An unusual presentation of Intussusception in a 4 year old child

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HISTORY

- 4 1/2 yrs-old Male presented in ER on May 2022
- c/o Intermittent, colicky abdominal pain since 8-10 days.
- 1 episode of non-bilious, non projectile vomiting
- No c/o fever, loose stools, blood in stools.
- No history of any previous illness.
- Birth history, Development history: normal
- Immunization History: Immunized till 18 months of age.

Patient Vitals

- HR 96 bpm
- RR: 26/min
- BP 118/80 mm Hg. Rt upper limb supine position
- SpO2 98% (room air)
- Peripheral pulses: well felt, no signs of dehydration.
- General examination was normal

Systemic Examination

Per Abdomen:

Inspection: Normal.

Palpation: Tense, diffuse tenderness on all quadrants, Palpable lump-right iliac fossa , 2X1 cm, firm, tender, mobile, no local rise of temperature, not fixed to surrounding structures.

Percussion: Tympanic over all quadrants.

Auscultation: Hyperperistaltic bowel sounds in upper two quadrants. Absent in lower quadrants.

CVS: S1S2 normal. No murmurs

RS: Bilateral Air entry present. No added sounds.

CNS: conscious, alert.

- **Differential diagnosis**:
- Intestinal obstruction
- Intussusception
- Appendicular abscess
- Appendicular mass
- Psoas abscess

Baseline Investigations

- Hemogram
- Hb: 9.9 g/dl
- TLC: 13700 cells/microL, PMN++
- Platelet Count: 3.4 lakh/cumm
- LFT,RFT, Coagulation profile: Normal

Imaging studies

- Xray Erect abdomen
- AbnormalAir-fluid levels are not seen.
- Density at the site of intussusception.

Usg Abdomen/Pelvis

- A well defined hyperechoic lesion of size approx. 35x20x30mm with significant internal vascularity in the epigastric region arising from the lateral wall of bowel loops.
- There is evidence of ring within ring appearance extending from the lesion till the right hypochondriac region likely suggestive of colo-colic intussusception with the above described lesion as the lead point.
- Malrotated ectopic and small sized right kidney.
- Mild ascites
- Mesenteric and retroperitoneal Lymphadenopathy.

Contrast Enhanced CT Abdomen/Pelvis

- Enlarged Mesenteric and Retroperitoneal lymph nodes, largest measuring 20 x 13mm,
- Soft tissue mass approx. 8 cm. in length with 35X20X30mm.
- Classical telescoping of bowel in bowel in the epigastric region having good vascularity suggesting an **ileo-colo-colic intussusception**.
- The SMA-SMV axis was normal.
- A small right malrotated Ectopic kidney.

OPERATIVE NOTES

Supine position, Laparotomy under G.A. with naso - tracheal intubation

Supra-umbilical right transverse muscle cutting incision.

12 to 15 cm. of ileo -colo -colic intussusception (Found & reduced)

3x2x1 cm. Cauliflower growth @ caecum involving the ileo -caecal valve.

Firm, occupying almost the entire lumen of terminal ileum. Mucosa intact.

A segmental localised resection of small bowel, appendix, caecum and IC junction along with the tumour mass

5 cm. margin on either side of tumour

Double-layered ileo -ascending anastomosis.

PRBC transfused and triple antibiotics started.

The post-operative recovery was good.

