SMALL INTESTINAL LIPOMATOSIS - A RARE CASE REPORT WITH REVIEW OF LITERATURE

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Abstract: Small intestinal lipomatosis refers to the presence of multiple lipomas in the small intestine. Inspite of lipomas being a common soft tissue tumor, they are very uncommon in the small intestine. Their presentation as multiple lesions and with intussusception is exceptionally rare. Here we report a rare case of small intestinal lipomatosis in a 35 year old female who presented with intussusceptions.

Keyword: Intestinal lipomatosis, Intussusception, GIT (gastrointestinal tract)

INTRODUCTION: Lipomas are benign neoplasms of mature adipose tissue and in gastrointestinal tract (GIT) they usually present as solitary lesion. The commonly involved sites in GIT in the order of decreasing frequency are large bowel, small bowel, esophagus and stomach. Intestinal lipomatosis is a condition characterised by the presence of numerous lipomas in the intestine giving rise to multiple sessile and pedunculated polyps. Lipomas and lipomatosis are usually asymptomatic and about one third of the cases present with symptoms like volvulus, intussusception, malabsorption and intestinal bleeding. As lipomatosis of small bowel is a very rare condition and only a few cases have been reported in the medical literature, we report a case of small intestinal lipomatosis in a 35 year old female who presented with intussuception.

CASE REPORT: A 35 year old female came to the surgery OPD with complaints of abdominal pain for the past 2 months. The pain was severe and increasing in intensity for the last 4 days. It was associated with vomiting, constipation and decreased urine output. She gives history of hysterectomy 13 years back. She was not a known diabetic or hypertensive. On examination, she was conscious, well oriented, vitals were stable and other general examinations were within normal limits. Local examination revealed tenderness in the epigastric and umbilical region. There was no palpable mass, guarding or rigidity. Bowel sounds were present. Ultra sonogram of abdomen revealed dilatation of small bowel loops, loculated collections in lower abdomen and minimal thickening of bowel in the lower abdomen suggesting small intestinal obstruction. The patient was taken up for diagnostic laparoscopy under general anaesthesia. Intra operative findings were:

- Omental adhesions to the anterior abdominal wall
- Simple right ovarian cyst
- Small bowel intussusception two feet from duodeno-jejunal flexure

After examination on the table, she was proceeded with midline laprotomy to reduce intussusception. Two feet of dilated small bowel loops was resected and primary anastomosis done? Right ovarian cyst was excised and along with small bowel segment was sent for histopathological examination.

GROSS FINDINGS: Received four segments of small bowel. Gross examination showed intussusception in one of the segments. On cutting open, all the segments revealed multiple sessile polyps studded throughout the intestine in sizes ranging from 0.5 cms to 4x3.5 cms. The polyps appeared yellow. Cut surface of the polyp was yellow, soft, greasy and lobulated (Fig 1).

The ovarian cyst was received in a separate container measuring 6.5x4x2.5 cms and the cyst had no solid areas.

Fig 1

Fig 2
MICROSCOPIC FINDINGS: Microscopic examination revealed wall of small intestine with the mucosa showing multiple polypoid projections. The polyps were covered by intestinal epithelium and the submucosa showed lobules of mature adipocytes with intervening fibro-vascular septae(Fig 2). Some of the polyps showed fibrosis. Mucosal erosions and ulcerations were also observed. Serosal congestion was present. Based on the above gross and microscopic findings a diagnosis of small intestinal lipomatosis was given.

DISCUSSION AND REVIEW: Neoplasms rarely occur in small intestine when compared to the rest of the gastro intestinal tract and it constitute about 1-5% of all gastrointestinal tumors. The estimated prevalence is 1.4/100,000 inhabitants/year. Of these tumors only one third are benign. Lipomas are slow growing, benign, encapsulated neoplasms composed of mature adipose tissue. They generally present as subcutaneous mass and GIT is an uncommon location for lipomas. When it occurs in GIT, large intestine is the most preferred site for lipomas followed by small intestine, esophagus and stomach in the order of decreasing frequency. They are solitary lesions mostly. The presence of multiple lipomas in the intestine is called intestinal lipomatosis. The number of lipomas required for the diagnosis of lipomatosis is not clear. This condition is rare when compared to solitary lesions and less than a dozen cases have only been reported so far. Small intestinal lipomatosis was first described by Hellstrom in 1906. It occurs equally in both sex and has a wide age distribution. The ileum is the most preferred site in small bowel. Aetiology and pathogenesis of this condition is still unclear. A positive familial history suggesting an autosomal dominant inheritance is seen in some patients. Hypercholesterolemia is often coexisting. Lipomatosis associated with hamartomatous syndromes is also known. Soft tissue lipomatosis has been reported to be associated with conditions characterized by mutation of the phosphatase and tensin homolog (PTEN) gene like Proteus syndrome, Cowden syndrome, and Bannayan-Riley-Ruvalca syndrome. A case of coexisting multiple gastro intestinal stromal tumors and intestinal lipomas has also been reported.

Most cases are asymptomatic. Some present with symptoms like abdominal pain, intestinal obstruction or blood in stools. Intestinal obstruction is due to volvulus or intussusception and rarely due to luminal obstruction. Lipomas or lipomatosis presenting as intussusception is infrequent. A review of adult intussusceptions done by Taranee Azar et al showed only 3 cases of lipomas causing intussusception in 44 cases of small intestinal intussusception. Erosions or ulceration of the overlying mucosa, torsion and gangrene are the reasons for blood in stools in these cases. It is a challenging task to diagnose lipomas radiologically. GI transit tests using barium sulfates, enteroclysis with intrawall-filling defects and CT scan are the methods used for radiological diagnosis of lipomas. Owing to the presence of fat, lipomas appear as well demarcated smooth radiolucent mass on GI transit tests. They also show ‘squeeze sign’ because they are fluctuant in size and shape. CT scan reveals characteristic masses with fat density. Grossly, they present as sessile or pedunculated mucosal polyps. The cut surface of these polyps shows fat tissue. In about 40% of cases, there is associated diverticulosis, probably due to weakening of the intestinal wall by the lipomas. Microscopic examination usually reveals lobulated mass of mature adipose tissue in the submucosa. It can be seen extending into the serosa or mesenteric fat. Muscularis propria is usually not affected.

Symptomatic cases are treated with segmental resection with primary end anastomosis. Endoscopic submucosal dissection can be considered for solitary lesions. In case of volvulus, if the intestinal wall is not affected after surgical removal of volvulus,