MACROCYSTIC LYMPHATIC MALFORMATION

(CYSTIC LYMPHANGIOMA)

(MACROCYSTIC LYMPHANGIOMA)

Edited by:
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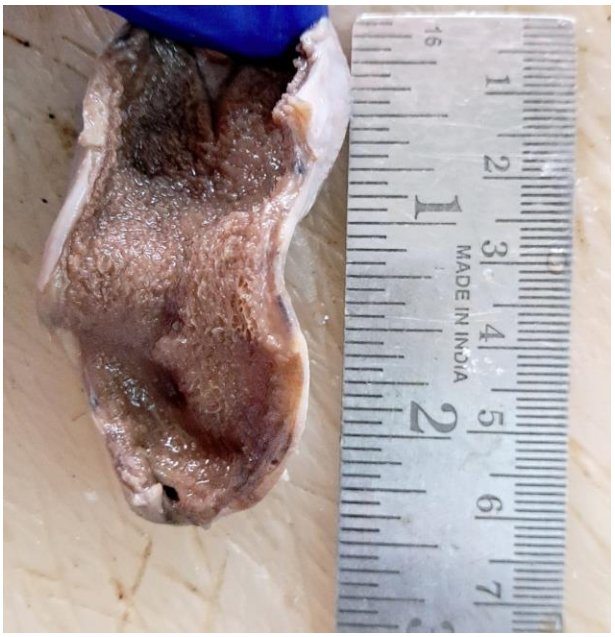
Ex Dean Goa University
President Asian Society of Cytopathology

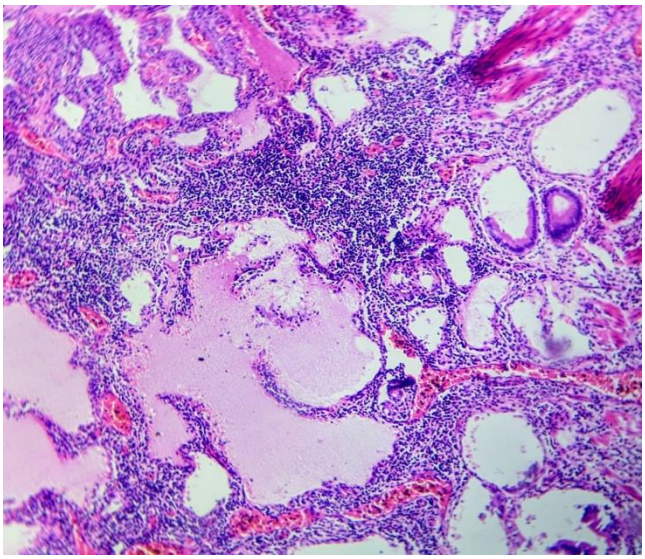
AUGUST 2024

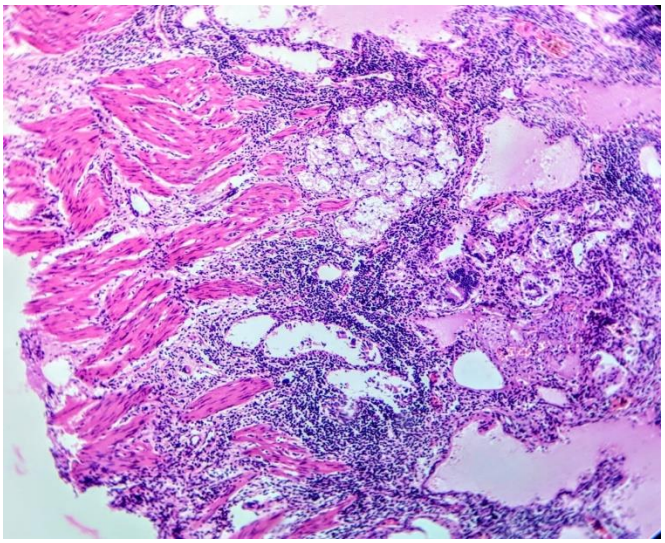
Case
Lymphangioma of Gall Bladder
30 y Female
Presented with Acute Cholecystitis with Cholelithiasis

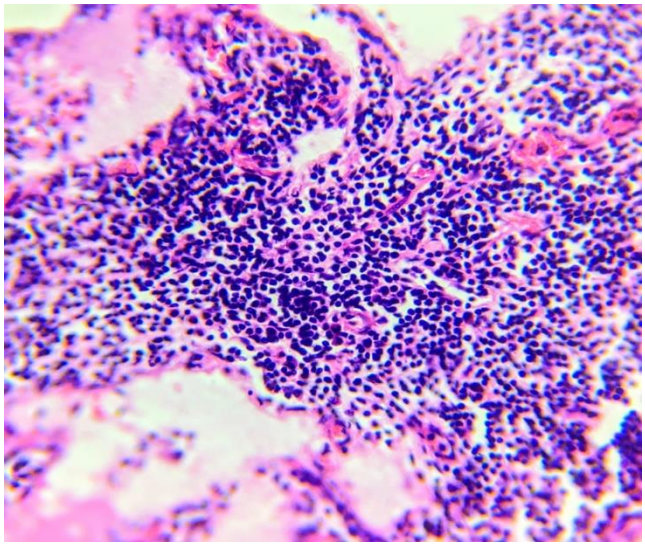
Reported by
Dr RGW. Pinto
Dr Ketan Suncthankar
Dt Peter Rodrigues
Dr Gaurangi Velip
Dr Shivshankar K
Dr Shubhra Amonkar