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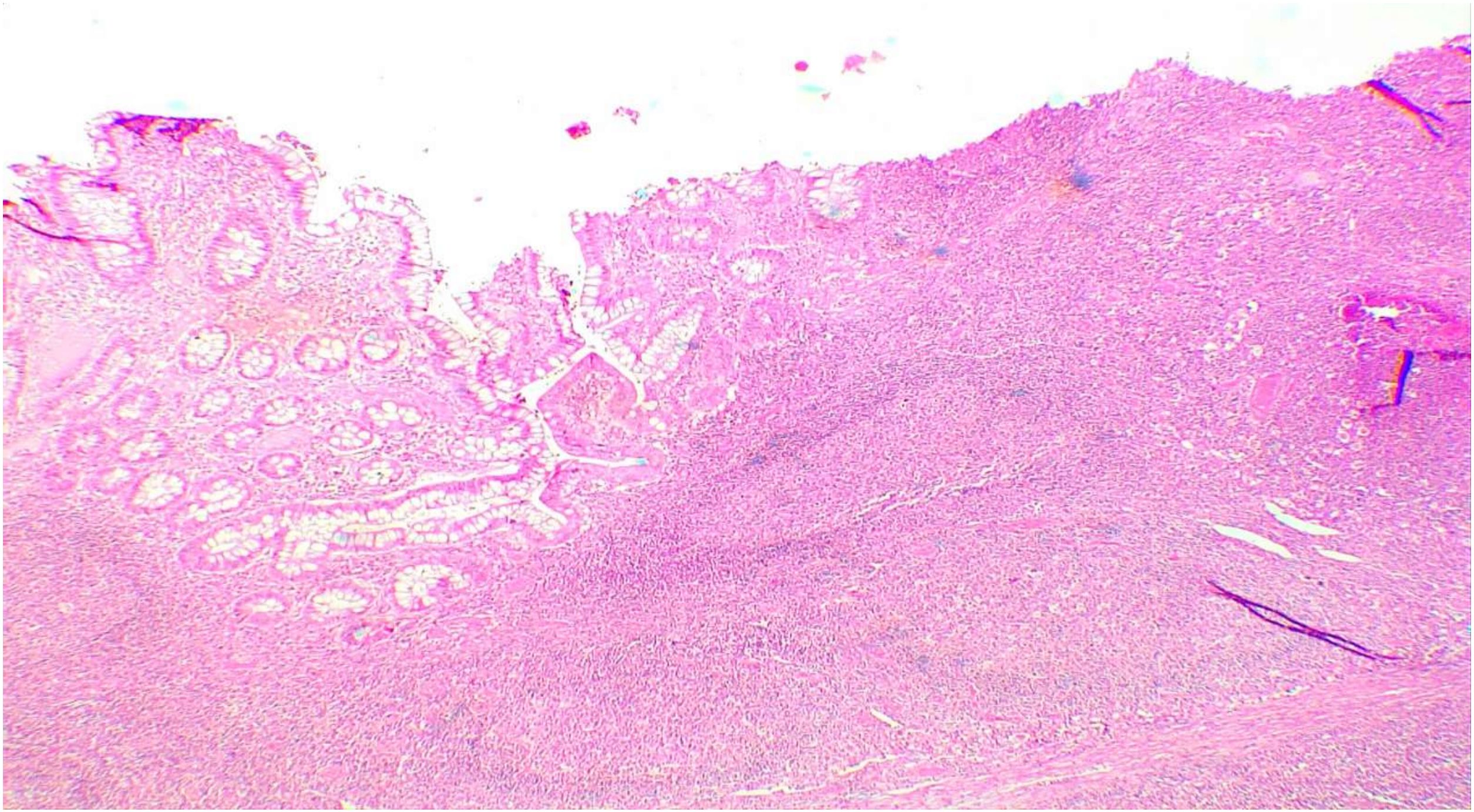
