



Case Report:

Primary Signet-Ring Carcinoma (Linitus Plastica) of the Colorectum presenting as Subacute Intestinal Obstruction

Sunil V Jagtap, Dept. of Pathology,
Dhiraj B Nikumbh, Dept. of Pathology,
Ashok Y Kshirsagar, Dept. of Surgery,
Swati S Jagtap, Dept. of Physiology
Shamima, Dept. of Pathology,
KIMS University and KHMRC, Karad, Maharashtra, India.

Address for Correspondence:

Dr. Sunil Vitthalrao Jagtap,

Associate Professor,
Department of Pathology,
Krishna Institute of Medical Sciences,
University of Karad - 415110,
Maharashtra, India.

E-mail: drsvjagtap@gmail.com

Citation: Jagtap SV, Nikumbh DB, Khirsagar AY, Jagtap SS, Shamima. Primary Signet-Ring Carcinoma (Linitus Plastica) of the Colorectum presenting as Subacute Intestinal Obstruction. *Online J Health Allied Scs.* 2012;11(1):14

URL: <http://www.ojhas.org/issue41/2012-1-14.htm>

Open Access Archives: <http://cogprints.org/view/subjects/OJHAS.html> and <http://openmed.nic.in/view/subjects/ojhas.html>

Submitted: Feb 28, 2012; Accepted: Mar 25, 2012; Published: Apr 15, 2012

Abstract: Primary Signet-ring cell carcinoma (Linitus Plastica) of the colon and rectum is a rare form of adenocarcinoma of the large intestine and has been reported to have an extremely poor prognosis. We report a case of Primary Signet-ring cell carcinoma of the colorectum in a thirty one year old man presented in Surgical OPD of our hospital with chief complaints of persistent pain in abdomen and vomiting since two days. Since the prognosis of primary signet ring cell carcinoma (SRCC) is extremely poor (in view of more malignant behavior than ordinary colorectal carcinoma), early diagnosis and aggressive treatment strategy are necessary.

Key Words: Signet ring cell carcinoma; Colorectal neoplasm; Linitus plastica

Introduction:

Signet ring cell carcinoma (SRCC) is uncommon in the colon and rectum, with a reported incidence ranging from 0.1% to 0.8%. [1,2] It is a rare but distinctive malignancy of the large bowel with more malignant behavior. More than 96% of the SRCC arises in the stomach, with the remainder arising from other sites involving colon, rectum, gall bladder, pancreas, urinary bladder and breast. [3] In general, SRCC shows the characteristic appearance of "Linitus Plastica" and behaves more aggressively than other histological types of carcinoma. [1,2] Primary SRCC were seen in younger patients (less than forty years of age) as per Tung SY et al. [3] As symptoms usually appear late, SRCC are commonly detected at advanced stages. Therefore, cases detected and treated at early stage are rare. In this case report, we report a thirty one year old man with SRCC in colorectum which was detected at early stage and treated aggressively.

Case Report:

A thirty one year old man presented in surgical outpatient department of our hospital with chief complaints of persistent pain in abdomen with vomiting since two days. He had past history of constipation and altered bowel habits, off an don since 3 months. Patient was a chronic tobacco chewer and potter by occupation with no history of diabetes mellitus, tuberculosis, inflammatory bowel disease, hypertension or ischemic heart disease. Family history was not relevant. Per

abdominal examination revealed distension with vague tenderness in epigastric and hypogastric region with free fluid and sluggish peristalsis. Other systemic examination was within normal limit. Per rectal examination did not reveal any abnormality. X ray chest was within normal limit. Plain abdomen radiography suggestive of large bowel obstruction and dilated bowel loops proximal to rectum. Gastroscopy revealed no abnormality. Laboratory investigations revealed mild neutrophilic leucocytosis with mildly increased creatinine. Rest of the biochemical parameters were within normal limits. With above findings, clinical diagnosis of subacute intestinal obstruction was made and laprotomy was performed.

Laparotomy revealed distended sigmoid colon. Serosal surface showed multiple tiny nodules of mucin. A stricture was palpable in distal sigmoid colon. Radical surgical resection of rectosigmoid colon with wide margin of 5 cm was done with end to end anastomosis. Peritoneal seeding was noted. Stomach, liver and spleen were normal. The resected rectosigmoid specimen was sent for histopathological study.

