ADULT INTUSSUSCEPTION WITH- A RARE ETIOLOGY

PRESENTER-

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CASE PRESENTATION

- 60y/F with no known comorbidities presented with:
- Right side lower **abdominal pain** since 10 days
- Multiple episodes of bilious **vomiting** since 3days
- Constipation since last 3 days
- No h/o fever, hematochezia/melena or weight loss.
- No surgical history in the past

ON CLINICAL EXAMINATION

- Conscious, oriented
- Vitally stable
- Afebrile

- P/A- soft, non-tender, diffuse abdominal distension with sluggish bowel sounds, no signs of peritonitis, no palpable mass.

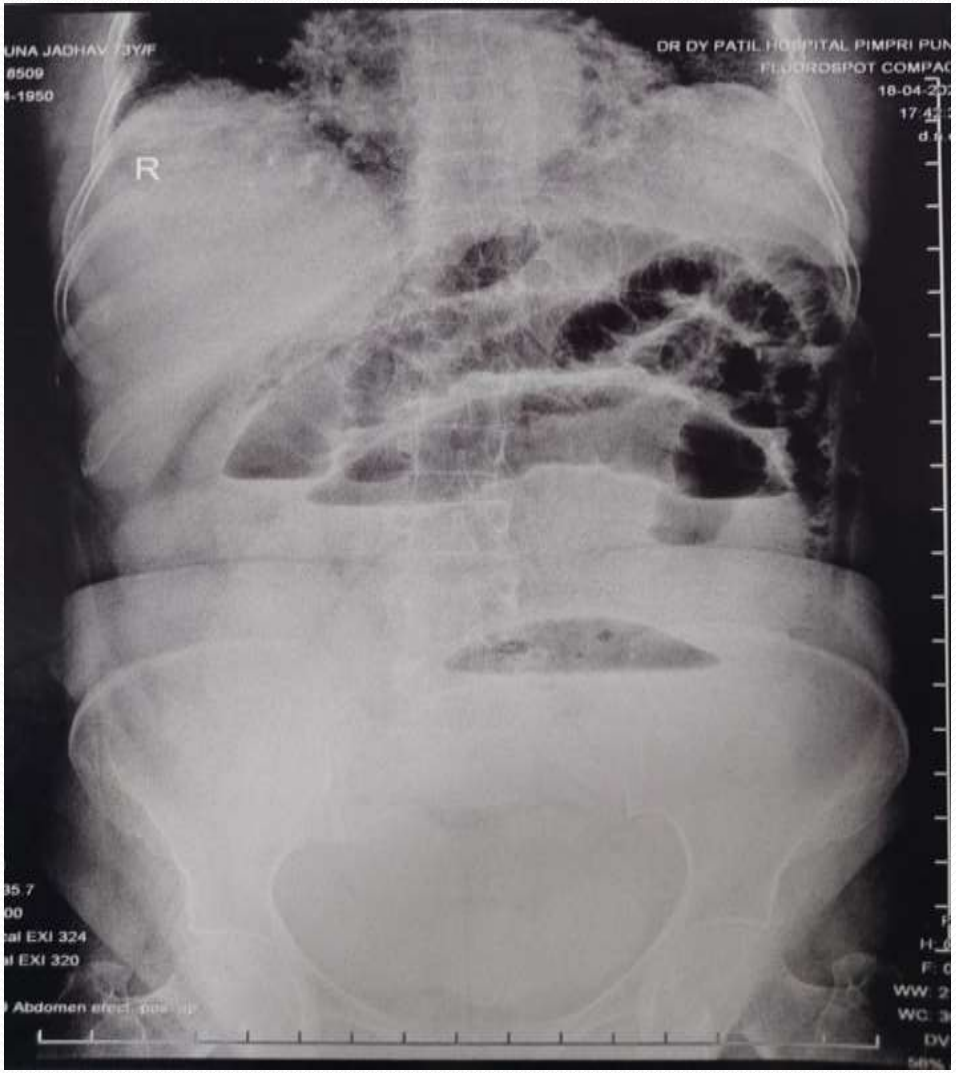
- P/R- Normal

- Examination of all other systems were grossly normal.

PLAIN ABDOMINAL RADIOGRAPH

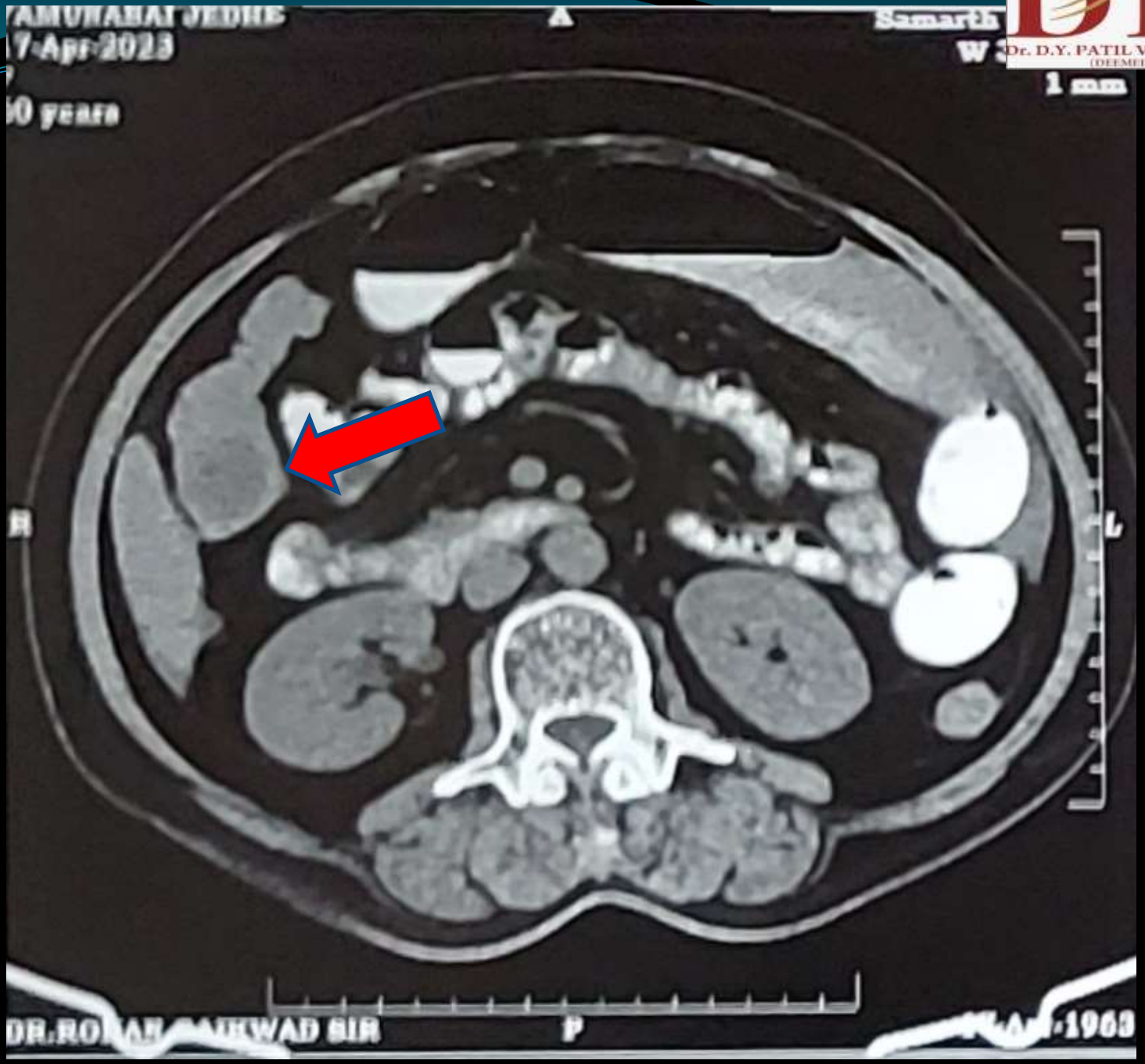
Xray abdominal erect: abdomen + pelvis shows