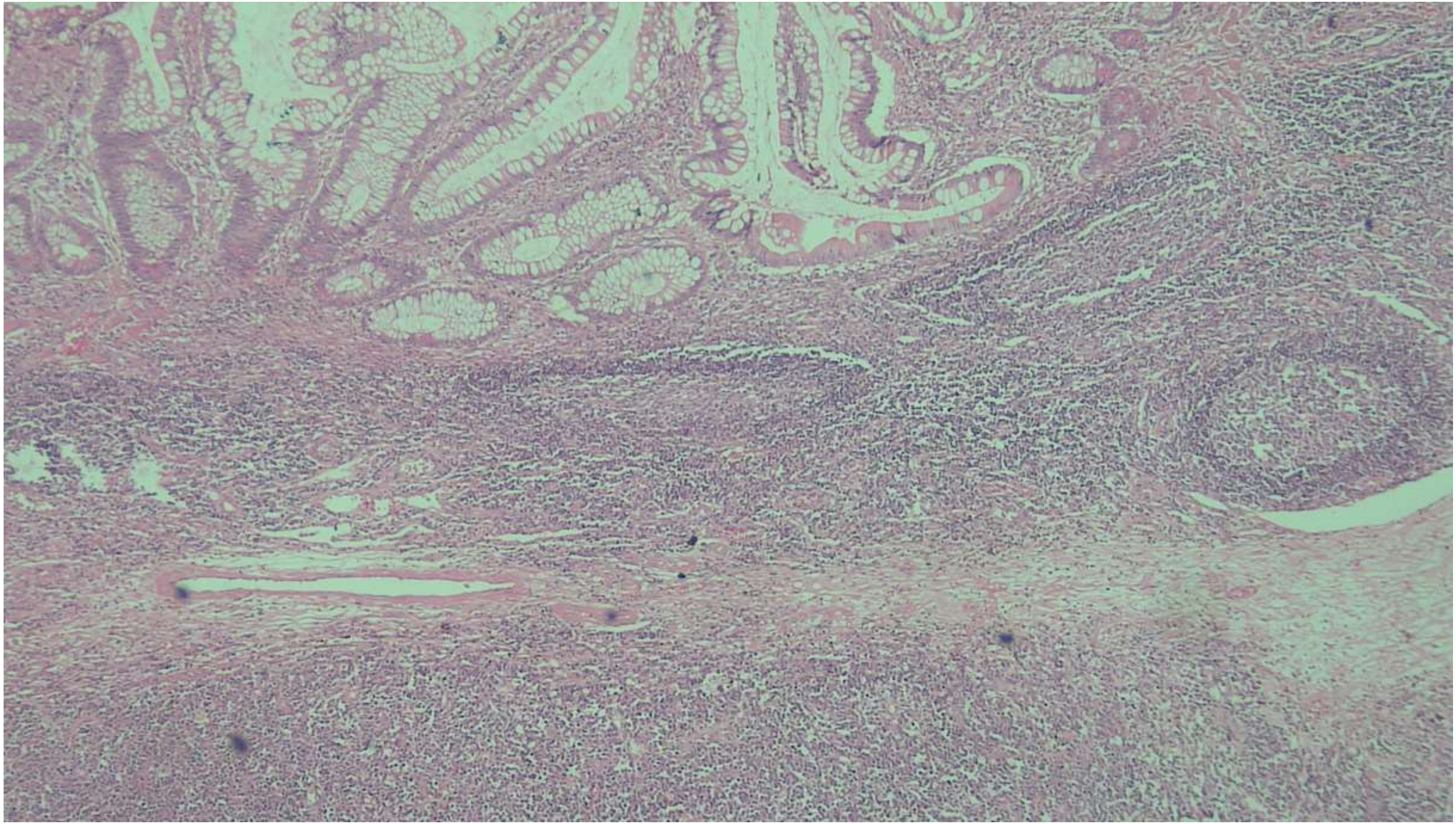
