Case Report

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Primary adenocarcinoma arising from ileo-cecal junction presented with a component of ileo-ileal intussusception

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ABSTRACT

Primary adenocarcinoma arising from small bowel is a rare entity account 1-2% of all GIT malignancies. Adenocarcinoma is the second most malignant lesion of the small bowel after carcinoid tumour followed by lymphoma and GISTs. Duodenum is the common site followed by jejunum and ileum within the small bowel. About 48years old male presented with symptoms and signs of intestinal obstruction i.e. pain and distension of the abdomen with bilious vomiting and absolute constipation. Case was diagnosed as intestinal obstruction provisionally. USG reported as ileocecal intussusceptions with mesenteric lymphadenopathy and was confirmed by CECT. After laparotomy, growth was found at ileocecal junction. Right hemicolectomy along with lymphadenectomy was done and specimen sent for HPE. HPE revealed well differentiated adenocarcinoma, arising from terminal ileum at ileocecal junction with ileo-ileal intussusception. Small bowel adenocarcinoma is rare, further rare in ileocecal junction, radiologically presented as ileo-cecal intussusception. However, lymph nodal enlargement put us in diagnostic dilemma. Author are interested to report this case because of its rarity (incidence of adenocarcinoma terminal ileum is extremely rare and presentation as intussusception is diagnostic challenging.

Keywords: Contrast enhanced computed tomography, Gastro intestinal stromal tumor, Histopathological examination, Ileocecal junction, Small bowel adenocarcinoma

INTRODUCTION

Primary small bowel malignancies are rare. It accounts 1-3% of all GIT malignancies and 0.1-0.3% of all malignant tumours of the body. Duodenum is the most common site for small bowel malignancy followed by jejunum and ileum. Pathologically carcinoid (44%) is the most common small bowel malignancy followed by adenocarcinoma (33%), lymphoma (15%) and GISTs (8%). SBAC is more common after 60 years and above, mean age is 70 years. It is common in both sexes with little male predominance. No known etiological factor is available; however, Crohn's and celiac diseases are

known risk factors. Smoking, alcohol consumption and obesity can also be a risk factor for the development of the SBAC.⁴

SBAC presents with nonspecific symptoms like vague abdominal pain, anorexia, weight loss and anaemia, resulting delay in diagnosis and poor survival rate.⁵ Patient usually presents with an advanced, complicating disease like sub-acute to acute intestinal obstruction, haemorrhage and perforation.⁶ Intestinal obstruction is most common complication, present with abdominal pain, distension, vomiting and absolute constipation. Complicated cases usually present to general surgeon on

emergency, necessitating laparotomy. SBAC is a histological diagnosis after excising the obstructing part. Duodenal growth near to ampulla of vator may cause obstructive jaundice. Pathologically the growth can be annular, nodular, ulcerated, irregular concentric causing partial to total obstruction. The tumour can be multi centric, synchronous primaries are not uncommon.⁷

Plain X-ray erect abdomen will show small bowel obstruction. X-ray barium meal follow through USG abdomen, computed tomography (CT) and magnetic resonance image, video endoscopic capsule and double balloon endoscopy will help in the diagnosis. However, video endoscopic capsule and X-ray barium meal follow through are not useful in obstructive cases. Surgery is the main stay of the treatment followed by chemo radiation. Final diagnosis is established only by histology after surgical excision.8 Right hemicolectomy is indicated the terminal ileum and ileocecal growth with end bloc dissection of lymph nodes followed by chemo radiation. Duodenal malignancies will require pancreaticoduodenectomy. Rest of the small bowel growths require wide local excision with negative margin along with regional lymphadenectomy.^{3,9} Adjuvant chemotherapy showed with improved survival rate. Regarding chemoradiation no substantiating reports are available in the literature.

CASE REPORT

48years old male was admitted with h/o pain abdomen, nausea and voting, anorexia with loss of weight and absolute constipation. Pain was in the right lower quadrant which was progressive and colicky in nature. Patient presented with vomiting 3-4 times a day, bilious in nature. On examination, abdomen was distended, mild tenderness was present in the right ileac fossa, no rigidity and guarding and no mass palpable, no free fluid and no organomegaly. Mild rectal dilatation was noted on per rectal examination. X-ray erect abdomen showed multiple air fluid levels of small bowel pattern (Figure 1).



Figure 1: X-ray erect abdomen-multiple air fluid levels signifies small bowel obstruction.

USG abdomen showed distended small bowel coils. Terminal ileum was thickened with mesenteric lymph adenopathy with free fluid in the peritoneal cavity, suggestive of ileocecal intussusceptions. CECT showed diffuse thickening of ileal loop at IC Junction with extension in to the colon with small fat density lesion in the afferent and efferent loops. Probably in favor of ileoileal/ileo-caecal intussusceptions (Figure 2) enlarged lymph nodes were noted at the root of mesentery. The provisional diagnosis was sub-acute intestinal obstruction secondary ileao-caecal intussusceptions.

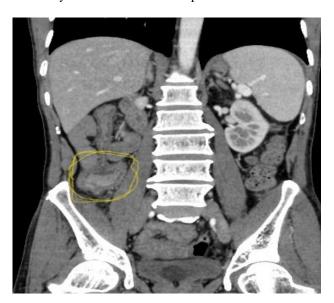


Figure 2: CT-ABD axial section ileocecal intussuception.



Figure 3: CT- ABD axial section lateral view ileocecal intussuception.