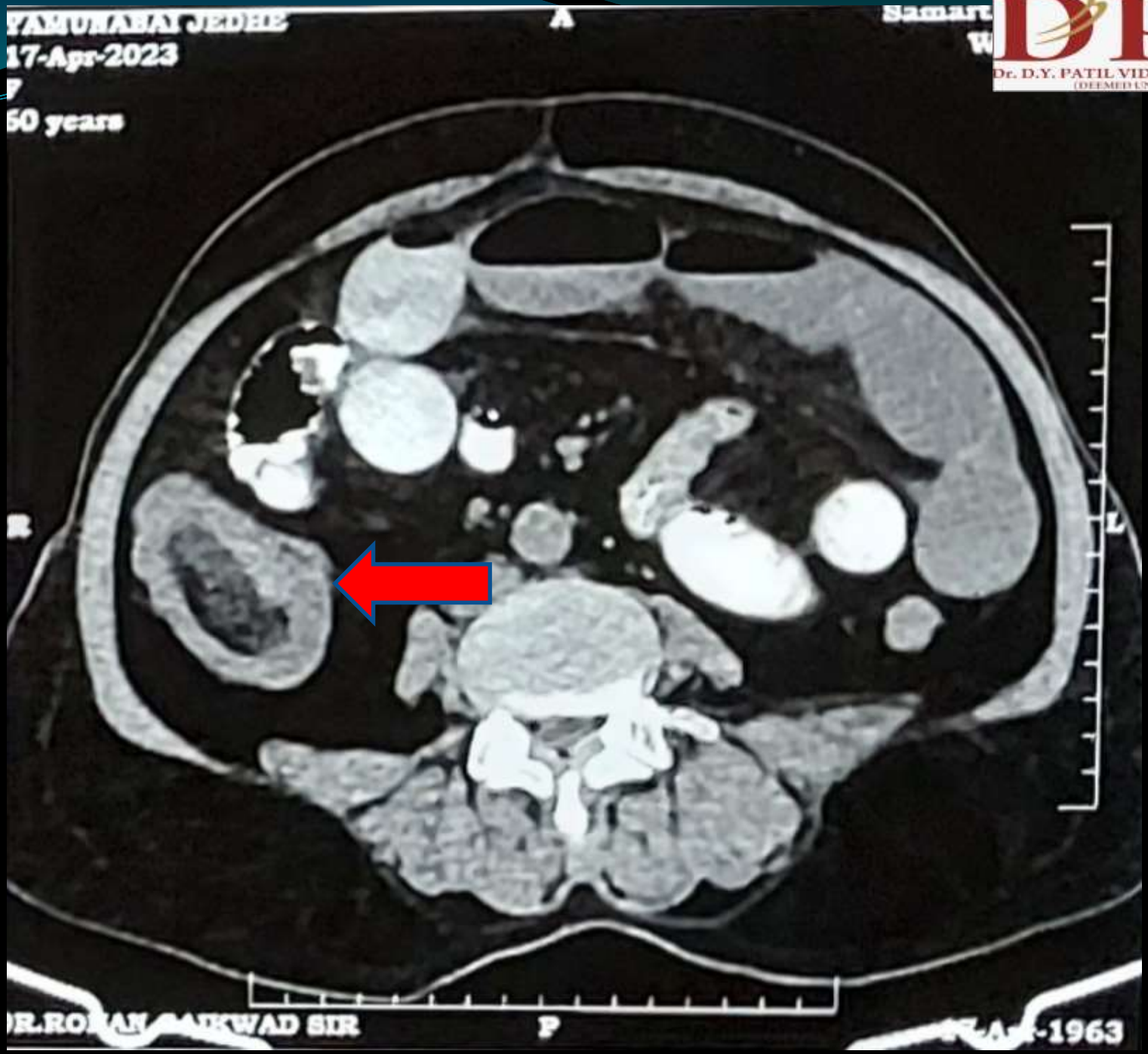
Multiple air-fluid levels arranged in a **step-wise pattern** without evidence of colonic gas.