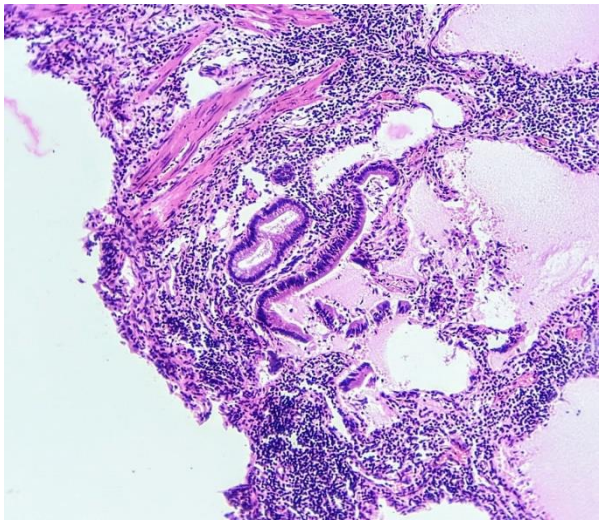
Operated by Dr Jude
Dr Reuben







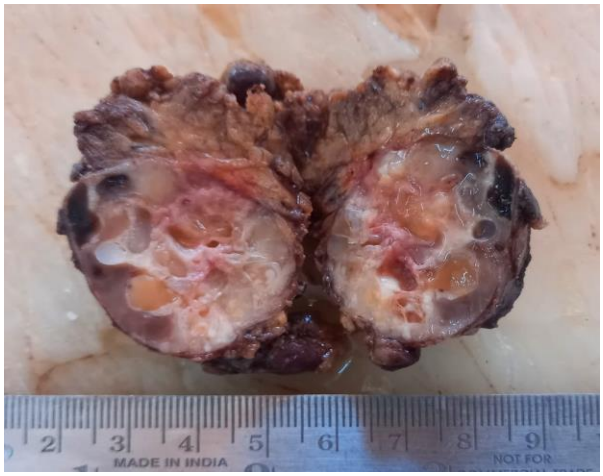


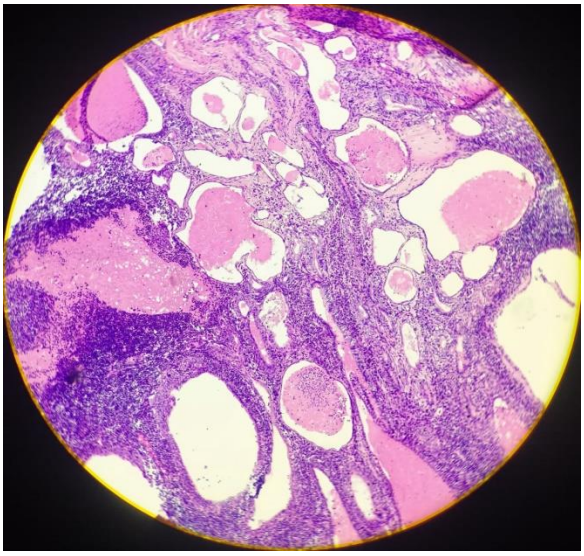


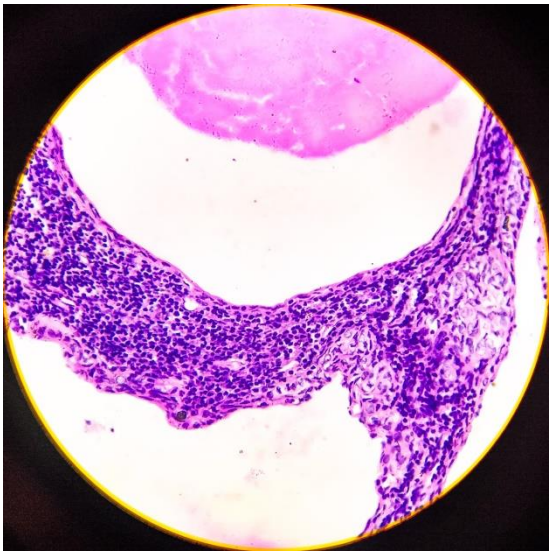
Case
Right Parotid Lymphangioma
75 y Female

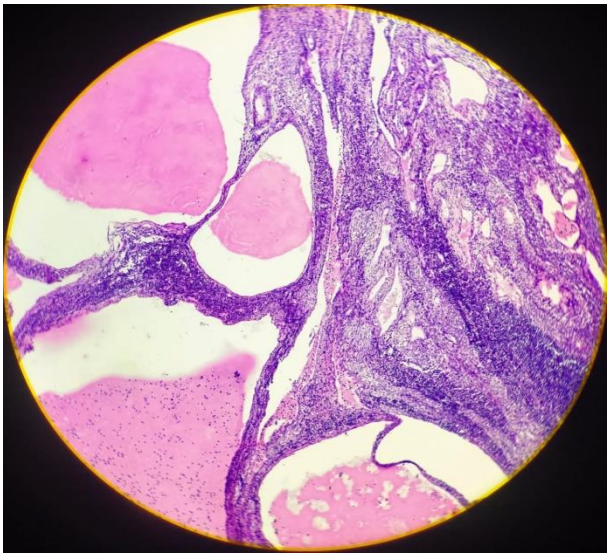
Operated by
Dr Jude Rodrigues

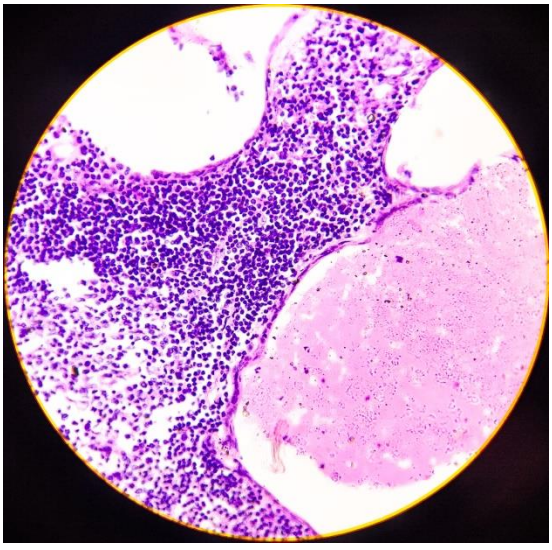
Reported by
Dr RGW Pinto
Dr Ketan Suncthankar
Dr Sanjana Lotlikar
Dr Amala Kudchadkar











Cystic Lymphatic malformation Of Sigmoid Mesocolon

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Tutor
Department Of Paediatric Surgery

Dr Aditya
Assistant Professor
Department Of Paediatric Surgery

CASE SUMMARY

- 10 months old female
- progressive abdominal distension and decreased appetite for 15 days

- P/A:
  - 10 x 12 cm firm, non-tender, ill defined intraabdominal mass
  - arising from pelvis and reaching till epigastrium,
  - not moving with respiration, minimal intrinsic mobility noted

Probable clinical diagnosis

Vague infra-abdominal asymptomatic mass in female child

- Ovarian lesion- benign ovarian cyst, teratoma
- Mesenteric cyst, omental cyst
- Rectal or colonic duplication cyst
- Pelvic tumors like rhabdomyosarcoma, neuroblastoma

INVESTIGATIONS

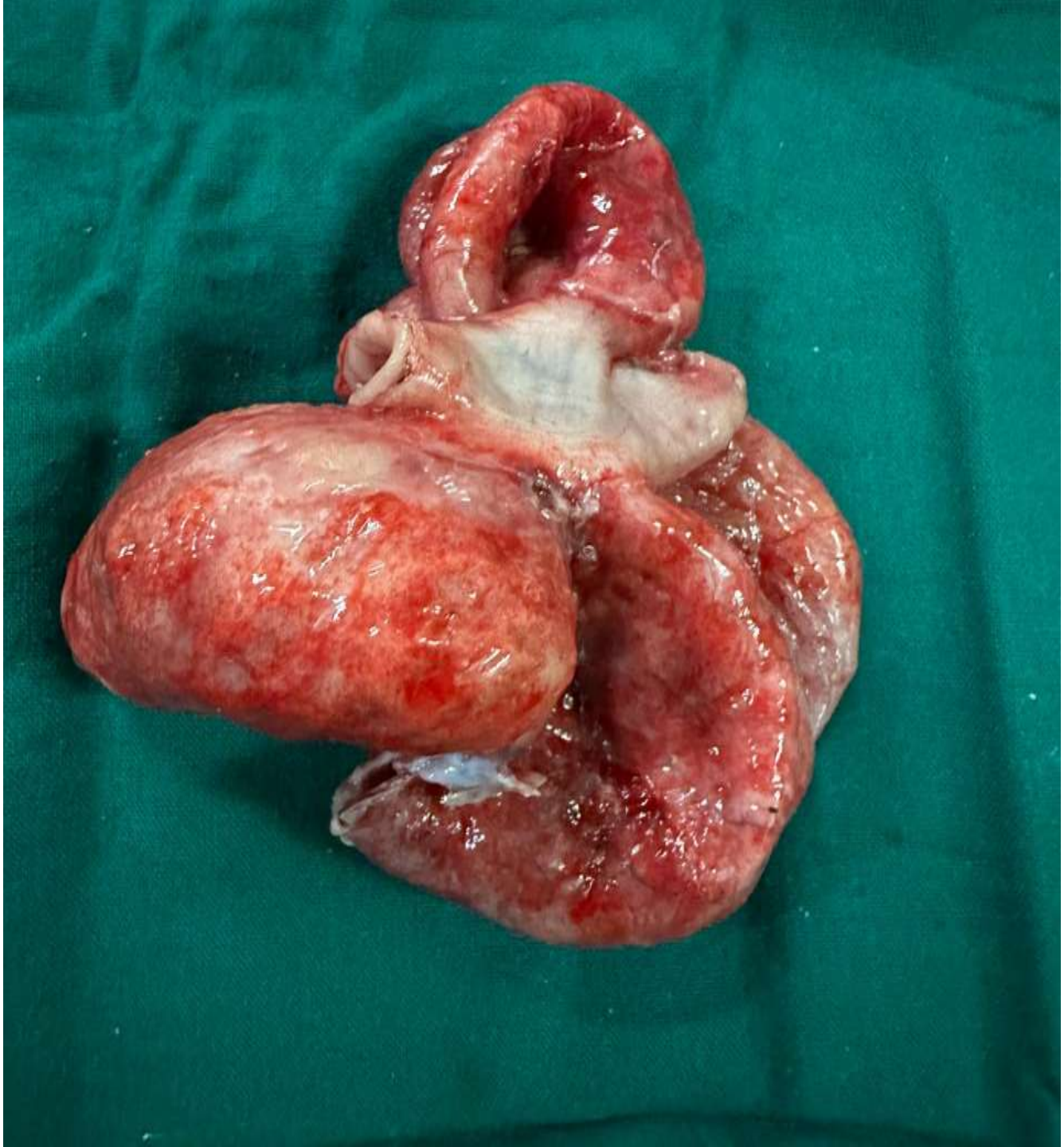
- BR, RFTe, LFTs were WNL
- Se Alpha Fetoprotein 9 ng/ml (WNL)
- Se Beta Human C**horionic Gonadotropin < 1.2 mIU/ml**
- **<u>USG abdomen</u>**
  - 12 x 6cm multiloculated cystic lesion
  - Occupying pelvis indenting the dome of bladder
  - Superiorly seen to reach upto the epigastrium
  - Uterus measures 5.1 x 1.3 cm in size
  - Both ovaries not well delineated