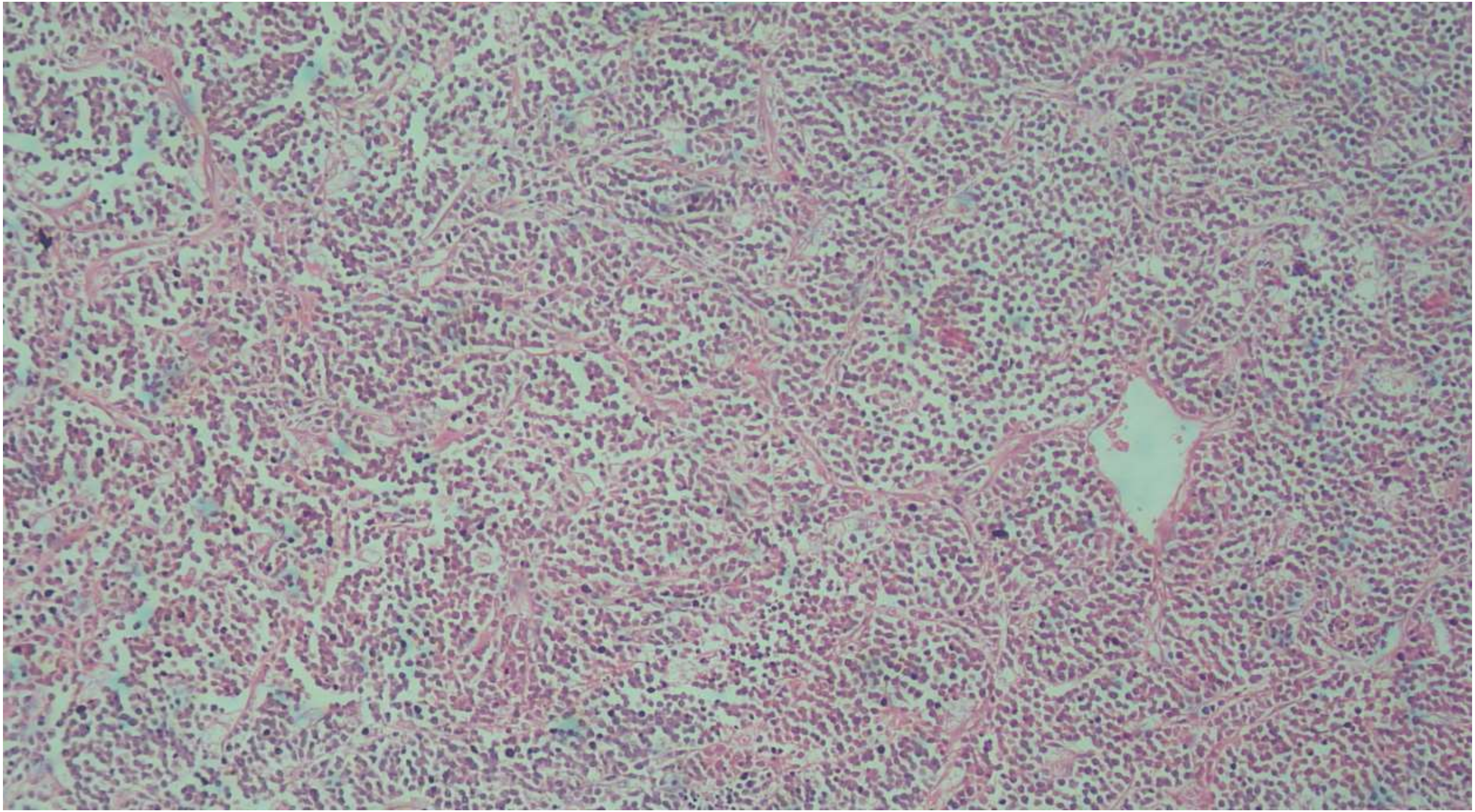
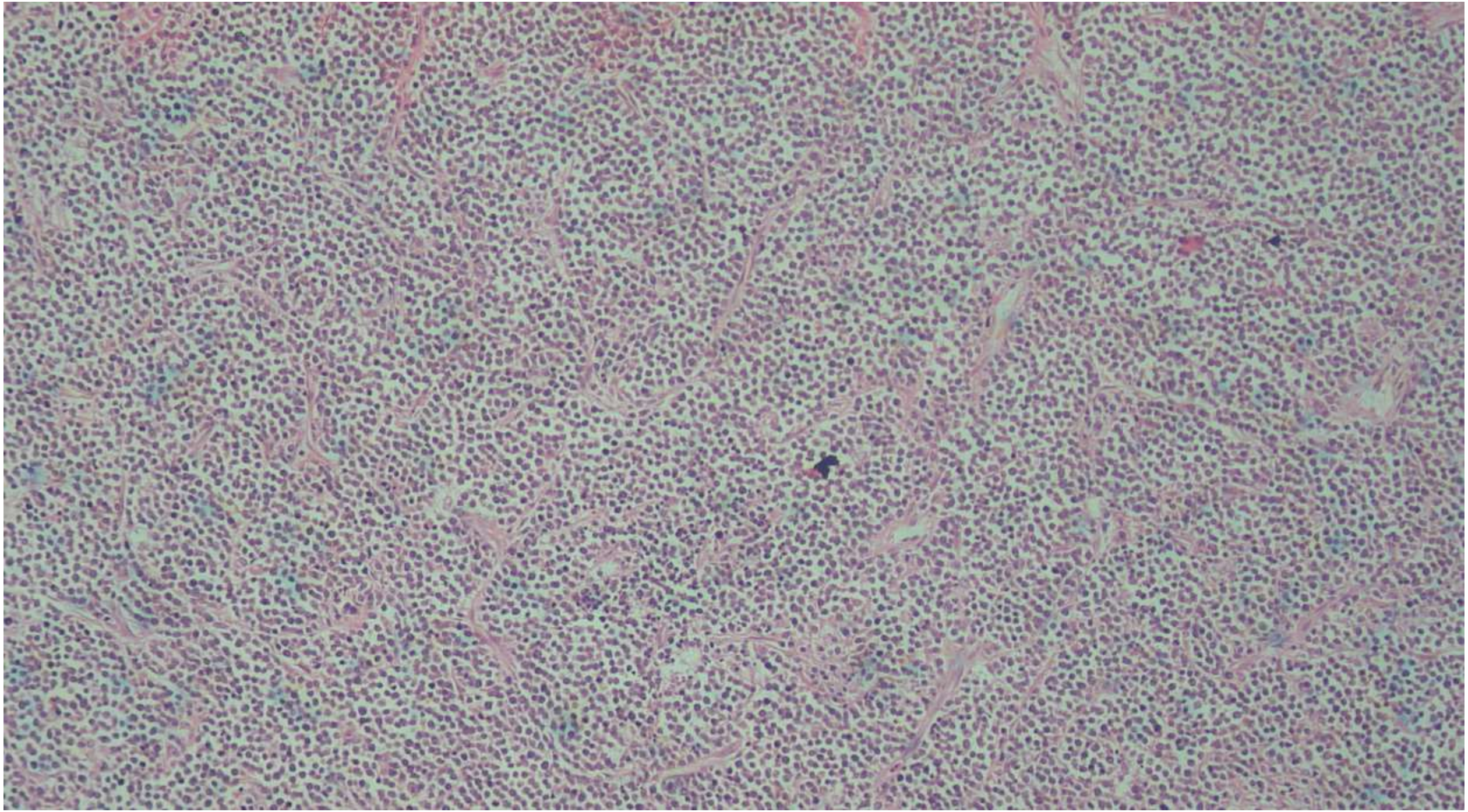
