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Case Report

A large inflammatory fibroid polyp of jejunum with unusual presentation: very difficult to diagnosis in paediatric patient

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ABSTRACT

Inflammatory fibroid polyps (IFP), also known as Vanek tumours, are one of the rarest groups of gastrointestinal tract polyps. They represent 0.1-3.0% of all polyps in this organ system. Most common location is the stomach, mainly the antrum (70%), ileum (19%), and colon (6%). Large polyps of the stomach can cause intermittent obstruction, described as "ball valve syndrome". This case report, reported in SMS medical college Jaipur in one year six months old child. We report a case of a 1 year 6 months female child with complaint of abdominal lump noted since 7 days, due to this reduced oral intake. On examination single, around 5×5 cm size non-tender lump is situated in left hypogastric region. On CECT abdomen shows mesenteric origin, may be desmoid tumour, Castleman disease, mesenteric haemangioma or inflammatory myo-fibroblastic tumour. Further MRI abdomen suggestive of inflammatory myo-fibroblastic tumour or NHL. Tumour marker study shows serum ferritin and LDH level raised. On exploration revealed a single, 6×5×4 cm lobulated jejunal mass encircle the jejunum, for this resection and anastomosis done. On histopathological report, suggestive of inflammatory fibroid polyp. Patient discharged successfully on 6th pod without any complication. Exploration done and a mass, which encircle the jejunum found. Resection and anastomosis done and patient discharged on 6th pod without any complications. Such an occurrence was incidental in the reported case, which can confuse our diagnosis, so knowledge about this type of disease is very important, especially in paediatrics population.

Keywords: Inflammatory fibroid polyp, Exploratory laparotomy, Jejunal polyp

INTRODUCTION

Inflammatory fibroid polyps (IFP), also known as Vanek tumours, are one of the rarest, idiopathic pseudo-tumorous lesion of gastrointestinal tract polyps, first described by Vanek in 1949 as an eosinophilic submucosal granuloma. They represent 0.1-3.0% of all polyps in this organ system.[1] They are defined as benign neoplastic lesions projecting from submucosa or mucosa, consisting of spindle stromal cells with infiltration of eosinophils. In that first report of 6 gastric lesions, Vanek called attention to the inflammatory nature of lesions and their submucosal origin. In 1953 Helwig and Ranier confirmed fibroblastic origin of proliferating spindle and stellate cells, and coined term inflammatory fibroid polyp which has remained generally accepted term.[2]

Incident wise slight prominence in male but reported in all age groups, but peak incidence id 50-70 years of age group.[1] Clinical presentation mainly depend in size and location of lesion. Stomach, mainly the antrum (70%) most affected site, ileum (19%), and colon (6%).[2] Natural history not known but rapid growth of lesion within few months also reported. Depending on size and location,

IFP can be asymptomatic or occurring with abdominal pain, gastrointestinal bleeding, weight loss, and vomiting. Large polyps of the stomach can cause intermittent obstruction, described as "ball valve syndrome".[3] There is also possibility of intussusception presenting with acute abdomen when polyp arises below Treitz ligament.[1,4]

CASE REPORT

We report a case of a 1 year 6 months female child with complaint of painless abdominal lump noted since 7 days, due to this reduced oral intake. Passing flatus and motion normally. No any history of vomiting, fever and coughing. No any history of constipation, haematuria and per-rectal bleed.

On examination single, around 5×5 cm size non-tender lump is situated in left hypogastric region. Lump is soft, non-tender, immobile. So initially diagnosed as mesenteric haemangioma/gastrointestinal stromal tumour.

On haematological evaluation-CA-125-25 u/ml, ferritin-265 ng/ml, LDH-857 IU/L, alpha-feto protein and beta-HCG normal range.

All other investigations are within normal range.

Ultrasonography-a highly vascular, hypoechoic, multilobulated mass with central calcification noted which is suggestive of haemangioma or GIST. Mild hepatomegaly also present.

