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Signet-Ring Carcinoma Colon in Young Age – A Case Report

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ABSTRACT

Colorectal adenocarcinoma (CRC) is the most common cause of death worldwide. one of the histologic subtypes of CRC is the signet ring cell carcinoma (SRCC) which has distinct molecular and tumor biology. primary SRCC diagnosed at an early stage is very rare, therefore overall prognosis is poor.16 year old female presented to the casualty ward with complaints of abdominal pain, vomiting, abdominal distention of 3 days duration with features suggestive of intestinal obstruction. Emergency laparotomy was done. patient had gangrene ascending colon and growth in descending colon. Right hemicolectomy with ileotransverse anastomosis and resection of growth in descending colon with proximal end colostomy and closure of the distal end of colon.HPE report revealed signet ring carcinoma and patient was started on 5 FU based therapy to prevent recurrence of disease.

KEYWORDS

Colorectal adenocarcinoma, signet- ring cell carcinoma, Right hemicolectomy, Colostomy

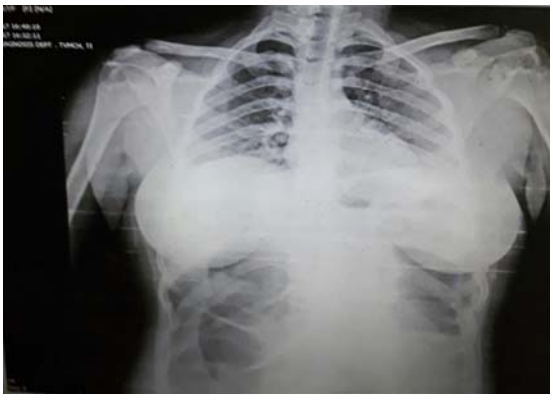
INTRODUCTION

Colorectal adenocarcinoma (CRC) is one of the most common cancers worldwide and the third leading cause of death in united states.CRC can be histologically sub typed into adenocarcinoma (AC), which accounts for large majority of cases; mucinous adenocarcinoma (MAC); signet ring cell carcinoma (SRCC) and even less frequent subtypes. primary SRCC of the colon is a rare entity, accounting for less than 1% of all CRC.SRCC is associated with a high grade tumor, young age, female patients and distinct molecular patterns, such as microsatellite instability(MSI) and activating mutations of the BRAF gene compared to mucinous adenocarcinoma. However since clinical symptoms tend to occur late in the course of SRCC, most cases are usually detected at an advanced stage with a poor overall survival rate. primary SRCC at an early stage is rare, and only 27 case have been reported.

CASE REPORT

A 16 yr old female presented to the casualty with complaints of abdominal pain, vomiting and abdominal distention of 3 days duration without any significant past medical and family history. on examination patient was conscious, oriented, afebrile, mild dehydration present and tachycardia present. Abdomen was distended, diffuse tenderness present with absent bowel sounds. Per rectal examination was empty. X-ray showed dilated bowel loops (Figure no:1). USG abdomen showed dilated bowel loops (Figure no:2). CT abdomen showed thickening of descending colon (Figure no:3). Emergency laparotomy was done. Intraop findings were gangrene of ascending colon upto hepatic flexure and growth in the descending colon with mucinous substance outside the bowel (Figure no:4). Right hemicolectomy with ileotransverse anastomosis and resection of the growth in the descending colon was done with proximal end colostomy and distal end closed. post op period was uneventful. HPE report revealed SRCC, transmural infiltration with resected margins free of tumor.4/4 lymph nodes metastatic carcinomatous deposits (Figure no:5). Due to patients young age an adjuvant 5 FU based therapy was commenced to prevent recurrence of the

disease. patient has completed 5 of the planned 8 cycles of chemotherapy without any haematological or non haematological toxicities.