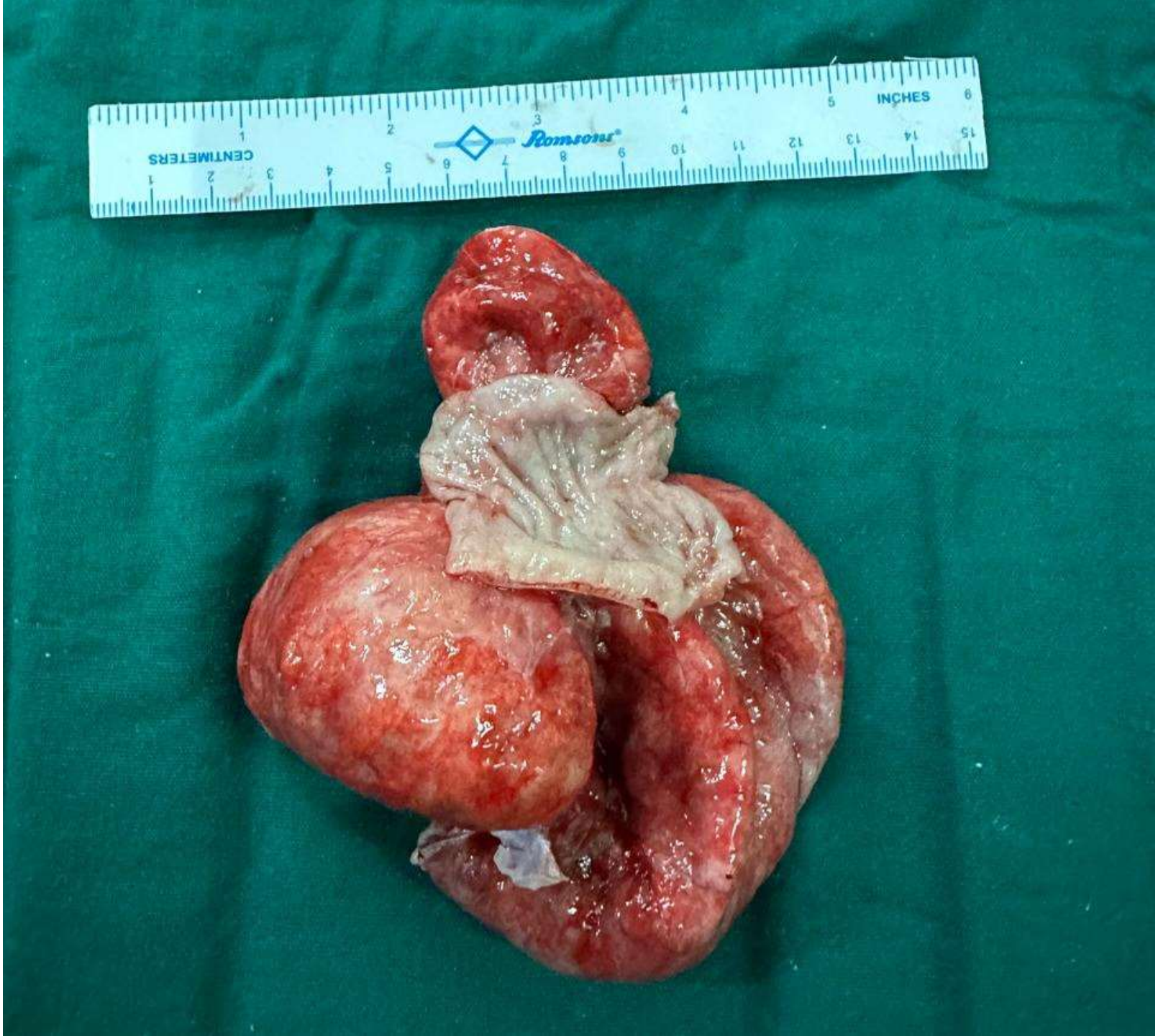
CECT ABDOMEN

- Large thin wall cystic mass is noted occupying the abdominal cavity, measuring 8 x 10.7 x 12.5 cm
- Multiple thin septations are noted within the cystic mass
- It is seen to insinuate between the sigmoid mesocolon indenting the sigmoid colon
- Seen to displace the bowel loops laterally
- Inferiorly it is seen to reach upto the pelvis, indenting the dome of bladder
- Anteriorly it is seen in close proximity with the anterior abdominal wall and is seen to protude through the umbilical defect which measures 1.5cm
- Uterus is normal in size, anteverted
- Both ovaries are not well delineated with respect to the mass
- D/D 1. Lymphangioma 2. Ovarian cystic neoplasm

Treatment- Sigmoid resection and anastomosis

- Diagnostic laparoscopy was done:
  - large multiloculated cyst, unable to identify the organ of origin
  - Mild ascites noted
  - Converted to laparotomy due to no available working space
- Laparotomy done by pfannensteil incision:
  - Multiple cysts arising from sigmoid mesocolon noted on either side of sigmoid colon sharing a common vascular supply
  - Largest cyst measuring 10 x 10 x 12 cm
  - Bilateral ovaries, uterus, fallopian tubes were normal
  - 7 cm of sigmoid colon resected along with the mesenteric cysts in toto
- Sigmoido-sigmoidostomy done




INCHES
CENTIMETERS
Romsons

HISTOPATHOLOGY REPORT

GROSS

- Shows multiloculation
- One of the cysts within shows yellowish gelatinous material
- Outer and inner wall is smooth, appears thickened
- Shows hemorrhagic areas

MICROSCOPY

- Section from colon shows congestion
- Section from wall of cyst shows fibrofatty tissue lined by flattened epithelium, lymphoid follicle, blood vessel showing congestion and chronic inflammation

Diagnosis: **Lymphangioma of Mesentery of intestine**

0UTCOME

- Course in hospital was uneventful
- Patient was discharged on post op day 5 on full feeds
- In opd follow up, baby was asymptomatic, tolerating feeds well, wound healthy

Mesenteric cysts- de Perrot Classification

- Lymphatic origin (cystic lymphatic malformations)
- Enteric origin (duplication cysts)
- Mesothelial origin (cystic mesothelioma)
- Rarely, urogenital cysts, cystic teratoma

May or may not extend into the retroperitoneum

Cystic lymphatic malformations

- Term “Lymphangioma” is not used currently as it is not a tumor. Lymphatic malformation is a more appropriate term
- Etiology: Benign ectopic lymphatics which lack communication with the remainder of the lymphatic system
- Most common site: head, neck and axilla (previously called cystic hygromas)
- Next common site: Abdomen
  - ileal mesentery (50-70%)
  - Omentum (10-30%)
  - Mesocolon (10-30%)
  - Retroperitoneum (10%)
- can be locally invasive or can encroach on vital structures.

