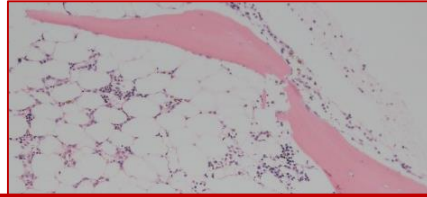
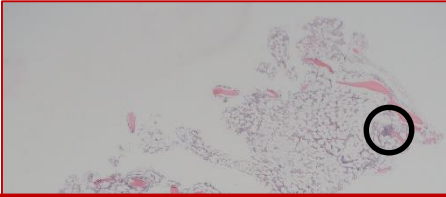
Gastric Marginal Zone B Cell Lymphoma of the Duodenum

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Stagingová trepanobiopsia odobratá 17.05.2018 (NOÚ)



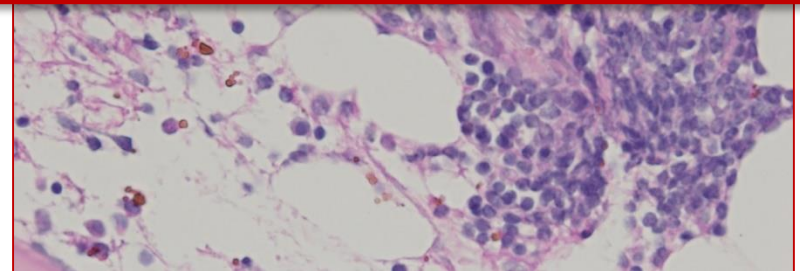
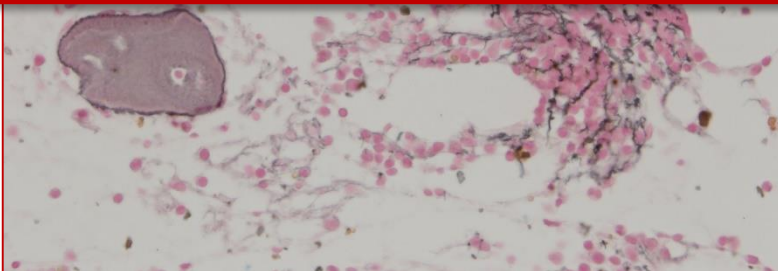
FCA 18.05.2018 (NOÚ), doručené s biopsiou KD **21.05.2018**

Vo vyšetrenej vzorke KD je normálne zastúpenie T-lymfocytov so zníženým pomerom CD4 / CD8.

Medzi nezmnoženými B-bunkami nachádzame malú monoklonálnu (*lambda*) populáciu elementov s nešpecifickým fenotypom, ktorý môže zodpovedať klinicky uvedenému B-NHL typu MALT.

Korelácia s biopsiou.

Prim. Dr. J. Gyárfáš, Mgr. A. Mlčáková





Telefonát s Dr. L. Homolovou (NOÚ) 24.05.2018 11.15 hod.



Pacient od detstva trpí každodennými hnačkami, „neriešil to“

Po našom náleze z resekátu čreva je na bezlepkovej diéte a bez hnačiek ...

(a potom že patológ nelieči ... 😊)

Je vo výbornom klinickom stave, napriek paraparéze

(pred 10 rokmi „double stroke“: ischemická + hemoragická CNS lézia)

Pod dohode s pac. riešia najprv onkologické ochorenie - je na onkologickej liečbe, bez i. nádorových ložísk, potom doriešia MAS



...