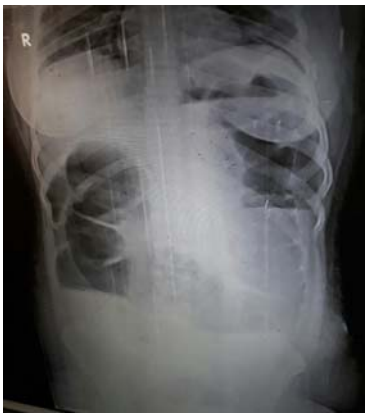
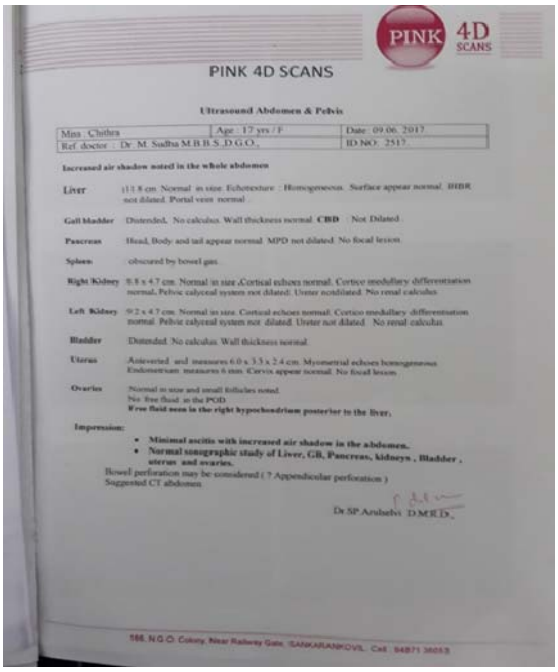


Figure no:1



PINK 4D SCANS

PINK 4D SCANS

Ultrasound Abdomen & Pelvis

Miss. Chithra	Age : 17 yrs / F	Date: 09.06. 2017
Ref. doctor : Dr. M. Sudha M.B.B.S.,D.G.O.,		ID NO: 2517.

Increased air shadow noted in the whole abdomen

Liver 11.8 cm. Normal in size. Echotexture : Homogeneous. Surface appear normal. IHBR not dilated. Portal vein normal.

Gall bladder Distended. No calculus. Wall thickness normal. **CBD** : Not Dilated.

Pancreas Head, Body and tail appear normal. MPD not dilated. No focal lesion.

Spleen obscured by bowel gas.

Right Kidney 8.8 x 4.7 cm. Normal in size ,Cortical echoes normal. Cortico medullary differentiation normal. Pelvic calyceal system not dilated. Ureter nondilated. No renal calculus.

Left Kidney 9.2 x 4.7 cm. Normal in size. Cortical echoes normal. Cortico medullary differentiation normal. Pelvic calyceal system not dilated. Ureter not dilated. No renal calculus.

Bladder Distended. No calculus. Wall thickness normal.

Uterus Anteverted and measures 6.0 x 3.3 x 2.4 cm. Myometrial echoes homogeneous. Endometrium measures 6 mm. Cervix appear normal. No focal lesion.

Ovaries Normal in size and small follicles noted.
No free fluid in the POD.
Free fluid seen in the right hypochondrium posterior to the liver.

Impression:

- **Minimal ascitis with increased air shadow in the abdomen.**
- **Normal sonographic study of Liver, GB, Pancreas, kidneys , Bladder , uterus and ovaries.**

Bowel perforation may be considered (? Appendicular perforation)
Suggested CT abdomen.

Dr.SP.Arubalvi D.M.R.D.,

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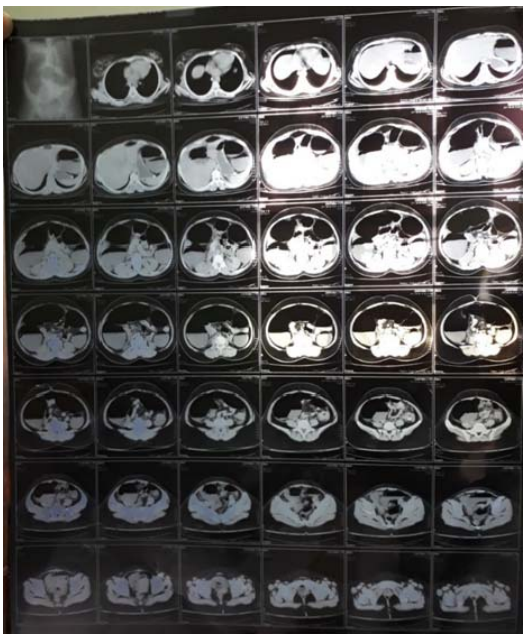
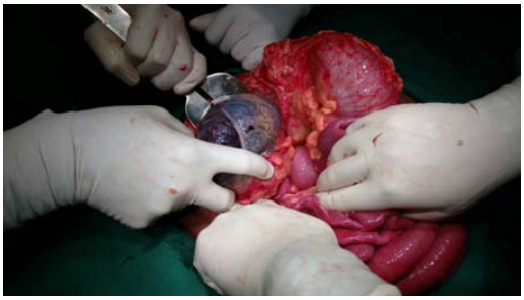
Figure no: 2