CLINICAL PRESENTATION

- Most are asymptomatic and incidentally detected
- Symptoms:
  - Abdominal pain is the most frequent symptom
    - Vague chronic dull aching/ Acute severe throbbing
  - Progressive abdominal distension
  - Subacute intestinal obstruction
- Malnutrition may be present in cases where intestinal lymphangiectasis is present
- On examination a palpable, freely mobile cystic mass may present of variable size

COMPLICATIONS

- Intestinal obstruction
- Volvulus
- Hemorrhage into the cyst
- Infection
- Rupture
- Torsion of cyst
- Obstruction of the urinary or biliary tract
- Malignancy (lymphangioendothelioma/ adenocarcinoma)

TREATMENT

- The Goal is a complete surgical excision with or without bowel resection (50-60% require bowel resection due to the blood supply)

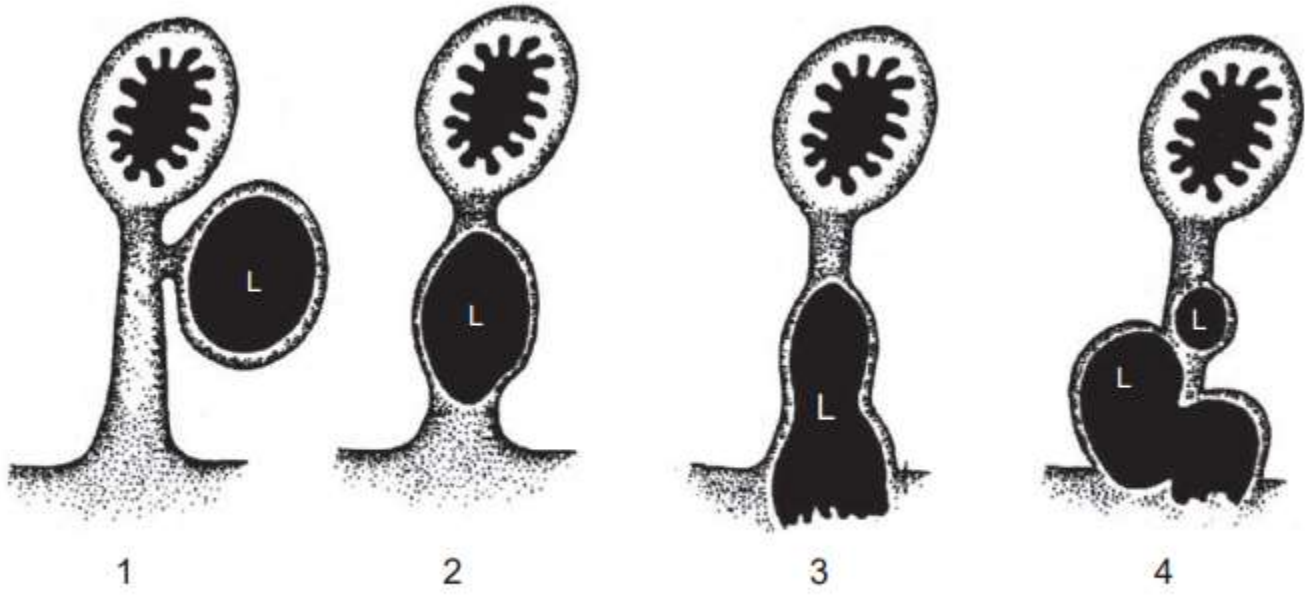


FIGURE 91-5 Classification of mesenteric cysts. Type 1—pedicled; easily resected. Type 2—sessile in leaves of mesentery; requires bowel resection. Type 3—extending into retroperitoneum; often incompletely resected. Type 4—multicentric; may require complex operations, sclerotherapy, or both. (From Losanoff JE, Richman BW, El-Sherif A, et al: Mesenteric cystic lymphangioma. J Am Coll Surg 2003;196:598.)

- In type 3 and 4 where complete excision is not possible, options are
  - Partial excision with marsupialization
  - Partial excision with sclerotherapy (25% glucose, ok432, tincture iodine, sterol, bleomycin)
- The intervention should be performed as soon as possible because of the risk of infection, torsion, hemorrhage, or obstruction

FOLLOW-UP

- Prognosis is excellent when the complete resection is feasible. However, relapses may occur if vesicles or part of the lesion remain unresectable
- Patient to be followed up every 3 months over a period of 2 years with USG abdomen to look for recurrence

MESENTRIC LYMPHANGIOMA

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Junior Resident- Pathology
Dr. R. G. W. Pinto
Professor and HOD Pathology department
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CASE

- A 10 months old female presented to OPD with progressive abdominal distension in the past 15 days.
- Investigation CBC, LFT, RFT and serum alpha fetoprotein were WNL.
- Serum beta-hCG level < 1.2mlU/ml.

Radiological Findings

- **On USG:** A large solid cystic mass noted in the abdomen measuring approx 12x16cm, seems to reach epigastrium , superiorly and pelvic region indenting the dome of bladder, inferiorly.

- Advised CE-CT for further evaluation.

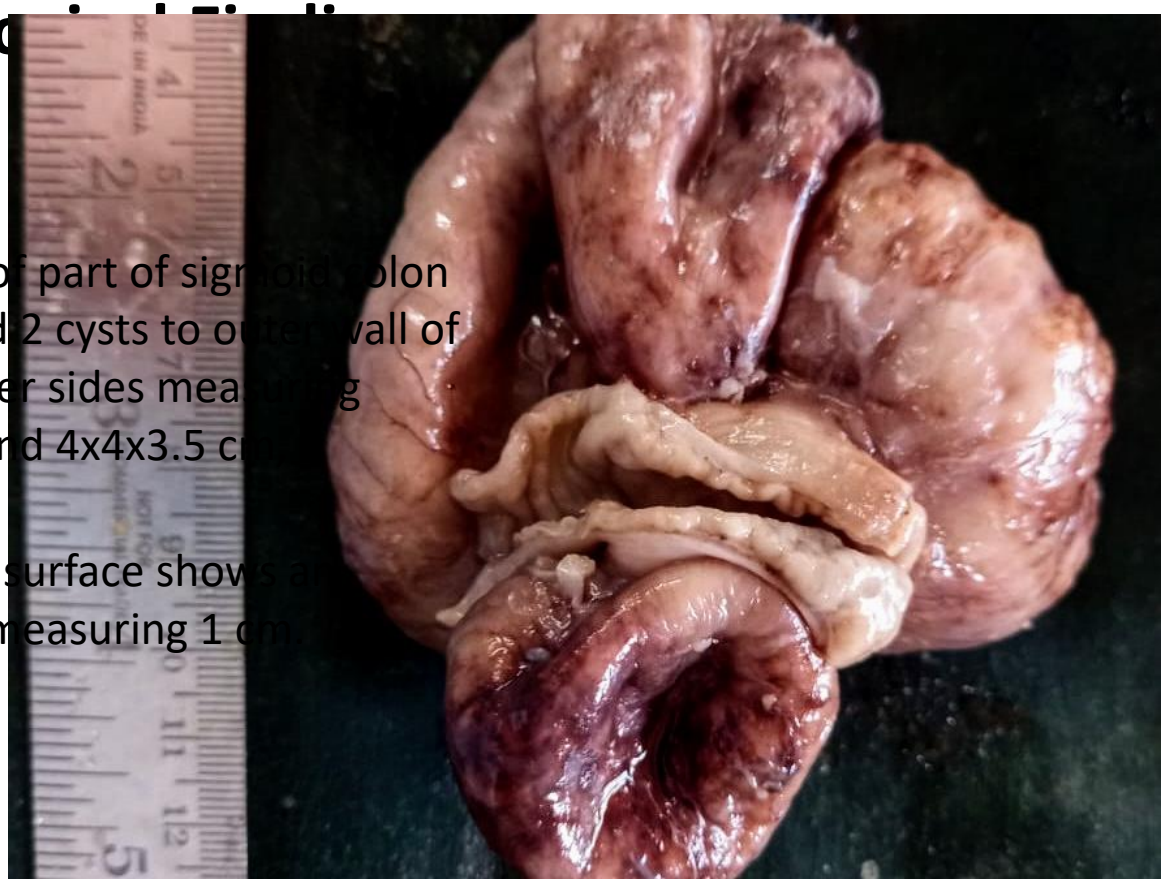
- **On CECT abdomen:** A large thin walled cystic mass with septae is noted occupying the abdominal cavity. The differential diagnosis is given:

  1. Lymphangioma
  2. Ovarian cystic neoplasm

OT Findings

- A large mutiloculated cystic away from sigmoid colon on either side of sigmoid colon showing common wall.
- 10- 15ml serous fluid aspirated.