Elective laparotomy was done after optimizing the patient. Mass was present at ileocecal junction which was mobile. Enlarged lymph nodes were noted in the

mesentery with minimal peritoneal fluid. Liver and peritoneum were free from metastasis. Right hemicolectomy was done along with lymph node dissection since patient condition was stable. Ileotransverse colostomy was done. Post-operative period was uneventful. Patient discharged on 7th post-operative period. The excised specimen (Figure 3) was sent for histological examination.

Histopathology: gross specimen was showing (Figure 4) greyish white tumour measuring 4x4cm size found at ileocecal junction, spouting in to caecum. ileo-ileal intussusception was seen at ileo-caecal junction. As it was common that any growth in the lumen of a small bowel initiates intussusception.



Figure 4: Intraoperative ileocecal.

Microscopic picture: tumour shows glandular formation. Epithelial cells show loss of nuclear polarity, enlarged hyperchromatic nucleus with nuclear crowding. Tumor invasion extended up to serosa. Metastases are noted in the lymph nodes. Resected margins were free from tumour invasion (Figure 5).



Figure 5: Ileocecal mass.

Impression: well differentiated adenocarcinoma of the ileum involving the serosa and lymph nodes. Patient was referred to oncology department where adjuvant chemotherapy was started. Patient received 8 cycles of capecitabine and tolerated well. Patient was followed for 9 months without any evidence of metastatic disease.



Figure 6: Hyperchromatic nucleus with nuclear crowding and tumor invasion upto serosa.

DISCUSSION

Primary small bowel malignancies are rare. It accounts 1-3% of all GIT neoplasms and 0.1-0.3% of all malignant tumours of the body. Duodenum is the most common site for small bowel malignancies (55%) followed by jejunum (30%) and ileum (15%). Carcinoid (44%) is the most common small bowel malignancy followed by adenocarcinoma (33%), lymphoma (15%) and GSTs (8%).^{2,3} The tumour can present with synchronous primaries and it is multi centric.^{4,7}

SBAC is more common in 6th to 8th decade of life with peak incidence are in 70years of age. It is common in both the sex with little male predominance. No known etiological factors are available. Crohn and celiac diseases are known risk factors however; smoking, alcohol consumption and obesity can also be a risk factor.^{4,5}

Patient presents with vague abdominal pain, anorexia, weight loss and anaemia, which are nonspecific symptoms resulting delay in diagnosis and poor survival rate. ^{5,6} One can suspect SBAC when patient present with weight loss and anorexia without symptoms of bowel obstruction. Patient usually presents with complicating disease like sub-acute or acute intestinal obstruction, haemorrhage and perforation. ^{6,7} Intestinal obstruction is most common complication, present with abdominal distension, vomiting and constipation. Complicated cases usually land up to general surgeon necessitating emergency laparotomy. ⁸ Duodenal growth near to ampulla of Vater may present with obstructive jaundice. ⁹ Plain X-ray erect abdomen will show the pattern of small

bowel obstruction. USG abdomen, computed tomography (CT) and magnetic resonance image helps to find out the site and extant of the growth. Varay barium meal follow-through and video endoscopic capsule is not useful when patient presents with intestinal obstruction. It is difficult to diagnose the mass lesion by USG and CT, if intussusception component is present. Author failed to diagnose the mass lesion on CT and USG, rather diagnosed it as intussusception. Final diagnosis can only be established by histological sections after surgical excision of the mass lesion. Variation of the mass lesion.

Surgery is the main stay of the treatment followed by chemo radiation. Right hemicolectomy with end bloc dissection of lymph nodes is indicated in growth arising from ileocecal junction and terminal ileum. Author did the same procedure in this case. Duodenal growth will require pancreatico-duodenectomy followed by triple anastomosis. Growth from rest of the small bowel requires wide local excision with negative margin along with regional lymphadenectomy.^{3,11}

Adjuvant chemotherapy will have improved survival rate however, no certain chemotherapeutic agents are evaluated properly. Chemotherapeutics which are used in other GIT cancers like 5FU, combined with leucovorin and oxaliplatin and other tyrosine kinase inhibitors are routinely given as adjuvant or neoadjuvant agents in small bowel carcinoma. Oxaliplatin and capecitabine are given as neoadjuvant chemotherapy in advanced cancer with improved survival rate. 12 SBAC is a histological diagnosis after excision of the obstructing part and sent for HPE. In this case, also patient presented with intestinal obstruction. Author did right hemicolectomy and it was histological reported as well differentiated adenocarcinoma of IC junction with a component of intussusceptions. Prognosis was poor because of late presentation due to lack of specific symptomatology. The reported 5year survival rate was 25-40%. 13,14

CONCLUSION

Adenocarcinoma of small bowel is a rare disease. Duodenum is the most common site followed by jejunum and ileum. It is difficult to diagnose preoperatively as it presents with nonspecific clinical features. Patient usually presents to a general surgeon on emergency basis due to small bowel obstruction. Growth is only found after emergency laparotomy. Plain X-ray erect film will confirm the small bowel obstruction. CECT fails to diagnose the mass lesion when intussusception component is present. Curative resection is planned if patient condition is stable otherwise bypass surgery in the form of ileo-transverse colostomy is carried out postponing the elective procedure.

Right hemi- colectomy is the choice if growth arising from ileocecal junction followed by ileo-transverse colostomy. Pancreatico-duodenectomy is the choice for duodenal growth followed triple anastomosis. Wide local excision with lymphadenectomy is planned in ileal growth. Adjuvant chemotherapy is not properly evaluated in the literature. GIT malignant chemotherapeutics are routinely given as an adjuvant therapy with reported improved survival rate. The same drugs are also given as neoadjuvant therapy in advanced cases.

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