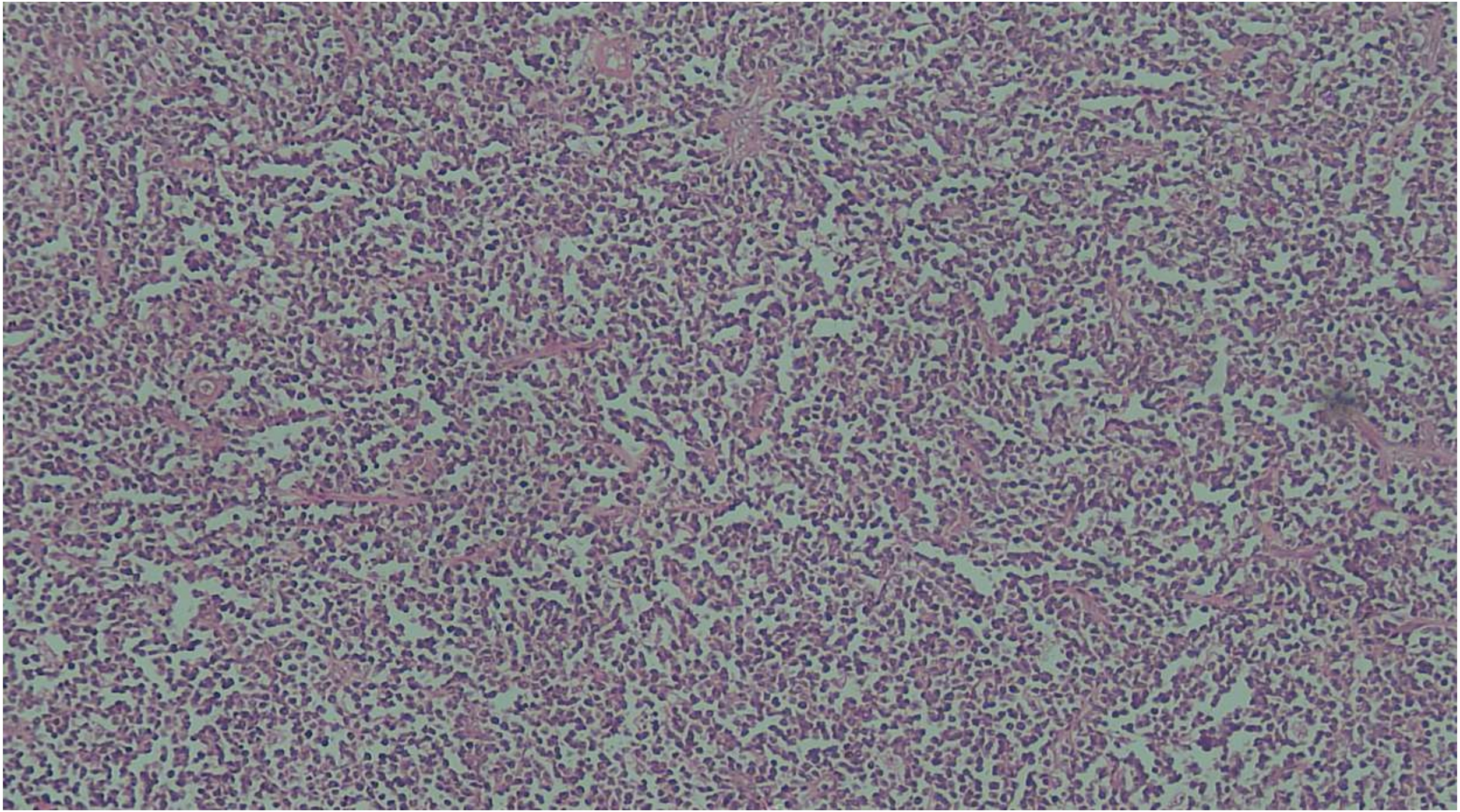
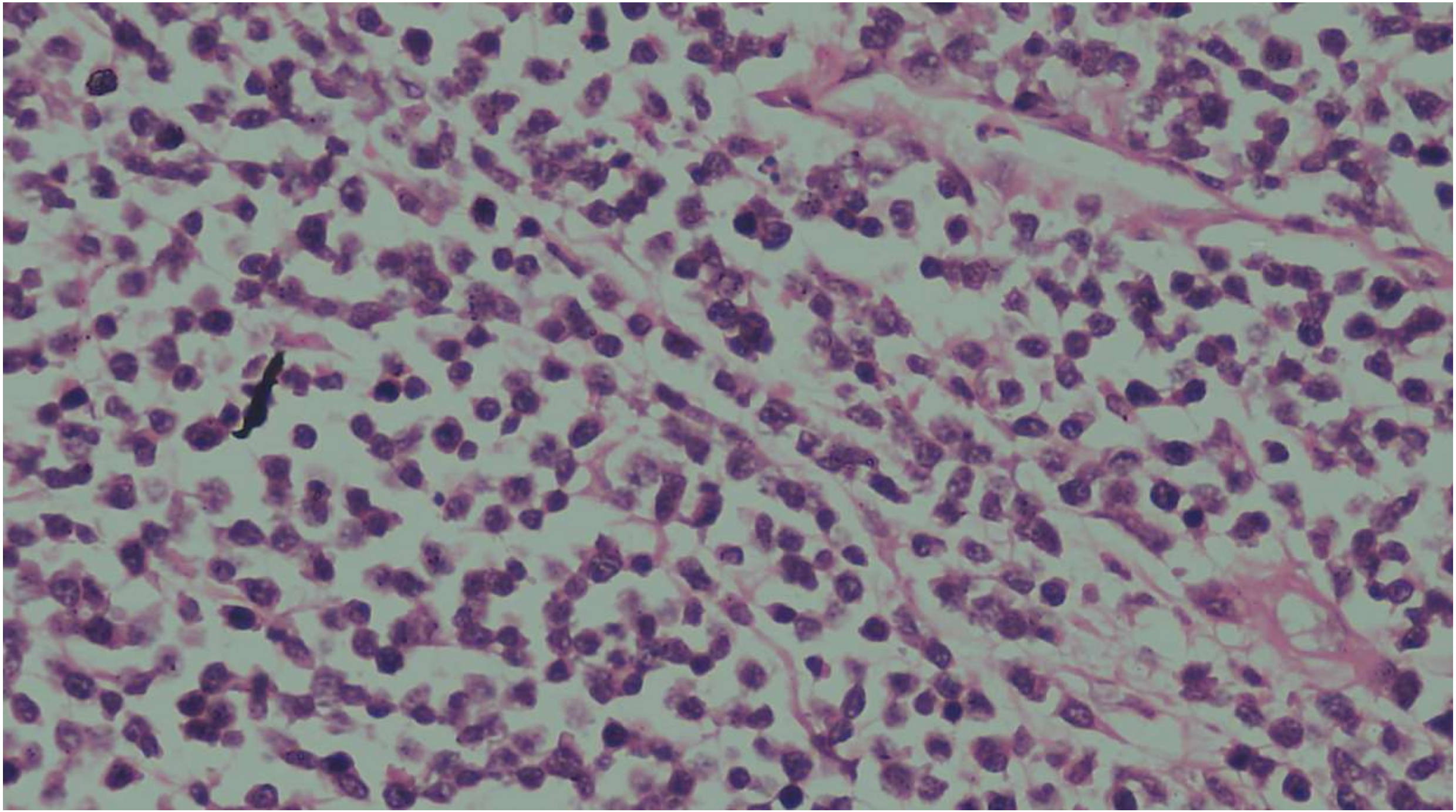