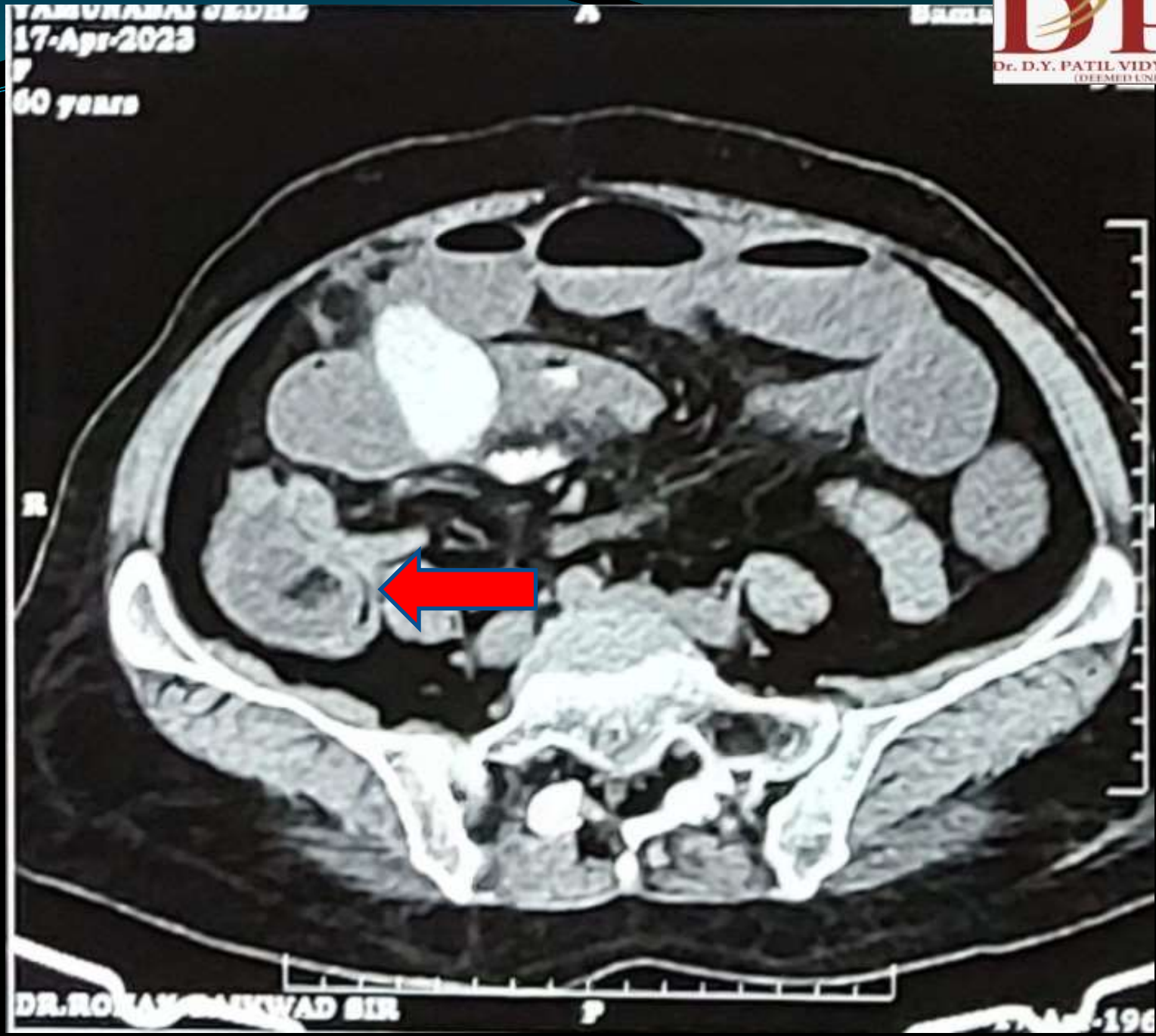


CECT ABDOMEN + PELVIS-

- **Solid nodule- 2.7x2.9cm,** homogenous, post contrast enhancement in the sub mucosal region of ascending colon at the hepatic flexure which is **protruding into the colon lumen appearing as the polypoidal mass**.
- Telescoping of ileal segments into cecum.
- **Intussusception** involving the segment of ascending colon and I-C junction along with the mesentry and appendix leading to Sub- acute small bowel obstruction.
- More likely possibility of large bowel lymphoma.