CECT whole abdomen-a well-defined lobulated mass lesion measuring 5×6×5 cm is noted in peritoneal cavity extending from L3-S1 level. Lesion is showing progressive enhancement with intense enhancement on delayed phase and central non-enhancing scar. Lesion is abutting sigmoid colon inferiorly, small bowel loops superiorly and laterally, anterior abdominal wall muscles without any sign of obvious invasion. Mild free fluid in pelvic cavity. Transient ileo-ileal intussusception in right upper abdominal quadrants. These findings are suggestive of-mesenteric desmoid tumour, unicentric Castleman disease, mesenteric haemangioma/inflammatory myo-fibroblastic tumour. CT and MRI reports of patients (Figure 1-5).

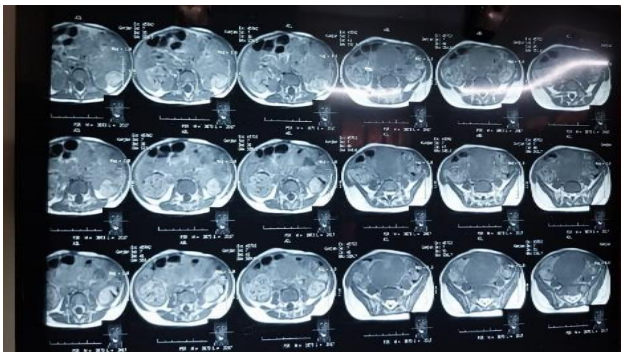

Figure 1: Preoperative Ct/mri film.

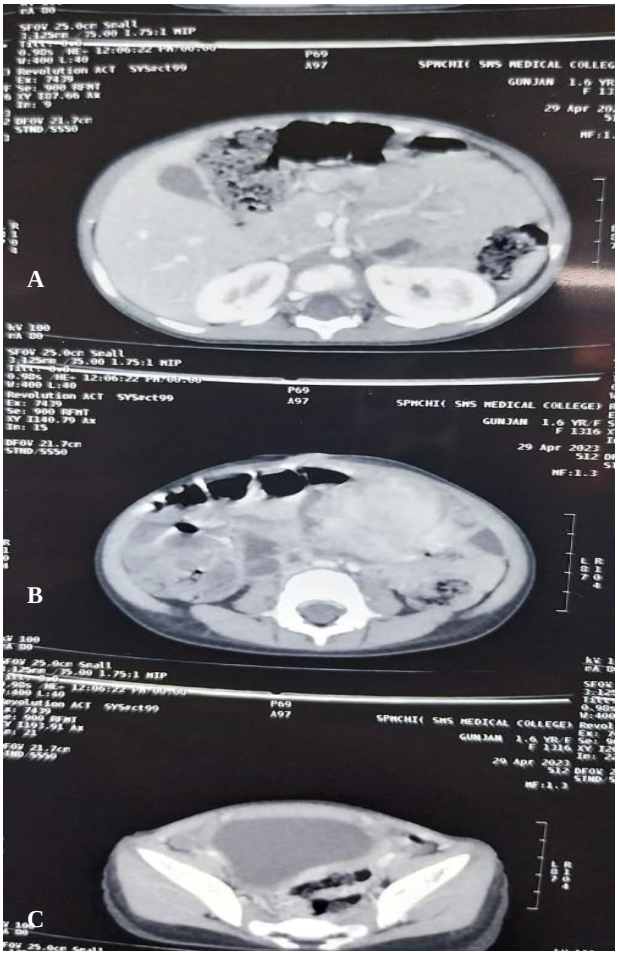

Figure 2 (A-C): Preoperative Ct/mri film.