Histopaholo

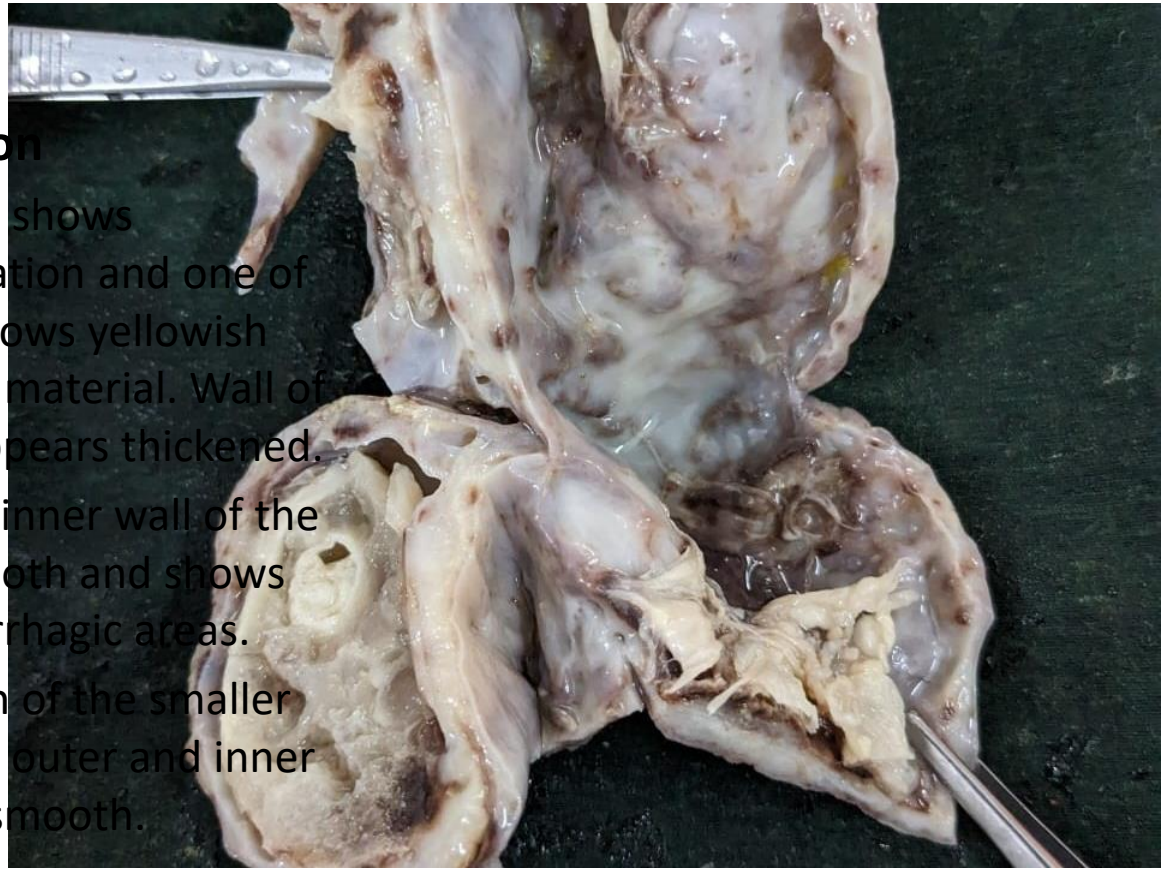


On Gross:

- A specimen of part of sigmoid colon with attached 2 cysts to outer wall of colon on either sides measuring 9x6x3.5 cm and 4x4x3.5 cm respectively.
- Larger cystic surface shows a perforation measuring 1 cm