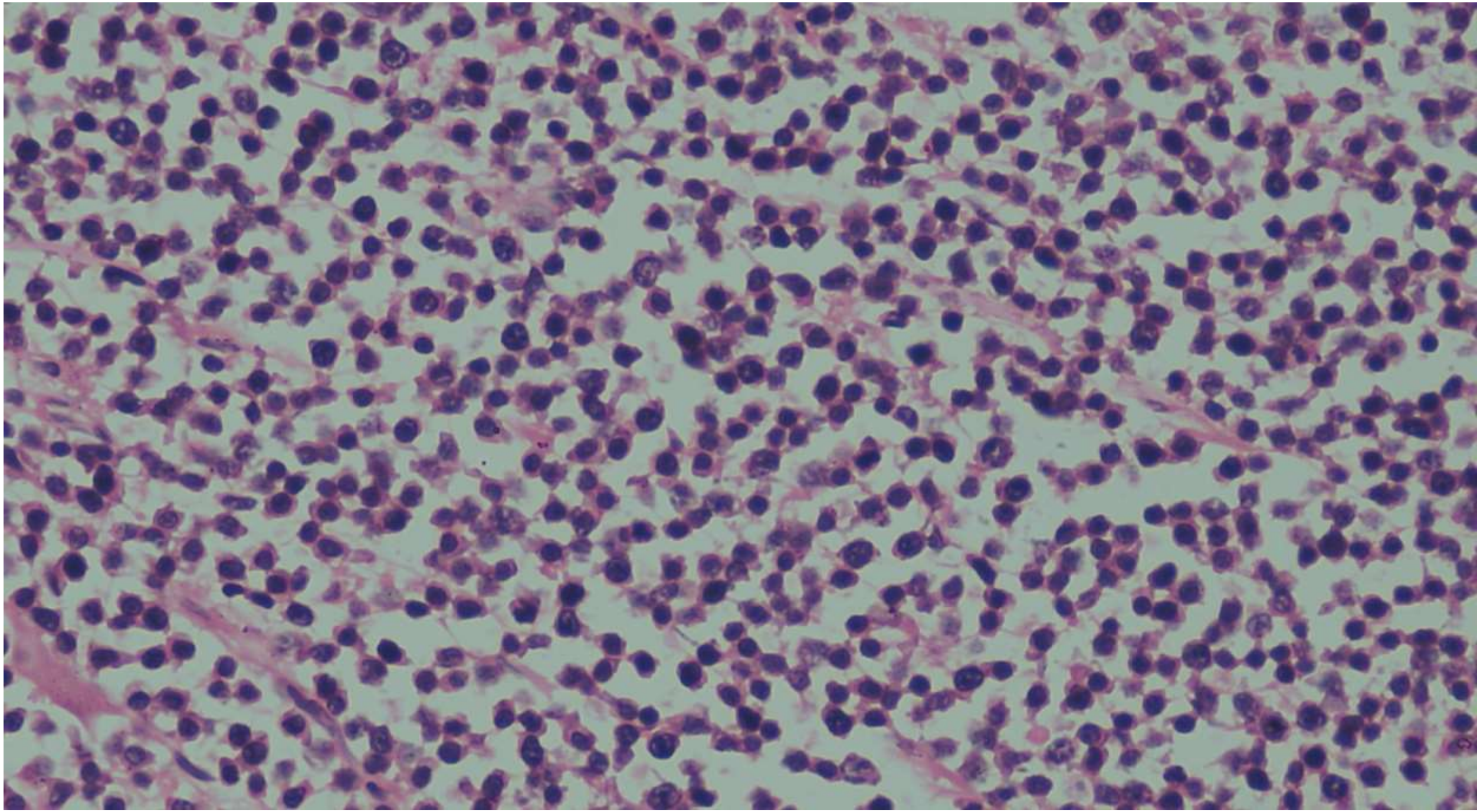


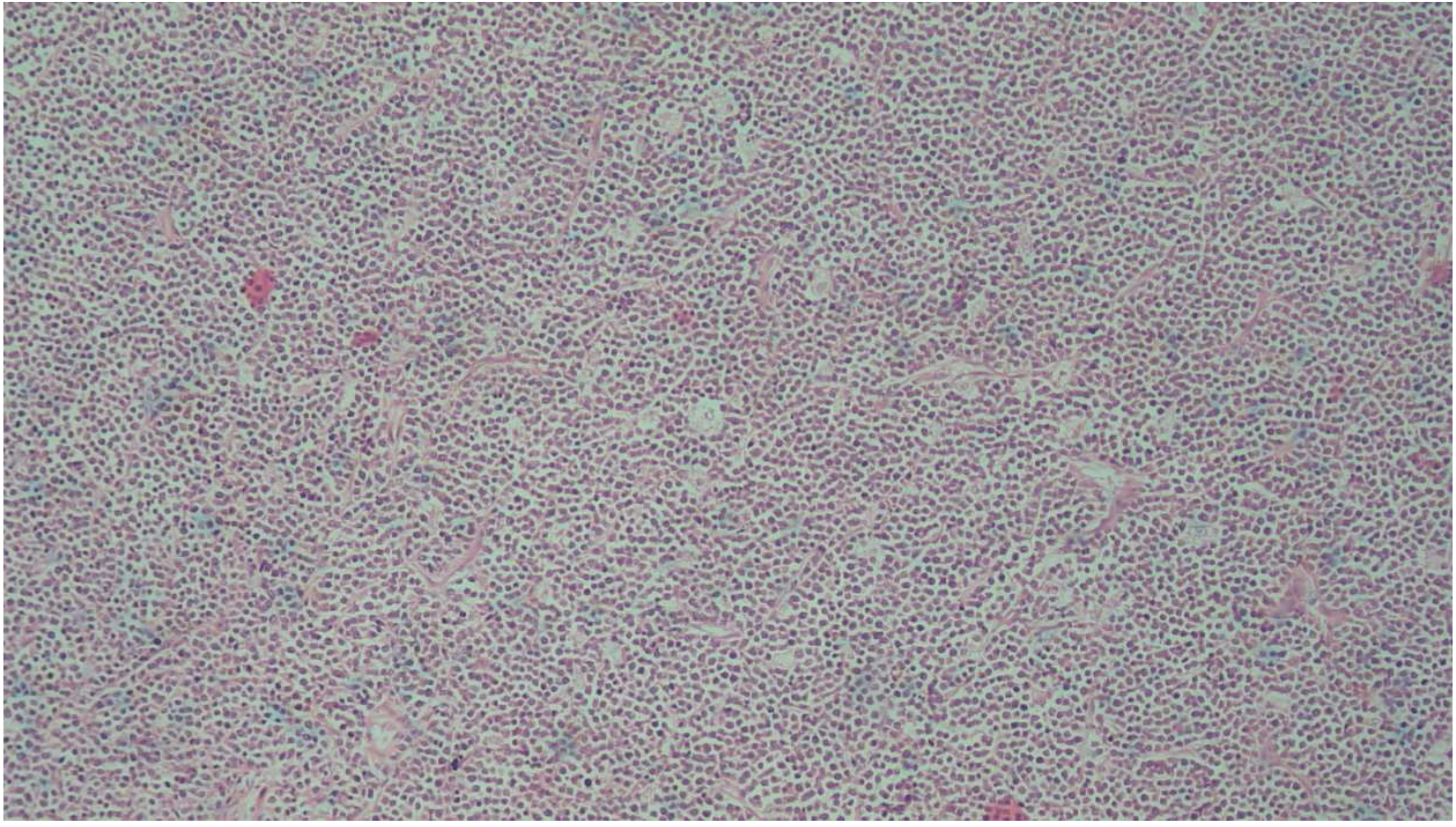












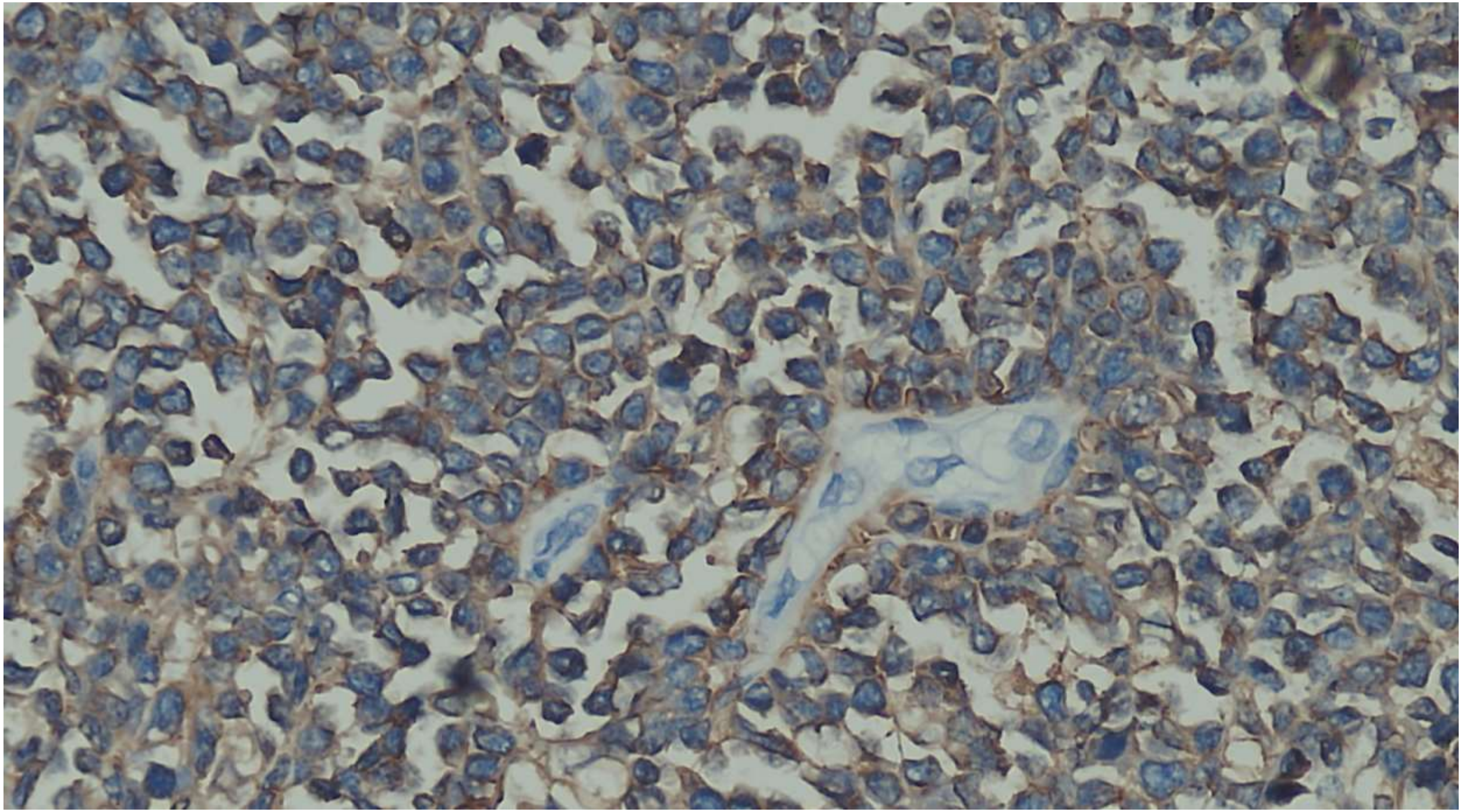
Available IHC marker’s were done

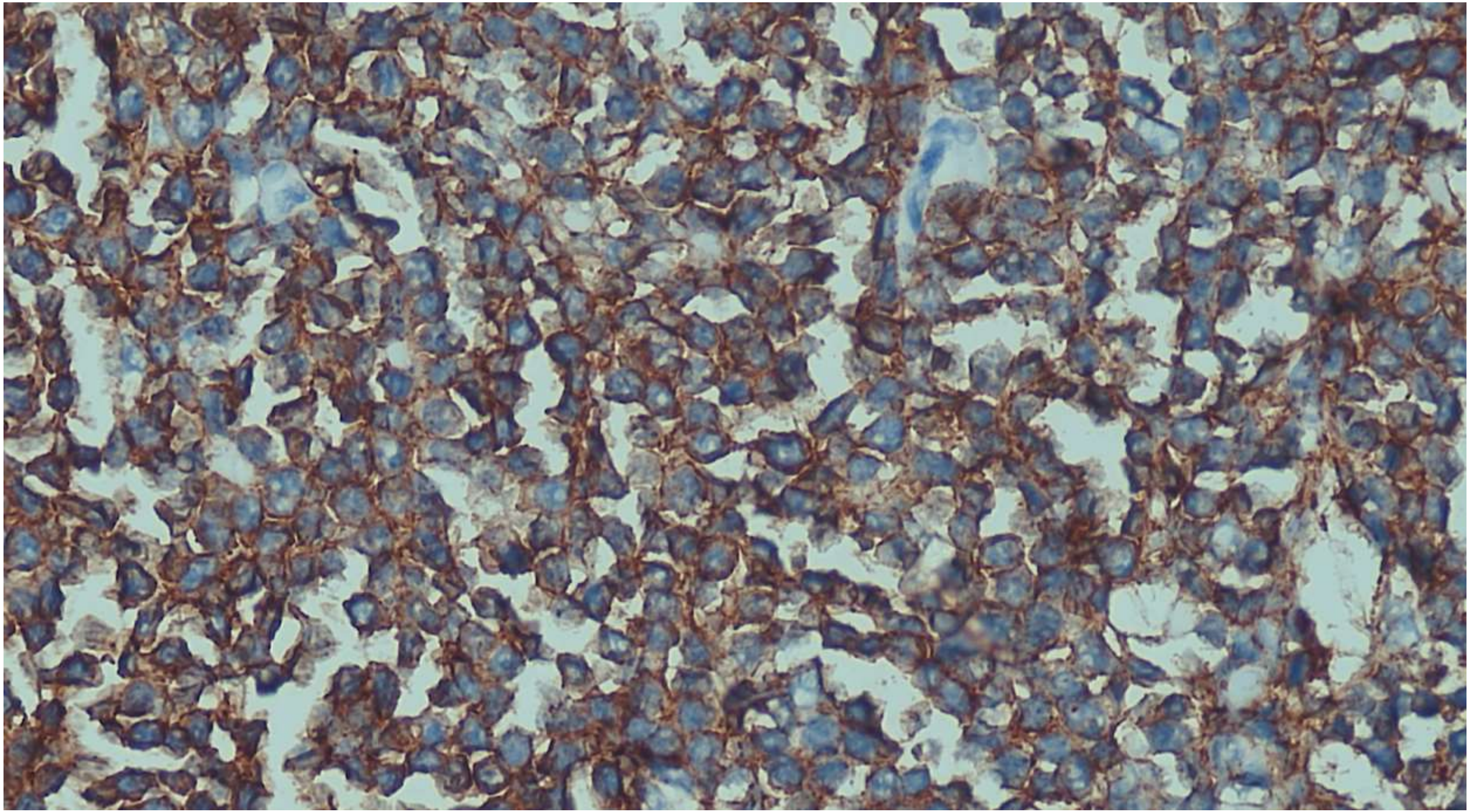
POSITIVE MARKERS:

- LCA : +
- CD20: +
- ALK : +

NEGATIVE MARKERS:

- SYNAPTOPHYSIN : -
- CHROMOGRANIN: -
- CD3: -
- BCL6: -





HISTOPATHOLOGICAL DIAGNOSIS:

EXTRANODAL LYMPHOMA OF ILEUM (NON-HODGKIN’S LYMPHOMA)
DIFFUSE B CELL LYMPHOMA AJCC/ ANN ARBOR STAGING- STAGE 1.