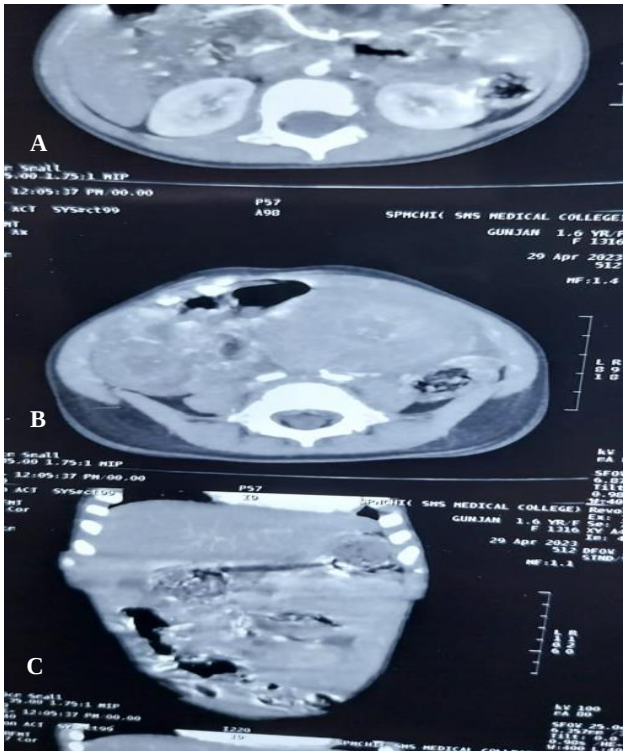

Figure 3 (A-C): Preoperative Ct/mri film.

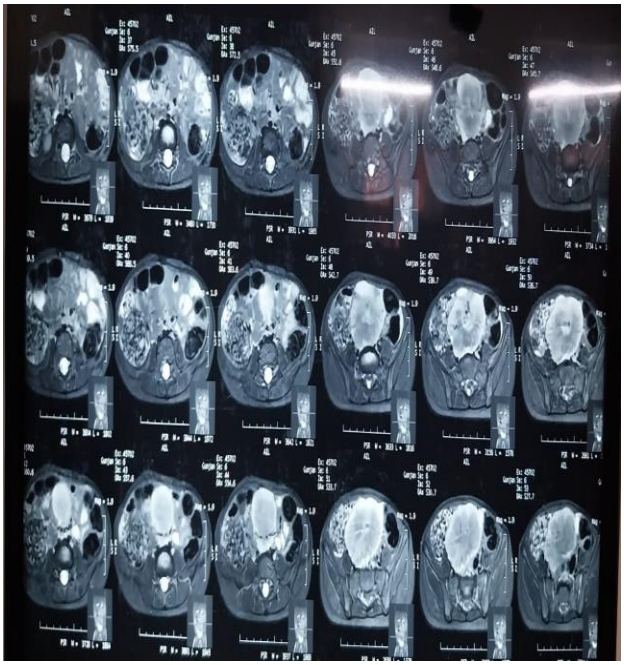

Figure 4: Preoperative Ct/mri film.

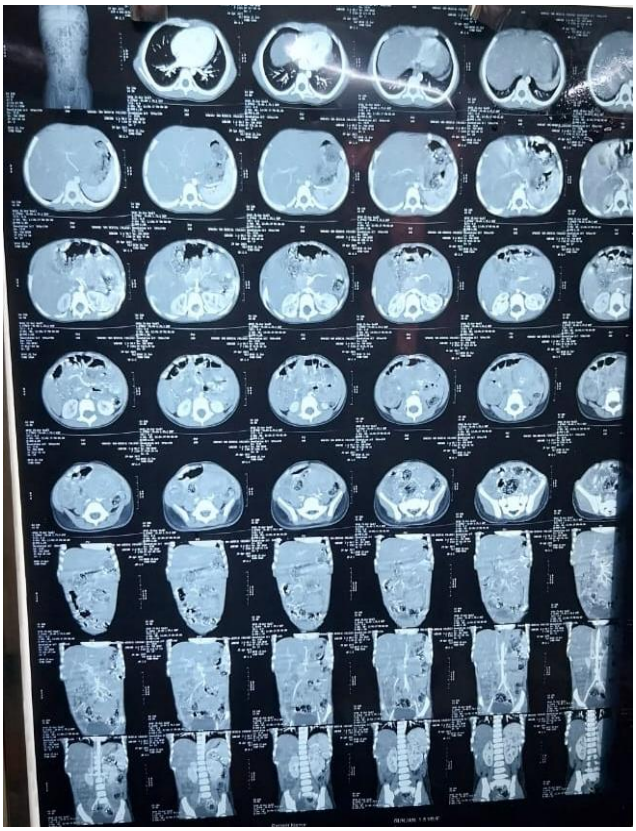

Figure 5: Preoperative Ct/mri film.