17-Apr-2023
50 years
A
R
L
P


17-Apr-2023
60 years
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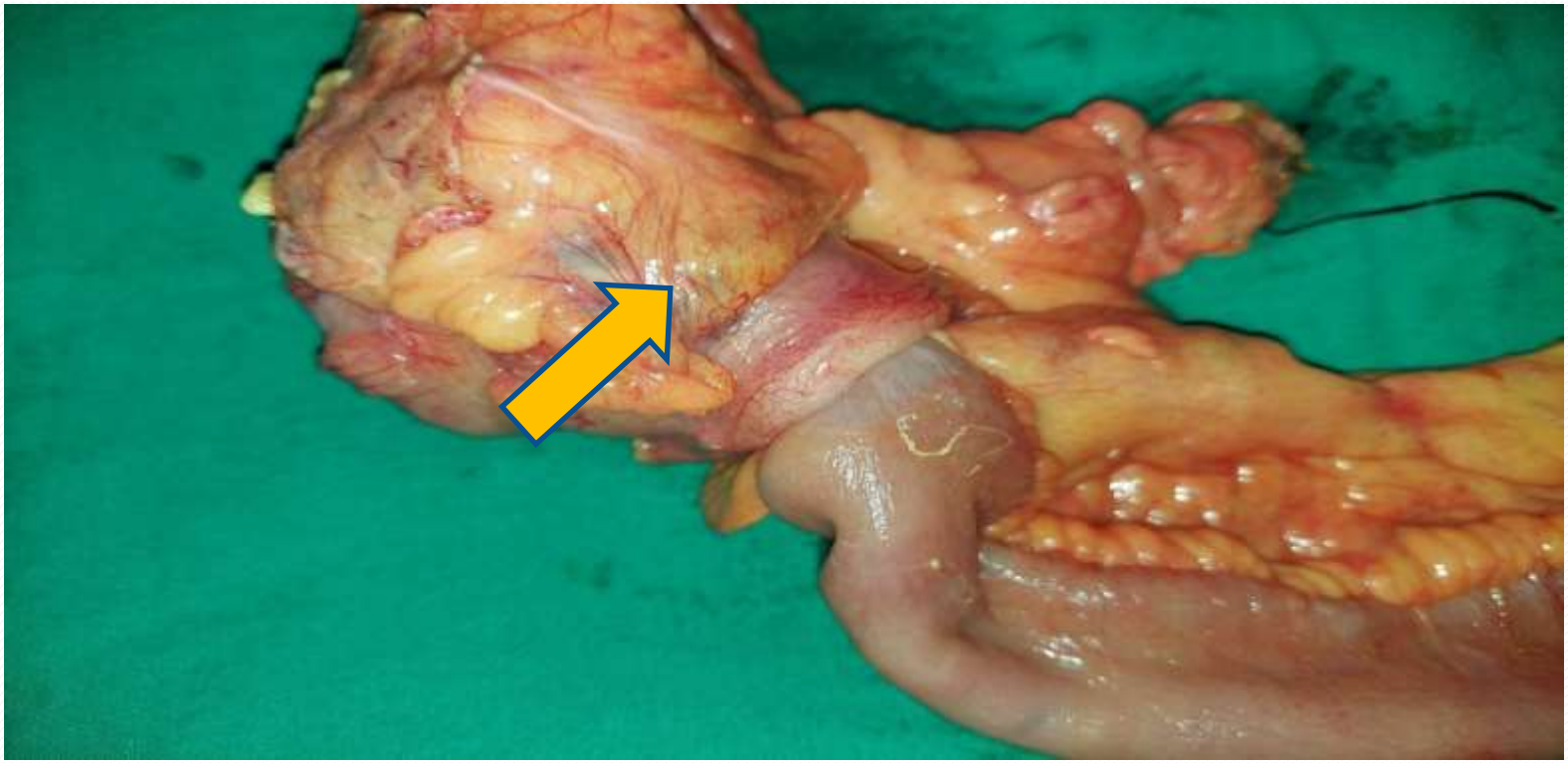
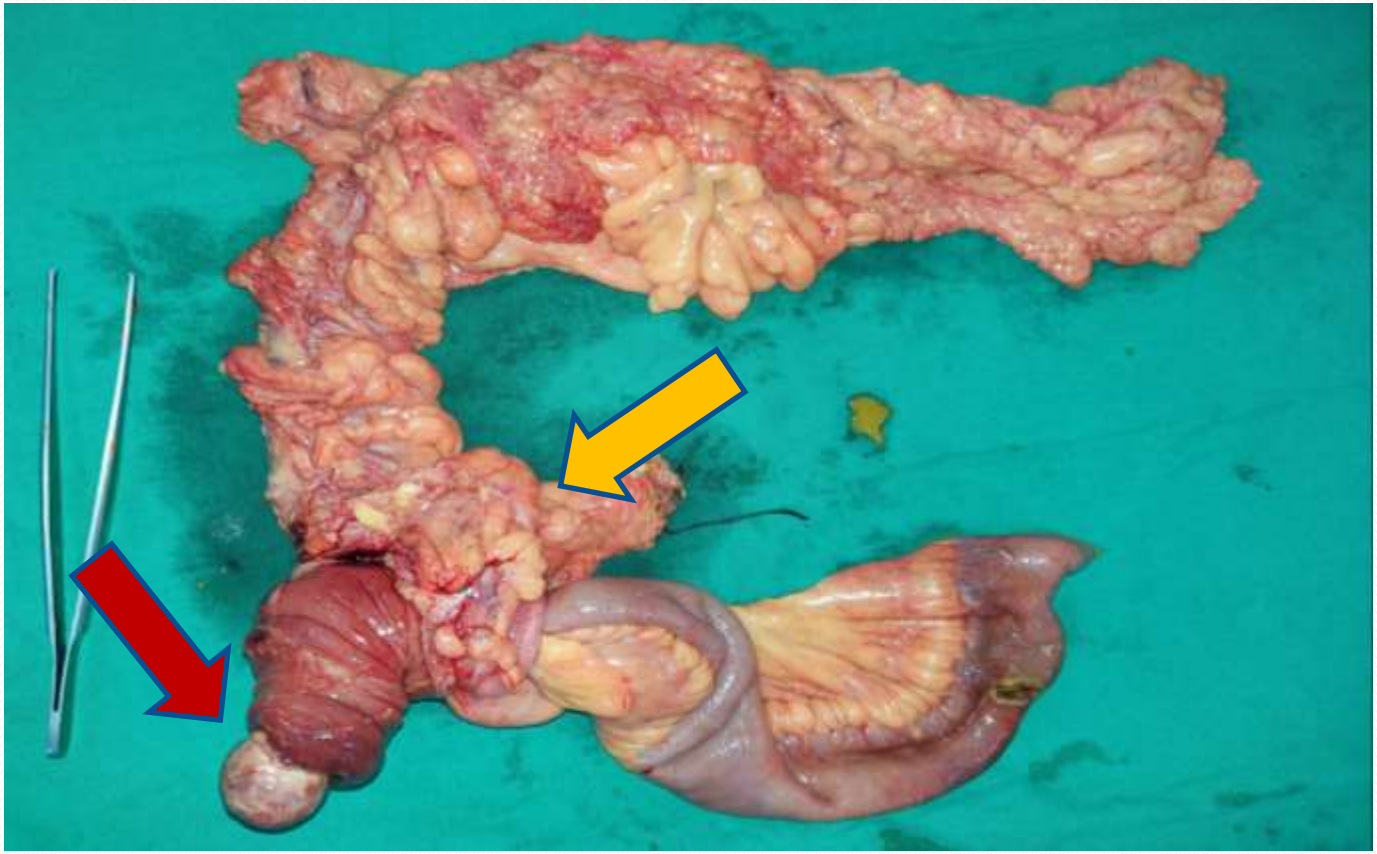
INTRAOPERATIVE IMAGE