Gross Features: Received a segment of recto-sigmoid measuring 15 cm in length, showing a constricted area measuring 2 cm in length and 6 cm away from distal surgical margin. Serosa showed congestion and many mucinous nodules (Figure 1). On cutting open, rectosigmoid showed a large single encircling gray-white ulceroinfiltrative growth measuring 3x2 cm (Figure 2). Wall was thickened by tumor and growth was seen reaching upto serosa. Cut section of the tumor mass was gray-white with mucinous areas. Also received peritoneal whitish nodules measuring 0.3 to 0.6 cm. On cutting open revealed gray-white, mucinous areas.

Light Microscopy: Multiple sections through rectosigmoid showed large intestinal mucosa with extensive areas of ulceration and tumor arising from it and infiltrating deeper tissue (Figure 3). The tumor was composed of neoplastic cells arranged in thick cords, nests, lobules and ill formed glands. Individual tumor cells are large, round to oval having mild pleomorphic hyperchromatic nuclei with abundant vacuolated or eosinophilic cytoplasm pushing the nuclei to periphery (signet ring cells) (Figure 4, 5). Many tumor cells floating in the pools

of extracellular mucin were noted. Solid areas of tumor at places were also seen. Tumor was seen infiltrating full thickness of mucosa and over serosal surfaces. Occasional tumor emboli, perineural infiltration were seen. Sections from peritoneal nodules showed fibroadipose tissue with SRCC. Both the surgical margins were free from tumor. The final histopathological diagnosis of Primary Signet ring cell carcinoma was given. The patient was on regular follow up.



Figure 1: Gross specimen of resected rectosigmoid with constricted area and serosal tiny mucinous nodules.



Figure 2: Cut surface of the rectosigmoid showing encircling gray white ulceroinfiltrative mass.

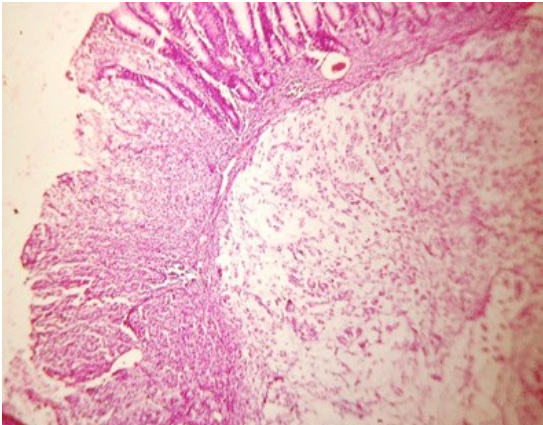
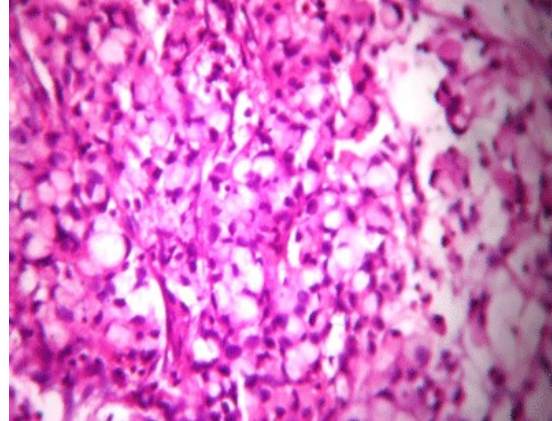


Fig 3: Photomicrograph showing large intestinal mucosa with ulceration and tumor arising from it and infiltrating the deeper structures. (H&E x100).

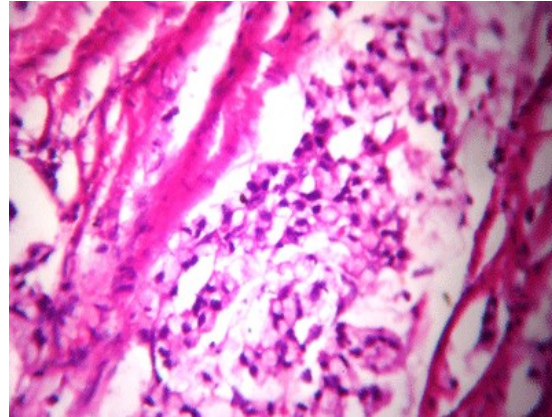
Discussion:

Primary Signet ring cell carcinoma (SRCC) of the colon and rectum is a rare variant of colorectal adenocarcinoma. SRCC has an aggressive clinical course and a poor prognosis.[4] There is high incidence of peritoneal metastases and relatively

low incidence of hepatic metastases, a characteristic feature distinguishing colorectal signet ring cell carcinoma from non-signet colorectal carcinoma.[4]



Figures 4 (above) and 5: Photomicrographs showing tumor cells arranged in thick cords, nests and ill defined glands with characteristic signet ring cell tumor cells. (H&Ex400).