The most helpful immunohistochemical profile to aid in diagnosis of Burkitt's lymphoma includes:

- CD20: POSITIVE
- CD10: POSITIVE

- BCL2: NEGATIVE
- Ki67: >95%

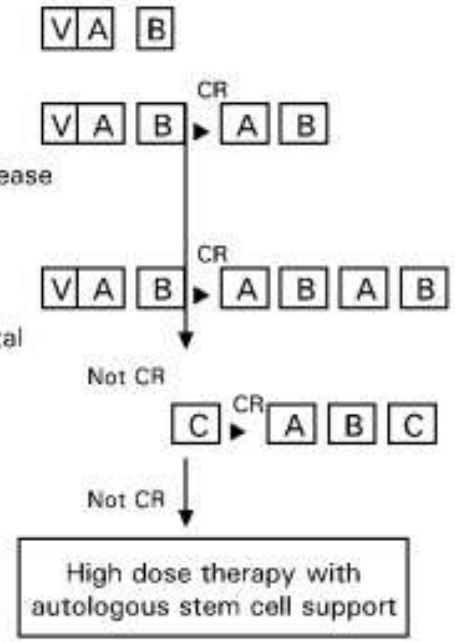
FISH is necessary to support the diagnosis of Burkitt’s lymphoma which includes:

- Presence of MYC translocation.
- Absence of BCL-2 and BCL-6 translocation.

Treatment and further Plan

- The commonest cause of intraabdominal tumor in this age group is Burkitt lymphoma.
- Patient was planned for further Immunohistochemistry on the tissue with marker's like c-myc and BCL2 to confirm Burkitt's Lymphoma.
- Treatment was planned according to BFM NHL protocol to be started on postoperative day 10/14.
- Staging investigations (PET Scan with bone marrow aspiration and biopsy) were planned to be done after wound healing.
- Wound healed and post operative recovery was good.
- Patient was discharged on day 10 postoperatively and was asked to follow up after one week.

Risk group	Treatment blocks
R1	Complete resection of primary tumour
R2	Tumour not resected LDH elevated or only extra-abdominal disease
R3	Abdominal disease and LDH elevated or bone marrow/CNS/sceletal involvement



V: Vorphase: cyclophosphamide 200 mg/m^2 i.v. day 1 -5, dexamethasone 4 mg x 4 day 1–5.

A: A-block: vincristine 2 mg i.v. day 1, methotrexate 3000 mg/m^2* i.v. day 1 (24 h infusion with calciumfolinate resque after 42 h), teniposide100 mg/m^2 i.v.day 4 -5, ifosfamide 800 mg/m^2 i.v. day 1 -5, cytarabine 150 mg/m^{2}x 2 i.v. day 4 -5, dexamethasone 4 mg x 4 p.o. day 1 –5 methotrexate 15 mg, prednisolone 10 mg and cytarabine 40 mg I.t. day 1 (and day 5 if CNS involvement).

B: B-block: vincristine 2 mg i-.v. day 1, methotrexate 3000 mg/m^2* i.v. day 1 (24 h infusion with calciumfolinate resque after 42 h) , doxorubicin 25 mg/m^2 i.v. day 4 -5, cyclophosphamide 200 mg/m^2 i..v. day 1 -5, examethasone 4 mg x 4 p.o. day 1 –5, methotrexate 15 mg, prednisolone 0 mg and cytarabine 40 mg i.t. day 1 (and day 5 if CNS involvement).

C: C-block: cytarabine 2000 mg/m^2 x 2 i.v. day 1-2 , vindesine 3 mg i.v. day 1 etoposide 150 mg/m^2 i.v. day 3 –5, dexamethasone 8 mg x 4 p.o. day 1 - 5, methotrexate 15 mg, prednisolone 10 mg and cytarabine 40 mg i.t. day 5.

*For patients above 50 years, methotrexate is reduced to 500 mg/m^2 i.v. for the first course and, if well tolerated, increased to 1500 mg/m^2 for the subsequent cycles.

Discussion

- Intussusception is a pathology in which telescoping of a proximal segment of bowel occurs into lumen of the distal segment.
- Most common cause of intestinal obstruction between 5 months and 3 yr of age and the most common emergency in children younger than 2 yr of age.
- Incidence:1.5-4 cases per 1000 live births,
- Male-to-female ratio 3:1.
- 60% - < 1yr, 80% - < 24months.
- Gastrointestinal infection or the introduction of new food proteins results in swollen Peyer patches in the terminal ileum. Lymphoid nodular hyperplasia is another related risk factor.
- Prominent mounds of lymph tissue lead to mucosal prolapse of the ileum into the colon, thus causing an intussusception.

Etiology

- Approx 90%: Idiopathic
- In 2–8% of patients,
- **recognizable lead points** for the intussusception are found, such as a
- Meckel diverticulum,
- intestinal polyp,
- neurofibroma, intestinal duplication
- cysts, inverted appendix stump, hamartomas, ectopic pancreatic tissue, anastomotic suture line, enterostomy tube
- Post transplant lymphoproliferative disease,
- Hemangioma
- Malignant conditions such as lymphoma or Kaposi sarcoma.
- NHL – 17%

Non Hodgkin's Lymphoma

- Subtypes
  - **Lymphoblastic lymphoma (LBL), -** T lymphocytes
  - **Mature B-cell lymphoma**, and
  - **Anaplastic large cell lymphoma** (**ALCL)**
- Mature B-cell lymphomas comprise
  - **Burkitt lymphoma (BL)** and
  - **Diffuse large B-cell lymphoma (DLBCL)**.

Ann Arbor Staging System

Stage I	**Single node region (I) or one extralymphatic site (Ie)**
Stage II	Two or more lymph node regions, on the same side of diaphragm (II) or local extralymphatic extension plus one or more lymph node regions on the same side of the diaphragm.
Stage III	Lymph node regions on both sides of diaphragm, which may be accompanied by extralymphatic extension.
Stage IV	Widespread Disease involving extra-lymphatic sites plus Lymph Node involvement
Modifiers	
A	Absence of symptoms
B	Fever, Night sweats, weight Loss, (>10% Body weight), fatigue, Bone joint Pains, itching.
X	Bulky disease (>10cm)

Outcome

- Prognosis of Burkitt’s Lymphoma
- Although it is an aggressive disease it shows good prognosis in initial stages.
- Stage 1: 90%
- Stage 2: 75-80%
- In advanced stage: stage 4- 50-60%

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