EVIDENCE OF STRICTURE WITH INTUSSUSCEPTION



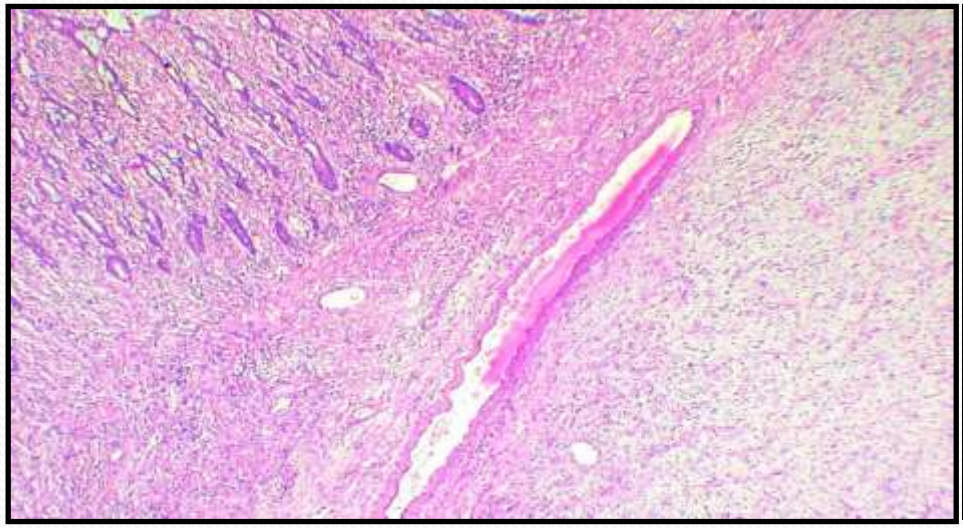
SPECIMEN OF RIGHT HEMICOLECTOMY

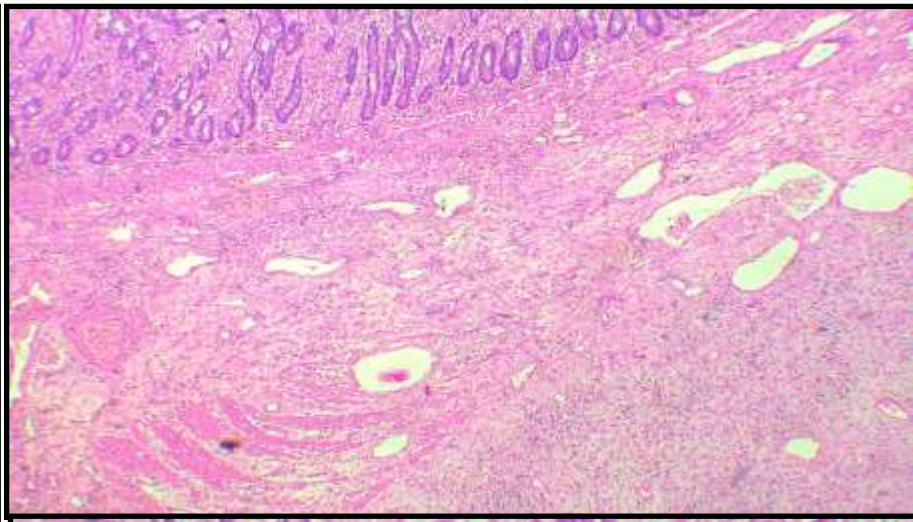


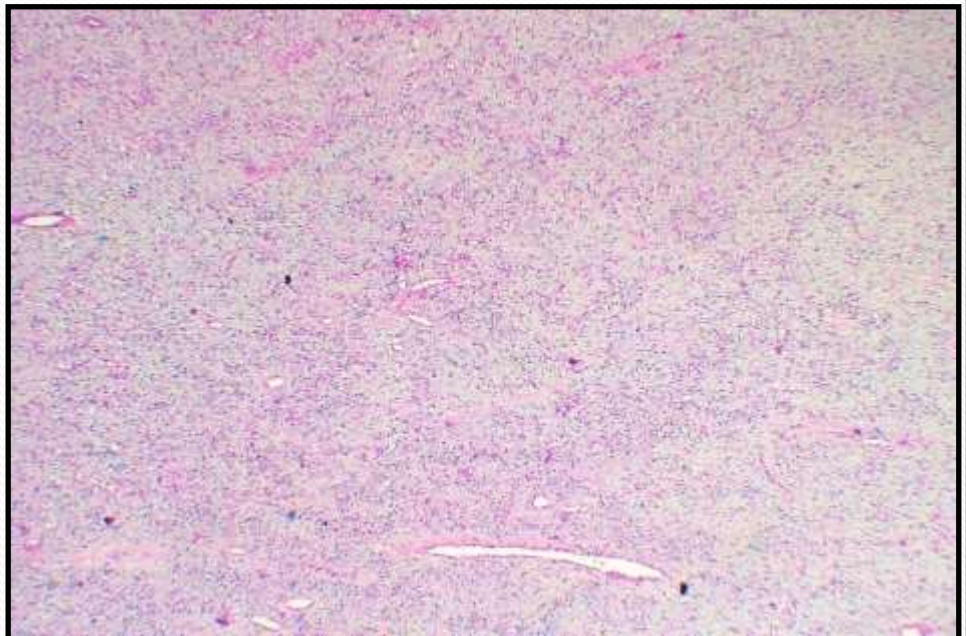
GROSS

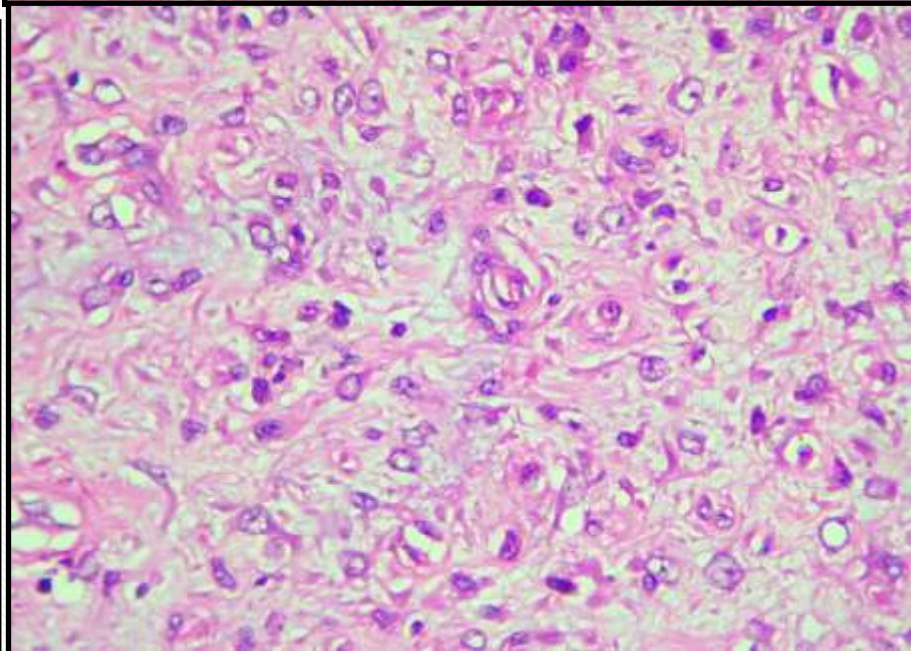
- **Right hemicolectomy specimen- 70 cms**
- **Ileal segment – 42 cm, appendix - 2.5 cm.**
- **Intussuscepted segment - 9.5 cm with markedly congested mucosa**
- **Nodular mass at the tip of ileum - 2.5 cm in diameter.**
- **35 lymph nodes were dissected.**

Microscopy- Lesion in ***submucosa*** composed short fascicles and bundles of ***spindle cells arranged in storiform pattern*** with scanty cytoplasm. ***Mitotic activity less than five.***