SRCC has been described in virtually all mucinous organs of the body, most frequently in stomach, but also in urinary bladder, prostate, breast, lung, and pancreas etc.[5] Within the gastrointestinal tract, up to 96% of SRCC are found in stomach. [6] In the colon, although small foci of signet ring cells can be seen in conventional adenocarcinoma, a primary SRCC with greater than 50% of tumor cells being signet rings, is extremely uncommon.[5-7]

Primary signet ring cell carcinoma of the colon and rectum as first described by Laufmann and Sufir in 1951 is rare entity [8]. The reported incidence is 0.1 to 0.8% only.[1,2] The histological appearance of the tumor is characterized by cells with abundant intracytoplasmic mucin and peripherally placed nuclei. SRCC of the colon and rectum are usually diagnosed at an advanced stage, because symptoms usually develop late. Thereafter cancers limited to the mucosal and submucosal layers are rarely detected, as seen in our case.

Primary SRCC of colorectum is diagnosed when the following criteria are satisfied:

1. The tumor is primary.
2. Histological material is adequate.
3. Signet ring cells present more than 50% of the cancer.[7]

All the three criteria were satisfied in our case. In our case, clinical history, gastro copy and laparotomy ruled out a primary growth in the stomach. Macroscopically SRCC shows annularly thickened and rigid bowel wall in long segment and stenotic bowel lumen, providing a linitus plastica appearance. Microscopically poorly differentiated or signet ring cells are

seen. Peritoneal dissemination is very common.[9] Our patient had all these features.

Immunostaining profiles for CK 7 and CK 20 have been used to characterize and differentiate SRCC of breast, stomach and colon.[10,11] CK 20 is a low molecular weight cytokeratin that is normally expressed in the gastrointestinal epithelium, urothelium and in Merkel cells.[12] Ck 7 is expressed by tumors of the lungs, ovary, endometrium and breast but not of the lower gastrointestinal tract.[4]

Conclusion:

Primary signet ring cell carcinoma of colorectum is a rare entity with extremely poor prognosis in view of more malignant behaviour than other colorectal carcinoma. Early diagnosis and aggressive treatment strategy are necessary for better management of patients.

References:

1. Giaccherio A, Aste H, Barachhini P, Conio M, Fulcheri E, Laperto G, Tanzi R. Primary signet-ring cell carcinoma of large bowel. Report of nine cases. *Cancer*. 1985; 56(11):2723-2726.
2. Almagro UA. Primary signet-ring cell carcinoma of the colon. *Cancer*. 1983;52:1453-1457.
3. Tung SY, Wu CS, Chen PC. Primary signet-ring cell carcinoma of colorectum :an age and sex matched controlled study. *Am J Gastroenterol*. 1996;91:2195-2199.
4. Sim HL, Tan KY, Poon PL, Cheng A. Primary rectal signet-ring cell carcinoma with peritoneal dissemination and gastric secondaries. *World J Gastroenterol*. 2008;14(3):2118-2120.
5. Maeham Y, Sakaguchi Y, Moriguchi S et al. Signet ring carcinoma of the stomach. *Cancer*. 1992;69:1645-1650.
6. Tendon M, Sostek M, Klein MA. Focus of signet ring-cell carcinoma in an adenoma of the sigmoid colon. *Arch Pathol Lab Med*. 1999;123:957-959.
7. Messerini L, Palomba A, Zampi G. Primary signet-ring cell carcinoma of the colon and rectum. *Dis Colon Rectum*. 1995;38:1189-1192.
8. Laufman H, Safir O. Primary linitus plastica type of carcinoma of the colon. *AMA Arch Surg*. 1951;62:79-91.
9. Nakahara H, Ishikawa T, Itubashi M, Hirota T. Diffusely infiltrating colorectal carcinoma of linitus plastic and lymphangiosis type. *Cancer*. 1992;69:901-906.
10. Goldstein NS, Long A, Kuan SF, Hart J. Colon signet ring-cell adenocarcinoma: immunohistochemical characterization and comparison with gastric and typical colon adenocarcinomas. *Appl Immunohistochem Mol Morphol*. 2008;8:183-188.
11. Tot T. The role of cytokeratins 20 and 7 and estrogen receptor analysis in separation of metastatic lobular carcinoma of the breast and metastatic signet-ring cell carcinoma of the gastrointestinal tract. *APMIS*. 2000;108:467-472.
12. Varadhachary GR, Abbnizzese JL, Lenzi R. Diagnostic strategies for unknown primary cancer. *Cancer*. 2004;100:1776-1785.