MRI abdomen with contrast-A well-defined lobulated lesion appearing T1 isointense, central T2/STIR iso to hyperintense with peripheral hyperintensity, measuring 39×55×49 mm (APXTRXCC) noted in lower abdomen and pelvis, extending from L4 to 52 level. Mass is showing moderate post contrast enhancement. There is no significant diffusion restriction on DWI/ADC images. Anteriorly mass is abutting anterior abdominal muscles, posteriorly mass is abutting right psoas muscle, bilateral iliac vessels and anterior vertebral body at L5-S1 level. Inferiorly mass is causing smooth indentation over urinary bladder without infiltration. Mass is encasing ileal branches of superior mesenteric artery without luminal narrowing. Lesion is causing mass effect in form of left lateral displacement of sigmoid colon with luminal narrowing.

Liver is normal in size, measuring 7.6 mm. Hepatic parenchyma appears normal, no focal or diffuse lesion noted intrahepatic IVC and hepatic veins are normal portal vein is normal, no IHBR dilatation.

These findings are suggestive of- inflammatory myo-fibroblastic tumour or non-Hodgkin lymphoma.

Intraoperative finding-on the basis of these investigatory findings, decided to operate. On exploratory laparotomy around 5×5 cm hard lobulated mass in jejunal mesentery, which is around 20 cm from duodeno-jejunal junction. For which resection of 10 cm jejunum involving mass with primary anastomosis done. Post-operatively no any complication found, the post-operative pictures (Figure 6-9).

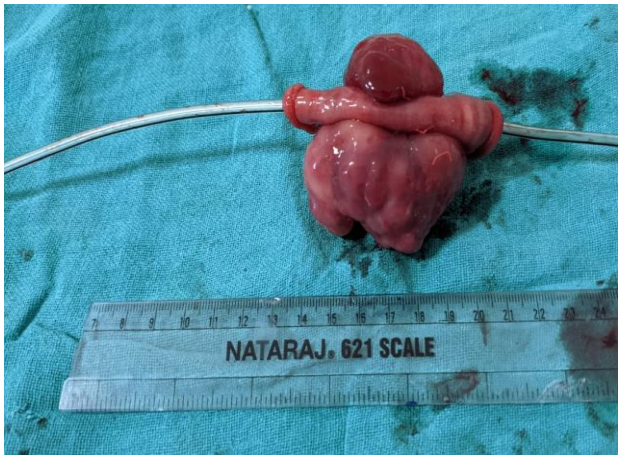

Figure 6: Anterior view of mass after resection.

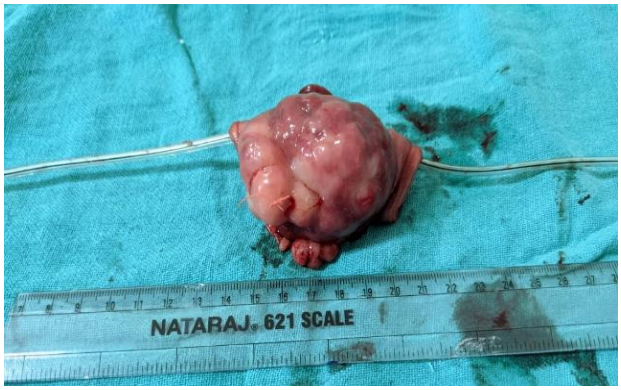

Figure 7: Posterior view of mass after resection.