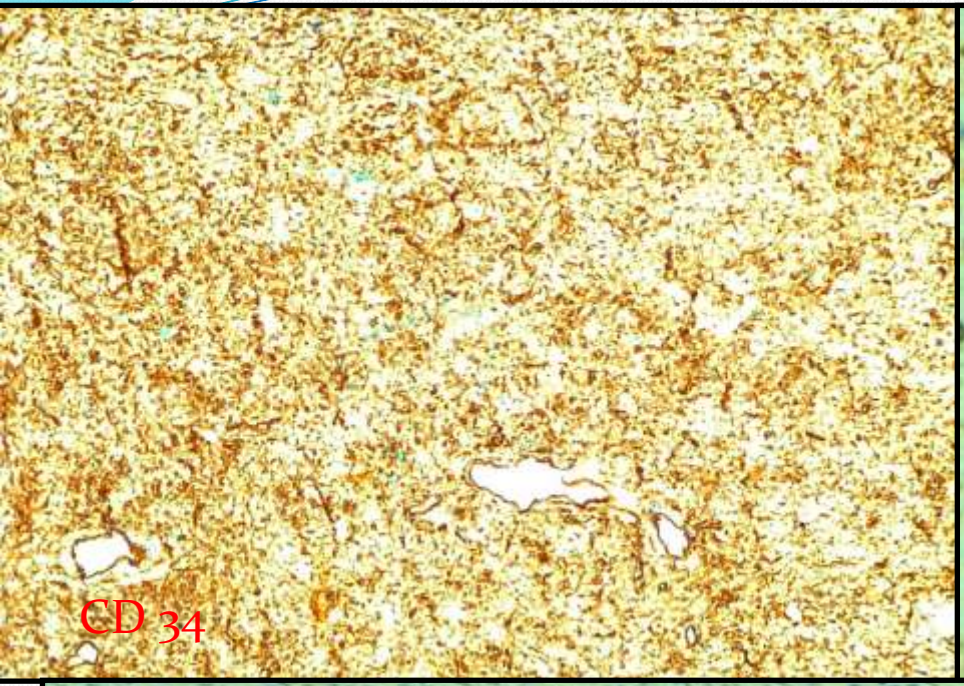
DIFFERENTIAL DIAGNOSIS

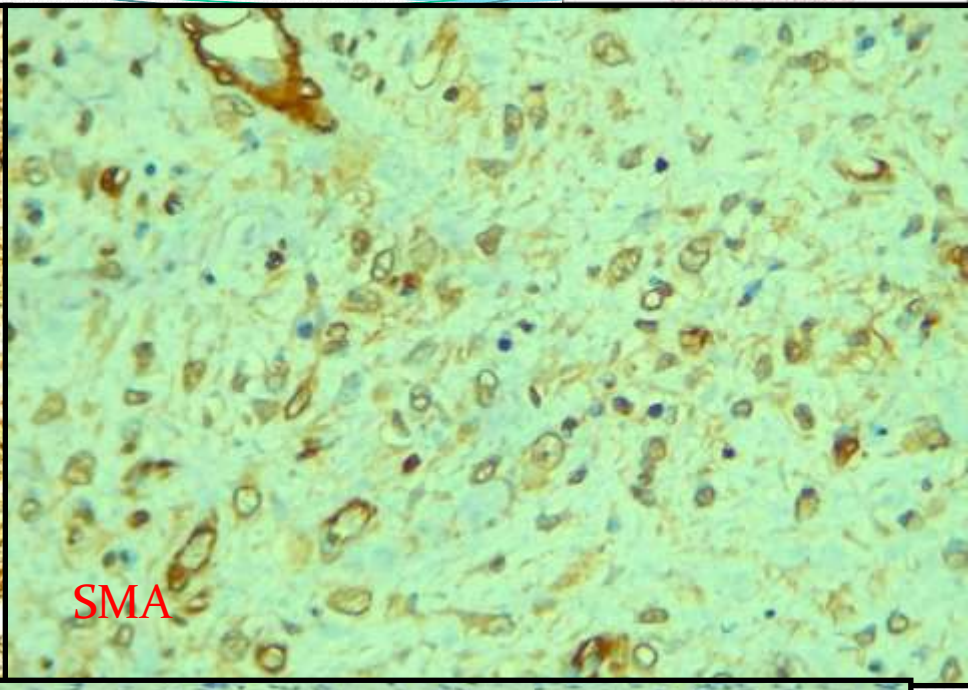
1. Gastro Intestinal Stromal Tumor (GIST)
2. Inflammatory Fibroid Polyp (IFP)
3. Inflammatory Myofibroblastic Tumor (IMFT) / Inflammatory Pseudotumor

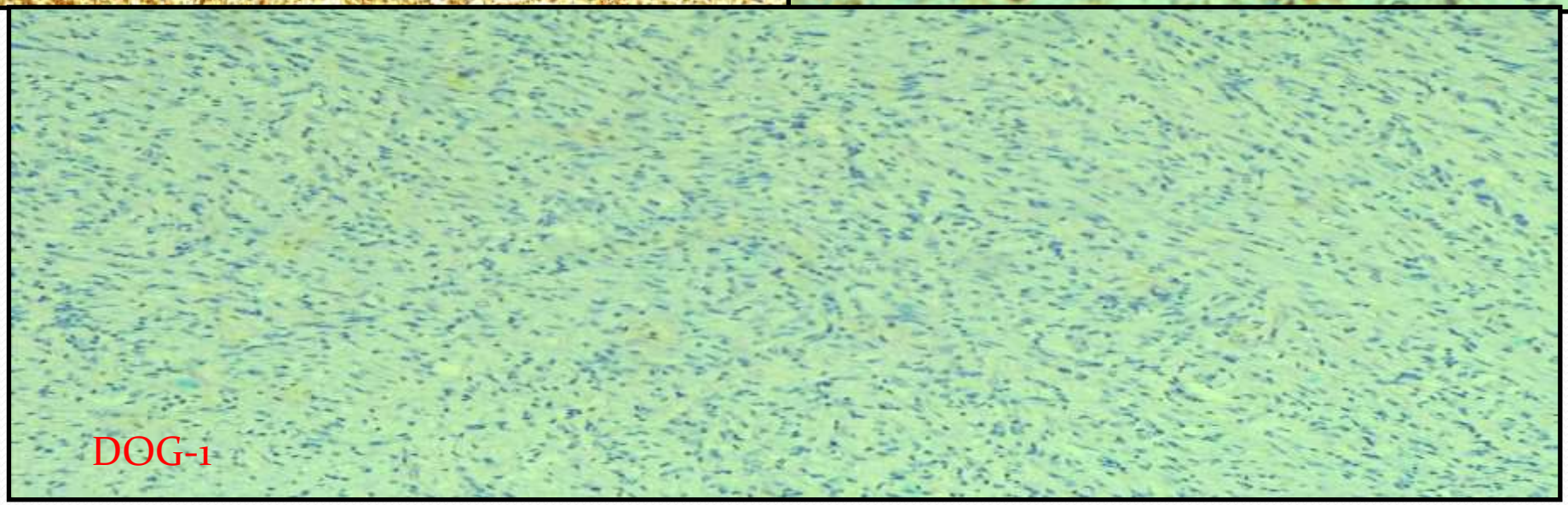
IHC-

- CD 117- Negative
- DOG1- Positive
- CD 34- Diffusely Positive
- SMA- Focally Positive
- S 100- Negative

DIAGNOSIS- Gastrointestinal Stromal Tumor of Ileum with low malignant potential


CD 34


SMA


DOG-1

Tumor Size	Mitotic Rate[2]	Predicted Biologic Behavior	Risk Per CAP[2]
≤2 cm	≤5 mitoses/50 HPFs	Metastasis rate: 0%	None
	>5 mitoses/50 HPFs	Metastasis rate: 50%–54%	Insufficient data - High (54%)
>2 cm to ≤5 cm	≤5 mitoses/50 HPFs	Metastasis rate: 1.9%–8.5%	Low (4.3%–8.5%)
	>5 mitoses/50 HPFs	Metastasis rate: 50%–73%	High (50%–73%)
>5 cm to ≤10 cm	≤5 mitoses/50 HPFs	Metastasis rate: 24%	Insufficient data - Moderate (24%)
	>5 mitoses/50 HPFs	Metastasis rate: 85%	Insufficient data - High (85%)
>10 cm	≤5 mitoses/50 HPFs	Metastasis rate: 34%–52%	High (34%–57%)
	>5 mitoses/50 HPFs	Metastasis rate: 71%–90%	High (71%–90%)

Data from Miettinen M, Lasota J. Gastrointestinal stromal tumors: pathology and prognosis at different sites. Sem Diag Path 2006;23:70-83.