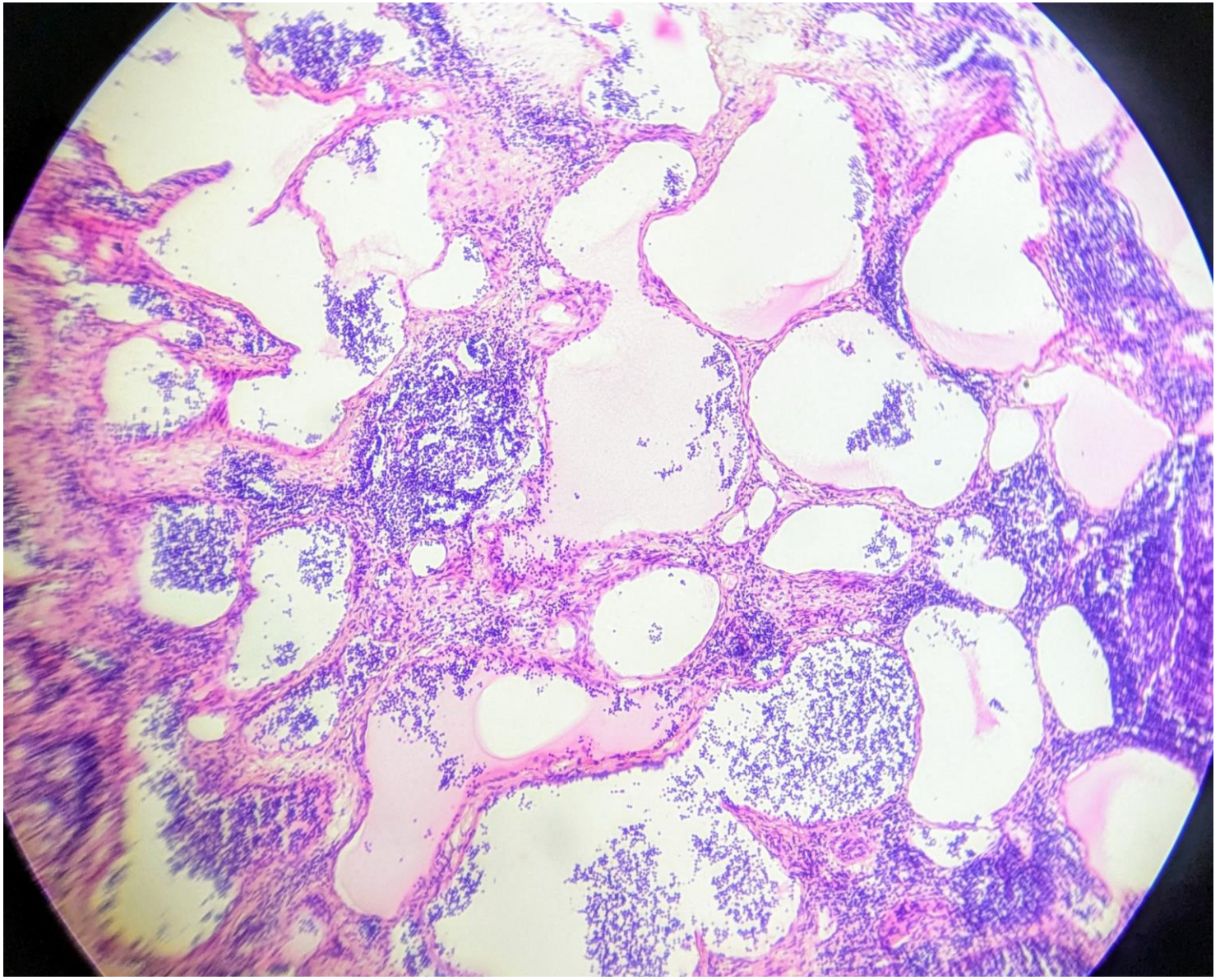
On Cut section

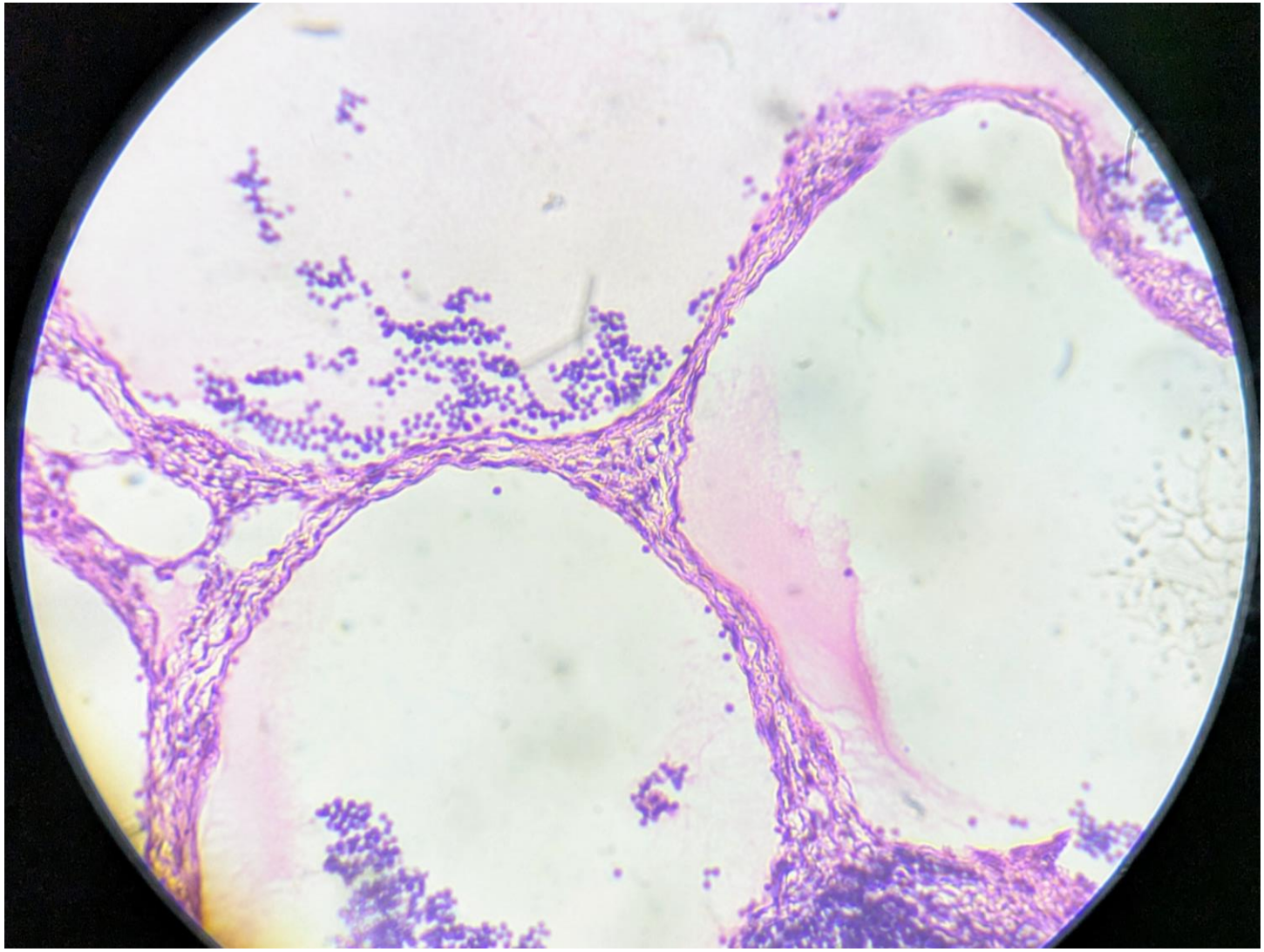
- Larger cyst shows multiloculation and one of the cyst shows yellowish gelatinous material. Wall of the cyst appears thickened.
- Outer and inner wall of the cyst is smooth and shows few hemorrhagic areas.
- Cut section of the smaller cyst shows outer and inner surface is smooth.