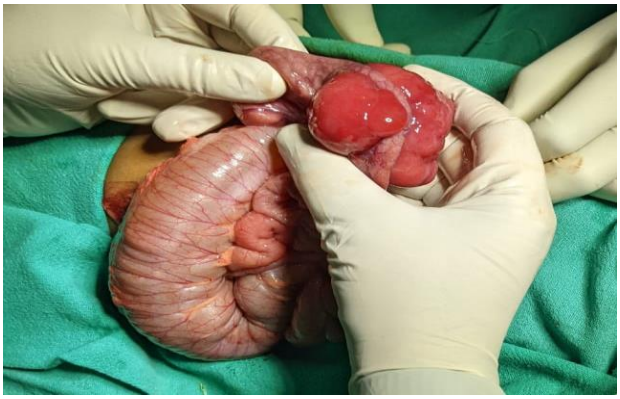

Figure 8: Lateral view of mass before resection.

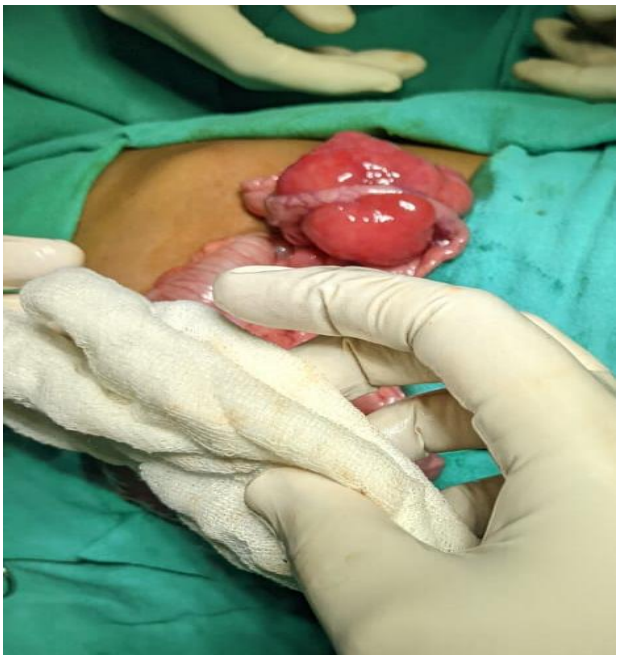

Figure 9: Anterior view of mass before resection.

On histopathology, gross-the specimen consists of segment of intestine measuring 9.5 cm, entrapped by nodular mass measuring 6×5.5×4 cm. Cut surface of mass is solid greyish white. Seven lymph nodes dissected ranging in size from 0.5 cm to 1 cm.

Microscopic-These show extensive proliferation of eosinophils admixed with large cells with irregular nuclear margin and prominent nucleoli. In most of the area's cells are arranged in isolated manner. Mitotic activity is sparse, extensive areas of necrosis seen. Lymph nodes show features of reactive hyperplasia. There is no evidence of malignancy.

Impression was inflammatory fibroid polyp.

On follow up patients found better health and wound healthy. Patient advise to review in oncology department for further management.

DISCUSSION

The incidence rate is slight predominance in men. IFP are reported in every age group, but the peak of incidence occurs at 50-70 years of age.[1] The clinical presentation depends on the size and location of the lesion. It can be asymptomatic or occur with gastrointestinal bleeding, abdominal pain, and vomiting. Polyps located in the upper gastrointestinal (GI) can cause dysphagia and gastroesophageal reflux, while those located below the ligament of Treitz can cause obstruction, diarrhoea, and intussusception, which may lead to acute abdomen.[1,4,6,7] IFP can also imitate acute appendicitis.[8,9] Asymptomatic polyps are also found incidentally during endoscopy performed for unrelated reasons.[8]