DISCUSSION

- GISTS are commonest mesenchymal neoplasm of GI tract (**1% to 2%** of GI malignant tumors).
- Derived from **Interstitial cells of Cajal (ICC**)
- **ICC- pacemaker** of GI motility, providing an interface between autonomic nerve stimulation and the muscle layer of the gastrointestinal wall.
- The hallmark of most GISTS is their **positivity for KIT(CD117) & DOG-1.**
- GISTs occur most commonly in the **stomach (40% to 60%).**
- GISTs located in small intestine is much rarer, and it represents only 20-25% of all cases.

Zhiqiang Liu et al Hindawi Gastroenterology Research and Practice Volume 2019, Article ID 3849850, 10 pages https://doi.org/10.1155/2019/3849850

- In a recent population-based analysis-
- ***10,046 GIST*** patients data was collected(2000 to 2015) based on Surveillance, Epidemiology, and End Results (SEER) database.
- Patients demographics, tumor characteristics, incidence, treatment, and survival were retrieved for analysis.
- Only ***25% were small intestinal*** GISTs and rest 75% were GIST from other sites.

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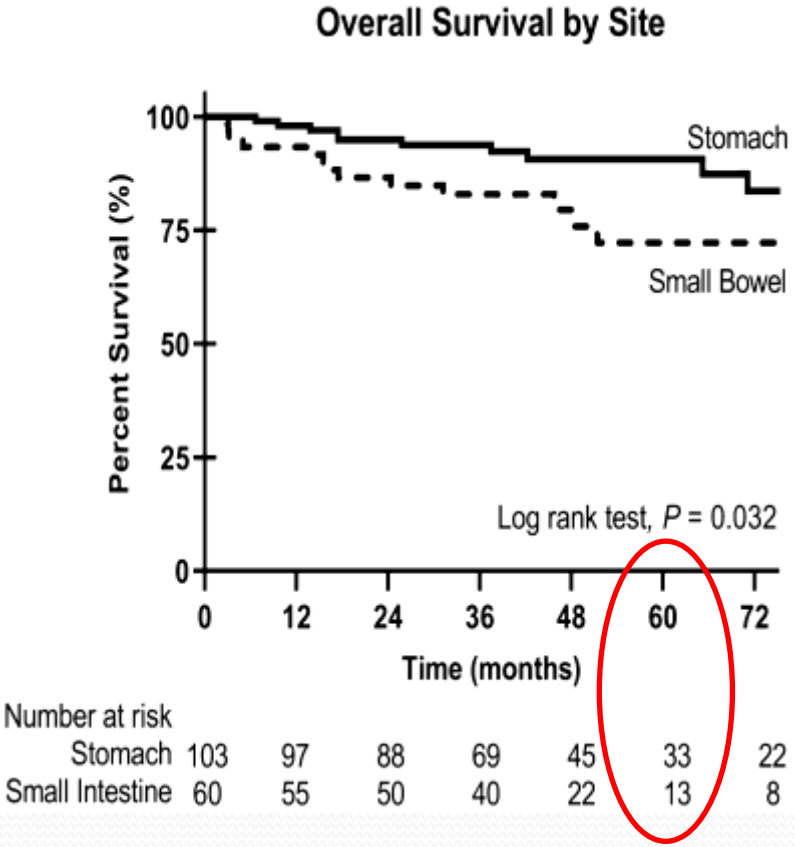
https://doi.org/10.1155/2019/3849850

- In a case series of **244 patients**, with wild-type *KIT* and *PDGFRA* data showed
-
- The primary site was **stomach (57.0%)**, followed by **small intestine (26%**), colon (3.3%), esophagus (2.0%), rectal (2.0%), and appendix (0.4%)
- 60 small intestinal GISTs were documented 45% duodenum, **55% were jejuno-ileal.**



Khan, T.M., Verbus, E.A., Rossi, A.J. et al. Patient demographics, clinicopathologic features, and outcomes in wild-type gastrointestinal stromal tumor: a national cohort analysis. Sci Rep 12, 5774 (2022). https://doi.org/10.1038/s41598-022-09745-1

- In a retrospective analysis of 32 cases with Small bowel GISTs, 4 tumors were located in the duodenum, 21 in the jejunum, and 7 in the ileum**.**

- Immunohistochemistry showed positive expression for CD117 (100%), CD34 (81.2%), DOG1 (93.8%).

Zhou L, Liao Y, Wu J, Yang J, Zhang H, Wang X, Sun S. Small bowel gastrointestinal stromal tumor: a retrospective study of 32 cases at a single center and review of the literature. Ther Clin Risk Manag. 2018 Aug 22;14:1467-1481. doi: 10.2147/TCRM.S167248. PMID: 30174429; PMCID: PMC6110632

- The usual IHC panel used for GIST includes *CD 117 and DOG 1, CD34, S-100, SMA..*
- In 1998, Kindblom **et al** reported that GIST expresses KIT tyrosine-kinase receptor. Since then, it is defined as KIT-expressing mesenchymal tumors of the GI tract.
- CD117(C KIT) a Kit protein encoded by C-KIT gene, an epitope of tyrosine kinase receptor(70%).
- CD 117 negative is typical of PDGFRA mutated GISTS(10%)
- These mutations can be Exon 11(70%), **exon 9(<10%)** of the KIT oncogene

.

Kindblom LG, Remotti HE, Aldenborg F, Meis-Kindblom JM. Gastrointestinal pacemaker cell tumor (GIPACT): gastrointestinal stromal tumors show phenotypic characteristics of the interstitial cells of Cajal. Am J Pathol. 1998;152:1259–1269. Corless C.L., Heinrich M.C. Molecular pathobiology of gastrointestinal stromal sarcomas. Annu Rev Pathol. 2008:557–586.

- Genotyping confirms KIT/PDGFRA-mutated GIST or to assess for WT GISTs.