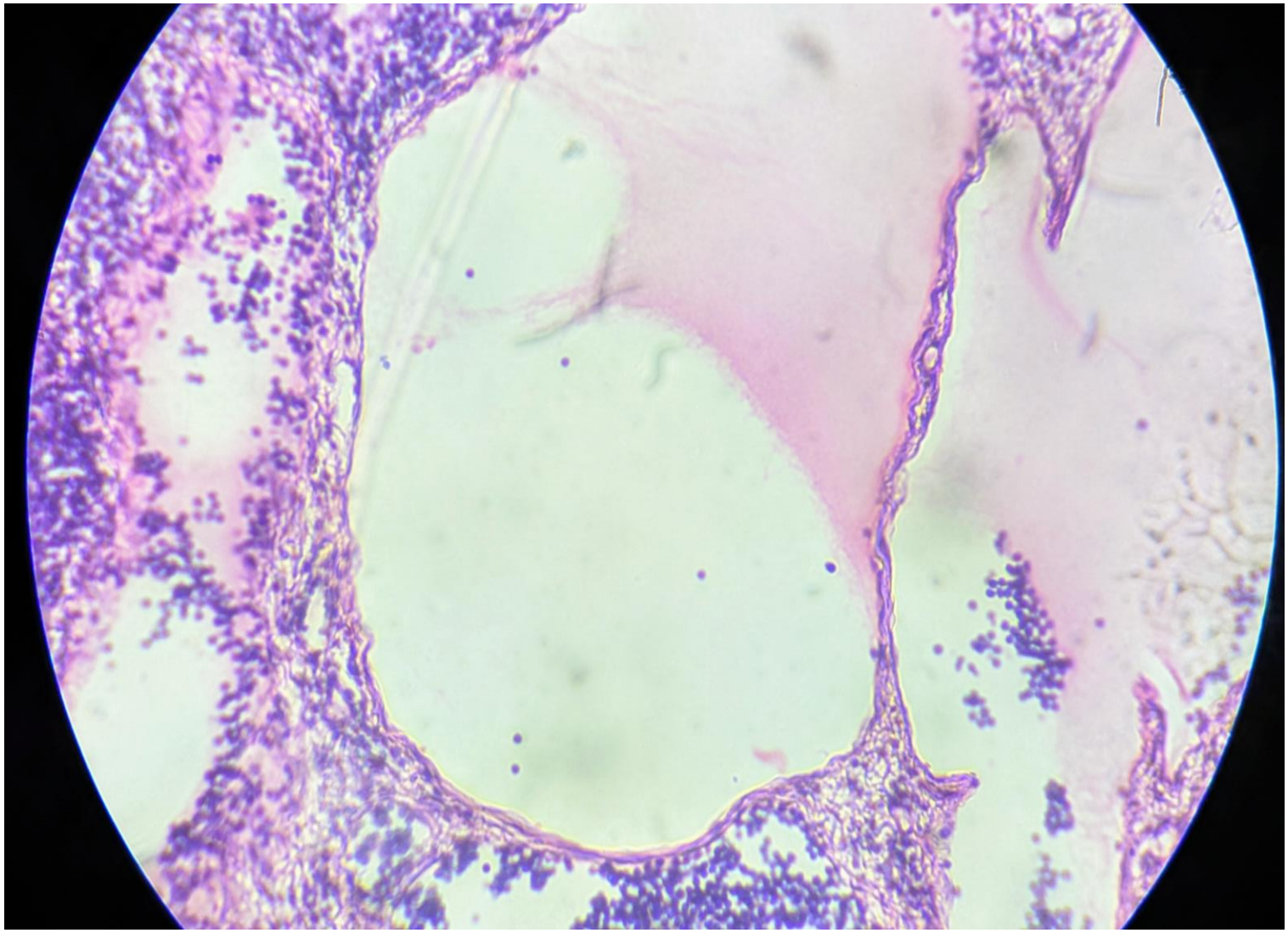


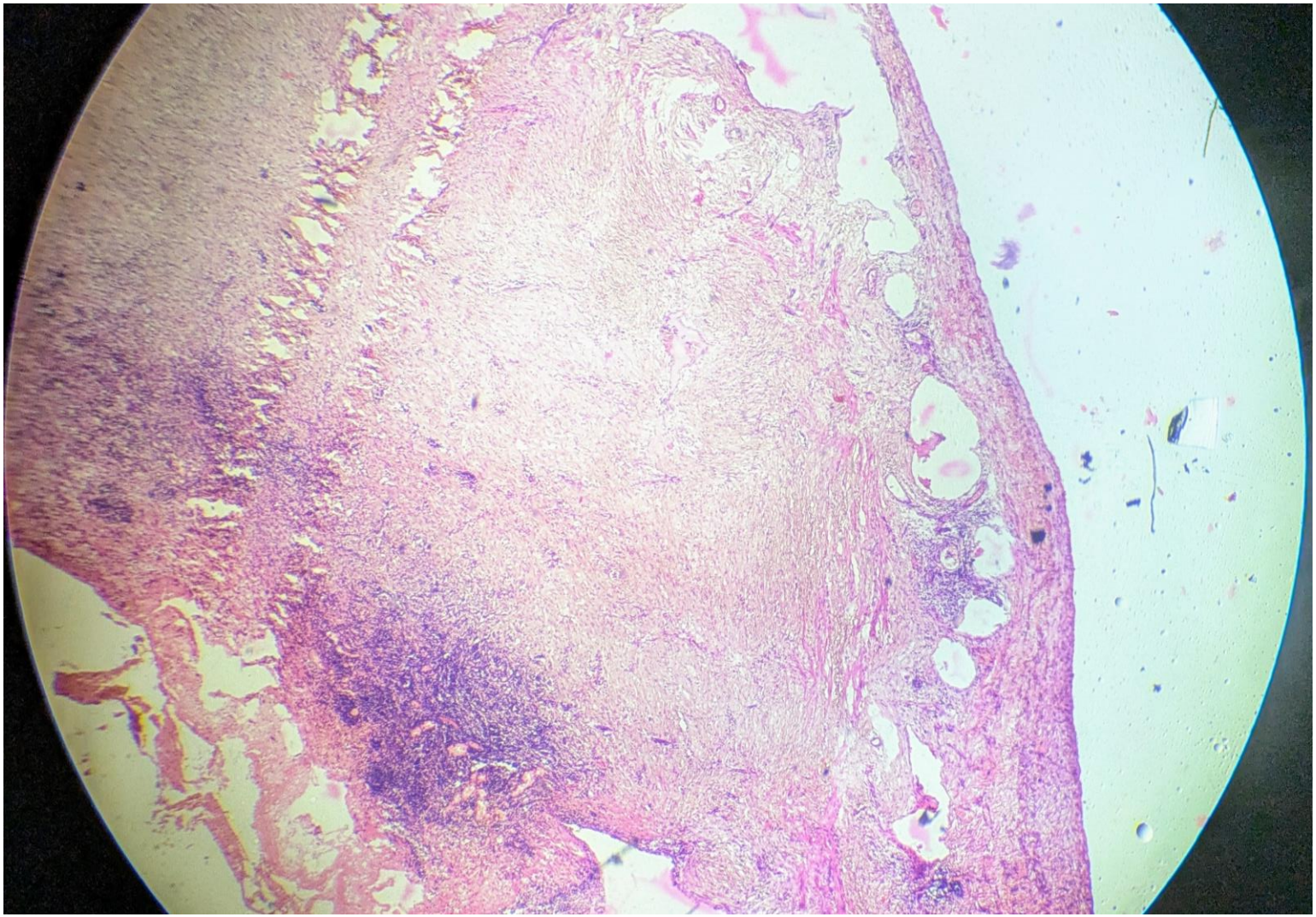
- **On Microscopy**

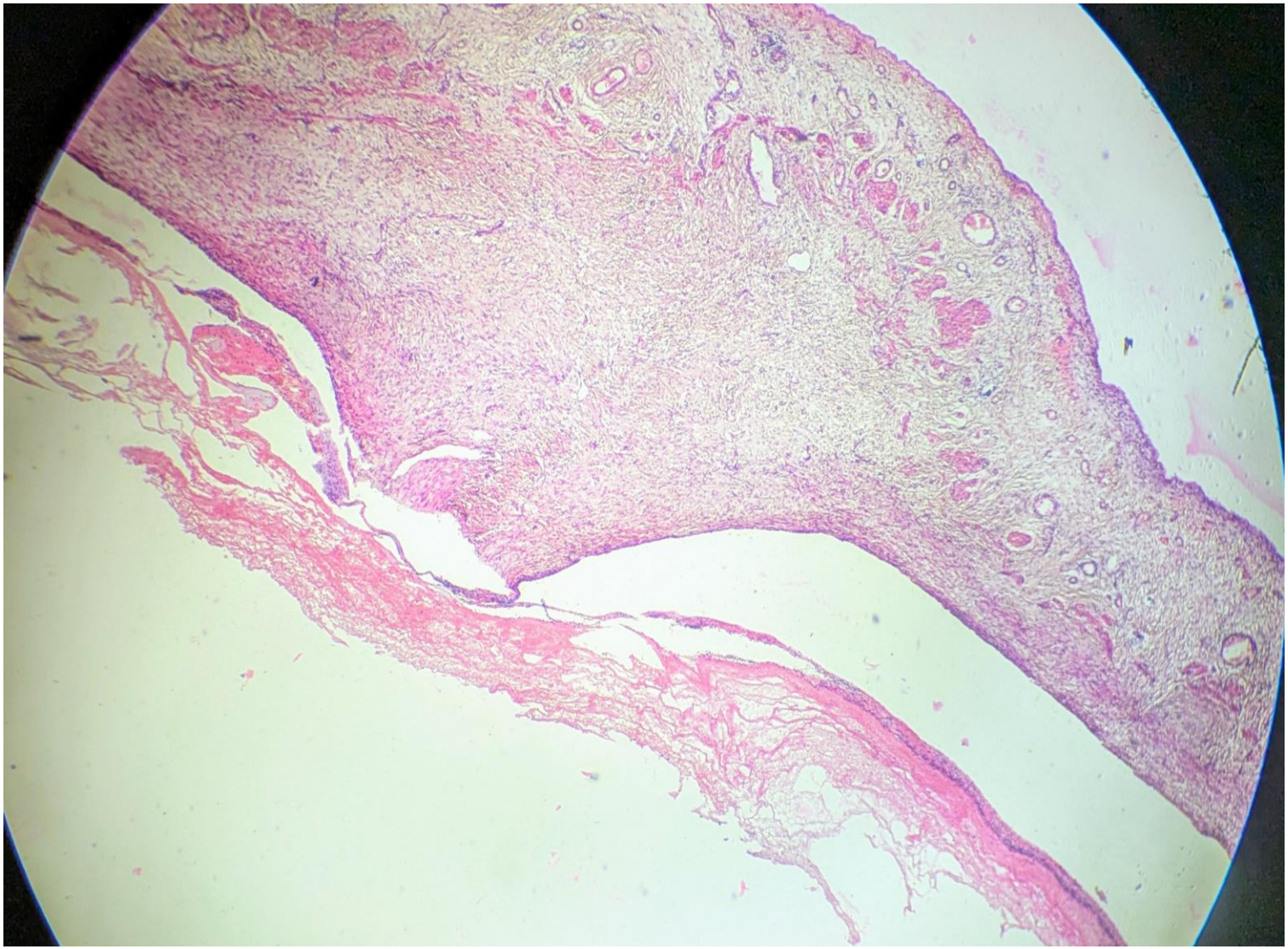
1. Multiple sections shows large irregular cystic spaces lined by flattened, bland epithelium and surrounded by few lymphoid follicles, few congested blood vessels and chronic inflammatory infiltrates.
2. Few cystic spaces contained eosinophilic acellular material (lymph) and they reach upto serosa of the bowel.
3. Section from colon shows congestion.
4. Sections from the margins are unremarkable.