IFP can be found during endoscopy but can also be found during laparoscopy or laparotomy.[4,9] Macroscopically they are usually a single lesions 2-5 cm in diameter, semi-pedunculated or sessile, covered with normal mucosa, with possible erosions.[10,11] Histologically IFPs are submucosal (but often expand to the mucosa) with a well-developed network of capillaries, characteristic "onion skin" arrangement of spindle cells around vessels, and infiltration of eosinophils and lymphocytes.[12] Histo-pathologically they should be differentiated with gastrointestinal stromal tumors (GISTs), schwannomas, and inflammatory myo-fibroblastic tumours.[13] Immunohistochemical staining of IFPs is usually positive for CD-34 and negative for CD-117 and S-100 protein.[12,13] This type of polyp is associated with mutation in the PDGFR-A gene, a feature shared with gastrointestinal stromal tumours.[12,13] Due to the submucosal character of the lesion, biopsy specimens taken during endoscopy may not confirm the final diagnosis before resection. In such cases endoscopic ultrasonography or computed tomographic imaging can be used.[10] While the most common removal method is endoscopic resection, there have been reports of infiltration of the muscularis propria layer of stomach, which requires a more radical approach and careful examination of surgical margins.[13]

Most frequently, they are localized in the gastric antrum. Other GI sites which are affected (in decreasing order of frequency) are the small bowel (mainly the ileum), colon, gallbladder, oesophagus, duodenum, appendix and the rectum. Patients with IFP in the stomach present with vomiting and bleeding, while intussusception and obstruction are the symptoms of the lesions which are located in the small bowel.[19] Weight loss, diarrhoea, bleeding and anaemia are seen in the colonic lesions.[20] Adult intussusceptions are relatively rare, constituting only 1% of the patients with bowel obstructions. They are mostly caused by tumours. Eighty percent of the tumours which are associated with small bowel intussusceptions are benign.[21,22] Lipoma is the most common benign tumour which causes intussusception. IFP rarely cause ileal intussusception. Wysocki et al presented a case of biliary obstruction caused by a duodenal IFP.[19] Duodenal

IFP accounts for 1% of the cases and presents with non-specific features. Dabral et al from our department reported a case of inflammatory fibroid polyp in a 7-year-old boy who presented with intestinal obstruction.[23] In the literature, IFP has been reported to be more common in the stomach. However, all the cases reported from our department were from the small intestine (Dabral et al and present study). The pathogenesis of IFPs is still unknown. The role of chemical, physical, or metabolic triggers have been suggested.[24] These polyps are usually solitary, sessile, 2-5 cm in diameter and show mucosal ulceration. Wysocki et al found a firm 4×1.6×2.5 cm mass in the second part of the duodenum, with an overlying small area of mucosal ulceration.[19]

Sometimes differentiation is difficult, especially the differentiation between the IFP and GIST.[25] To conclude, IFP of the small intestine are rare, polypoid lesions which can present as intussusceptions, leading to intestinal obstruction. Clinically, they may be confused with neoplastic polyps, especially in adults and pre operative diagnosis is difficult, requiring histopathological examination for confirmation.

Vanek tumours are rare lesions and occur individually, in contrast to the most common singular or multiple (in 40% of cases) epithelial gastric polyps, known as fundic gland polyps (FGP, Elster glandular cysts).[14,15] The genesis of FGPs is not known, and they are treated as hamartomatous lesions or a particular form of hyperplastic polyp.[15]

For practical reasons FGPs were divided into sporadic type and those associated with familial adenomatous polyposis (FAP). Sporadic FGPs have no clinical significance but their presence is often the reason for referring patients for endoscopic polypectomy. Because there is no real risk of transforming into stomach cancer in such cases, follow-up endoscopy is not recommended.[16] Studies also did not find increased risk of colorectal adenomas in patients with sporadic FGPs.[16,17] In patients with FAP, the incidence rate of FGPs is significantly increased, and they are even treated as a symptom of FAP. Therefore, all patients with incidentally diagnosed FGPs, who are under 40 years old, should undergo colonoscopy.[16,18]

CONCLUSION

IFP is a rare, idiopathic pseudo-tumorous lesion of the gastrointestinal tract. While mostly reported as solitary gastric lesions, multiple cases of small bowel IFPs are also reported. we can assume that such an occurrence was incidental in the reported case, which can confuse our diagnosis, so knowledge about this type of disease is very important, especially in paediatrics population. Due to the rarity of IFP there is no available literature describing co-occurrence of IFP and other types of polyps; therefore, we can assume that such an occurrence was incidental in the reported case.

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