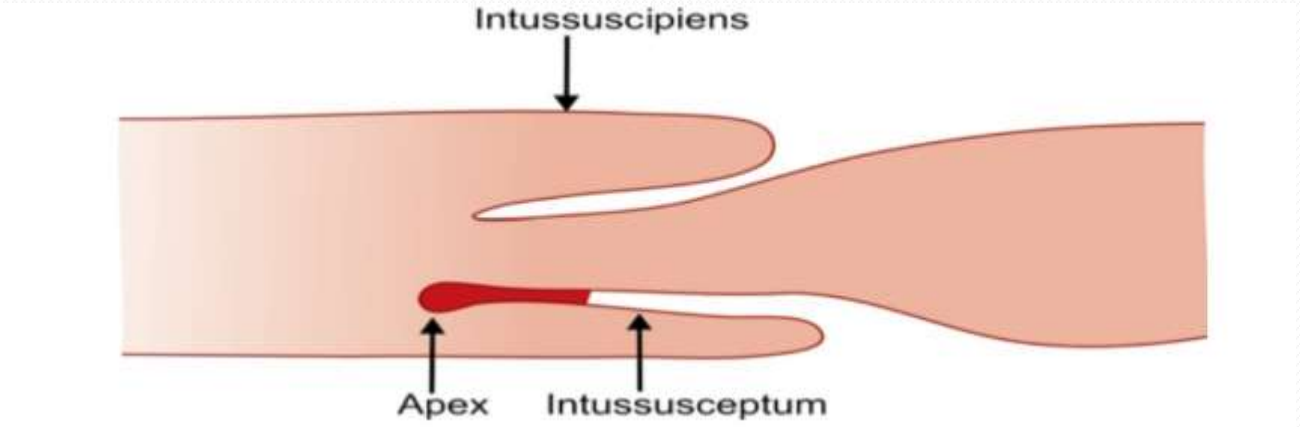
- In KIT and PDGFRA negative GISTS, the IHC status of SDH is assessed.

- All SDH-deficient patients have a risk of paragangliomas (annual whole-body magnetic resonance imaging [MRI]), whereas those with nonsporadic germline SDHX mutations require genetic counseling.

- For all these reasons, although the GISTs with low risk of relapse and not candidates for any medical therapy also, a mutational analysis is currently felt as a companion to virtually any pathologic diagnosis of GISTs.

- Intussusception is the telescoping of intussusceptum into an intussuscipiens.

- Intussusception mostly occurs in childhood and is rare in adults with the incidence of approximately 2-3 per 1L per year, causing only 1% of all bowel obstruction in adults.



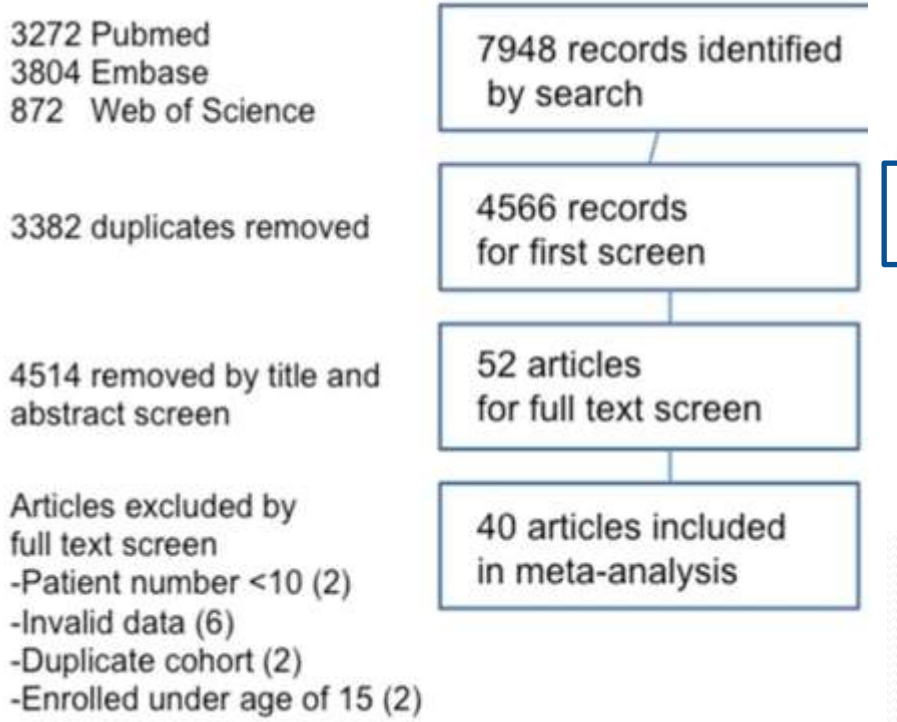
Wang N., Cui X.-Y., Liu Y., et al. Adult intussusception: a retrospective review of 41 cases. World Journal of Gastroenterology: WJG. 2009;15(26):3303–3308. doi: 10.3748/wjg.15.3303.

Kashfi S. M. H., Behboudi Farahbakhsh F., Golmohammadi M., et al. Jejunojejunal intussusception caused by a jejunal villous adenoma polyp in an adult. Annals of Colorectal Research. 2014;2(4) doi: 10.17795/acr-25420.

- In adults with intussusception, 55-60% of reported cases were due to malignant tumor as a pathologic lead point.

- Primary adenocarcinoma is the most common cause.

Honjo H, Mike M, Kusanagi H et al (2015) Adult intussusception: a retrospective review. World J Surg 239(1):134–138



Symptoms and signs	Range of proportions reported by primary studies, %	Pooled propor-tion, %	95% CI (%)
Pain	15–100	82.6	75.6–87.9
Nausea or vomiting	12–100	49.7	42.0–57.5
Absence of gas	6–74	35.9	23.9–49.9
Abdominal mass	4–63	21.9	16.9–27.9
Diarrhea	7–41	20.1	15.8–25.2
Hematochezia	0–54	18.9	14.7–24.1
Fever	4–29	13.9	10.5–18.2

	Enteric (ref = 16)	Ileocolic (ref = 14)	Colonic (ref = 15)
Primary adenocarcinoma	16.6 (8.9–28.8)	61.7 (46.1–75.3)	78.8 (64–88.6)
Metastatic carcinoma	48.7 (31.8–66.0)	13.4 (6.6–25.0)	14.4 (7.3–26.4)
Lymphoma	26.2 (15.2–41.1)	28.1 (16.6–43.5)	16.8 (8.9–29.6)
GIST	21.3 (12.0–34.9)	14.8 (7.3–27.7)	0[a]
Others	23.1 (13.4–36.8)	16.0 (8.3–28.7)	0

Hong KD, Kim J, Ji W, Wexner SD. Adult intussusception: a systematic review and meta-analysis. Tech Coloproctol. 2019 Apr;23(4):315-324. doi: 10.1007/s10151-019-01980-5. Epub 2019 Apr 22. PMID: 31011846

- Between 1983 and 2018, only 18 cases of GIST and Intussusception were reported.

- 72% were women.
- 56% of GISTs were located in the stomach.

- 94% were CD117-positive, and 6% i.e only 1/18 was CD117-negative.

- None was ileo-colic intussusception.

CONCLUSION-

- Bowel obstruction due to intussusception in adults is rare entity.
- Small intestinal GIST is a rare solid tumor with poorer prognosis than Gastric GISTs.
- Intussusception with small intestine GIST is a rare entity and with CD 117 negative GIST is even rare.

THANK YOU