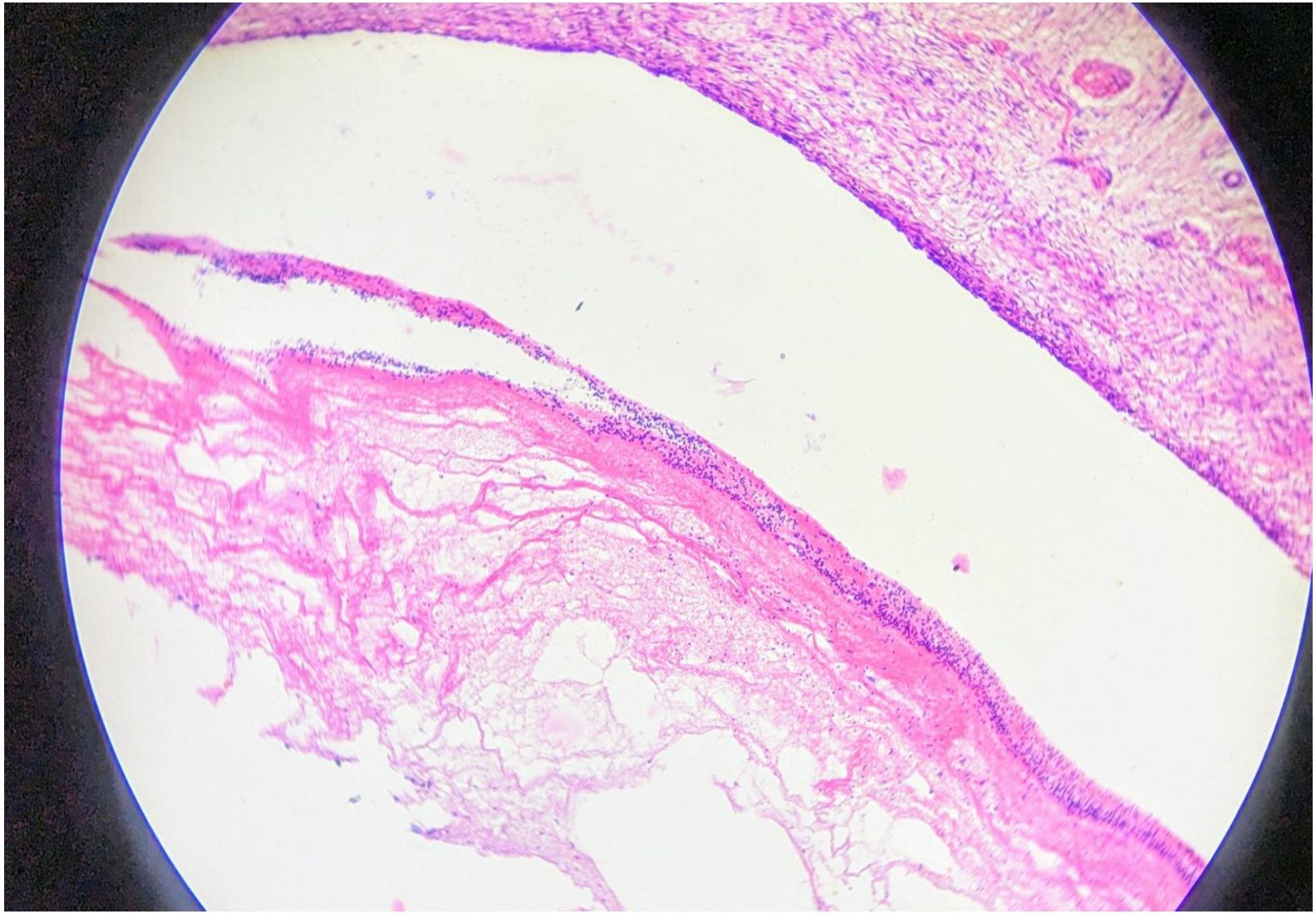


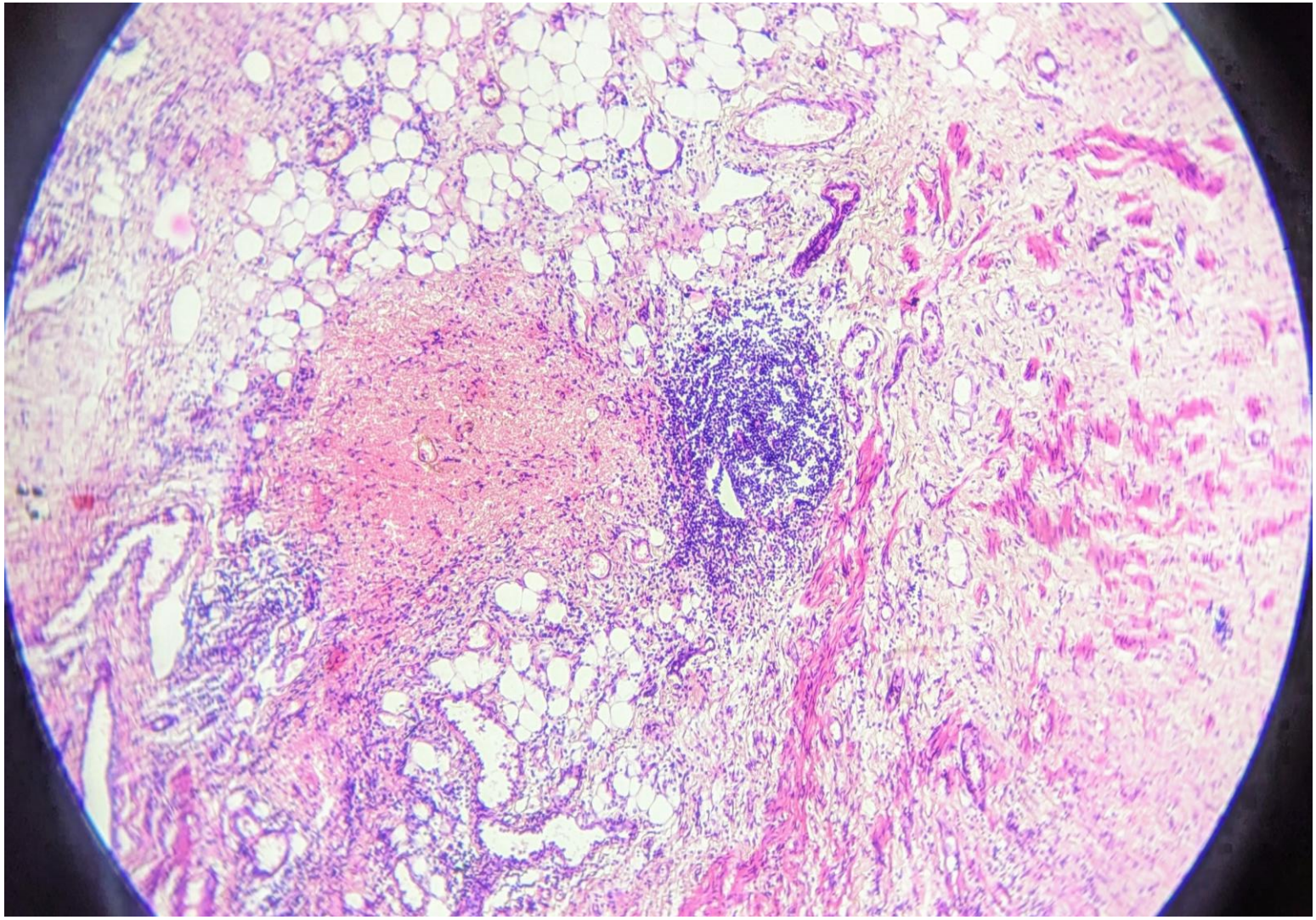












Discussion

- **Introduction**

- Lymphangioma is an uncommon congenital malformation of the lymphatic system that manifest as benign tumor.
- Lymphangioma are most commonly seen in children and rarely in adults.
- Mesenteric lymphangiomas are cystic lesion in nature and are therefore known as mesenteric cyst.
- Incidence of intra-abdominal cystic lymphangiomas is low i.e <1/1,00,000.

- **Site**

- Most common site is head, neck and axilla and called as cystic hygroma.
- Intra-abdominal lymphangiomas are commonly located in small bowel mesentery (66% cases) followed by ascending and transverse colon mesentery (33% cases).
- Less than 1% of cases have been reported in the mesentery of descending colon, sigmoid or rectum.

- **Etiopathogenis**

- Lymphangiomas are commonly caused by congenital abnormality of the lymphatic system which leads to sequestration of the lymphatic tissue during embryological development.
- However, Abdominal trauma, lymphatic obstruction , inflammatory causes, surgery, or radiation therapy can also lead to the formation of such tumor.

- Lymphangiomas are classified as

1. Simple lymphangioma
2. Cavernous lymphangioma
3. Cystic lymphangioms

- Simple lymphangiomas are situated superficially in the skin and are composed of small thin walled lymphatic vessels.
- Cavernous lymphangiomas are composed of dilated lymphatic vessels and lymphoid stroma and have connections with spaces of various normal adjacent lymphatics.
- Cystic type consist of lymphatic spaces of various sizes that contains fascicles of smooth muscle and collagen bundles, but has no connections with adjacent normal lymphatics.
- However, cystic lymphangiomas is not always clearly differentiated from cavernous type because the cystic type may also contain cavernous areas.

- **Clinical features**
- Most are asymptomatic and found incidentally.
- Non- specific symptoms like abdominal pain, vomiting and constipation.
- **Investigations**
- Radiological Investigations are **gold standard** for diagnosis of mesenteric lymphangiomas. Majority of lymphangiomas are discovered incidentally on imaging.
- **Treatment**
- Surgical excision of the tumor is gold standard method of treatment of lymphangiomas.

THANKYOU