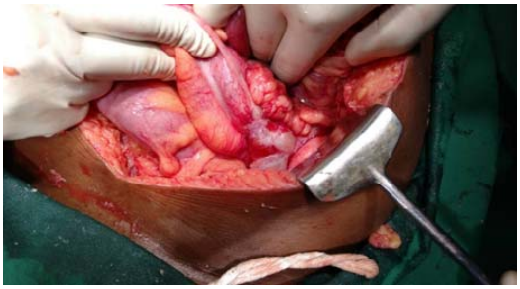
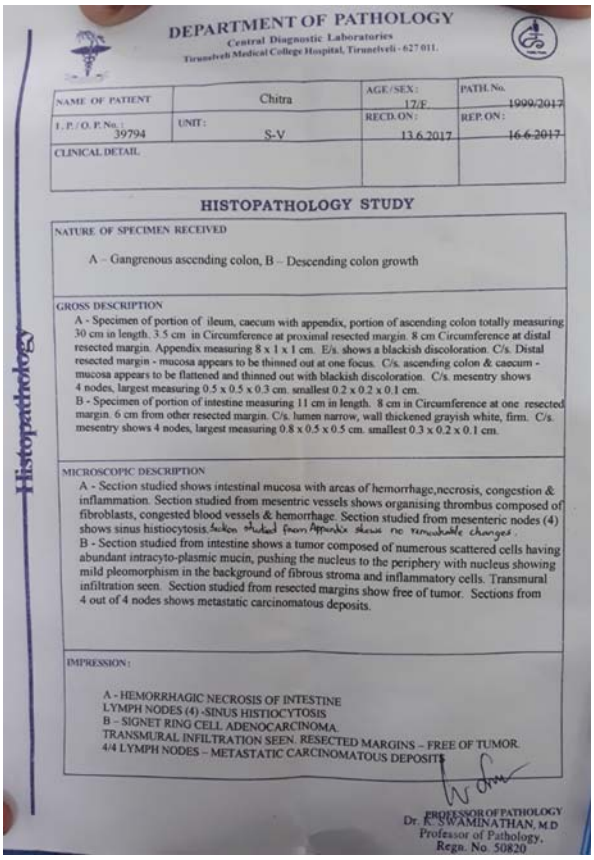


Figure no: 4



DEPARTMENT OF PATHOLOGY
Central Diagnostic Laboratories
Tirunelveli Medical College Hospital, Tirunelveli - 627 011.

NAME OF PATIENT	Chitra	AGE/SEX: 17/F	PATH.No. 1999/2017
I.P./O.P.No. 39794	UNIT: S-V	RECD.ON: 13.6.2017	REP.ON: 16.6.2017
CLINICAL DETAIL			

HISTOPATHOLOGY STUDY

NATURE OF SPECIMEN RECEIVED

A – Gangrenous ascending colon, B – Descending colon growth

GROSS DESCRIPTION

A - Specimen of portion of ileum, caecum with appendix, portion of ascending colon totally measuring 30 cm in length. 3.5 cm in Circumference at proximal resected margin. 8 cm Circumference at distal resected margin. Appendix measuring 8 x 1 x 1 cm. E/s. shows a blackish discoloration. C/s. Distal resected margin - mucosa appears to be thinned out at one focus. C/s. ascending colon & caecum - mucosa appears to be flattened and thinned out with blackish discoloration. C/s. mesentry shows 4 nodes, largest measuring 0.5 x 0.5 x 0.3 cm. smallest 0.2 x 0.2 x 0.1 cm.
B - Specimen of portion of intestine measuring 11 cm in length. 8 cm in Circumference at one resected margin. 6 cm from other resected margin. C/s. lumen narrow, wall thickened grayish white, firm. C/s. mesentry shows 4 nodes, largest measuring 0.8 x 0.5 x 0.5 cm. smallest 0.3 x 0.2 x 0.1 cm.

MICROSCOPIC DESCRIPTION

A - Section studied shows intestinal mucosa with areas of hemorrhage,necrosis, congestion & inflammation. Section studied from mesentric vessels shows organising thrombus composed of fibroblasts, congested blood vessels & hemorrhage. Section studied from mesenteric nodes (4) shows sinus histiocytosis. Section studied from Appendix shows no remarkable changes.
B - Section studied from intestine shows a tumor composed of numerous scattered cells having abundant intracyto-plasmic mucin, pushing the nucleus to the periphery with nucleus showing mild pleomorphism in the background of fibrous stroma and inflammatory cells. Transmural infiltration seen. Section studied from resected margins show free of tumor. Sections from 4 out of 4 nodes shows metastatic carcinomatous deposits.

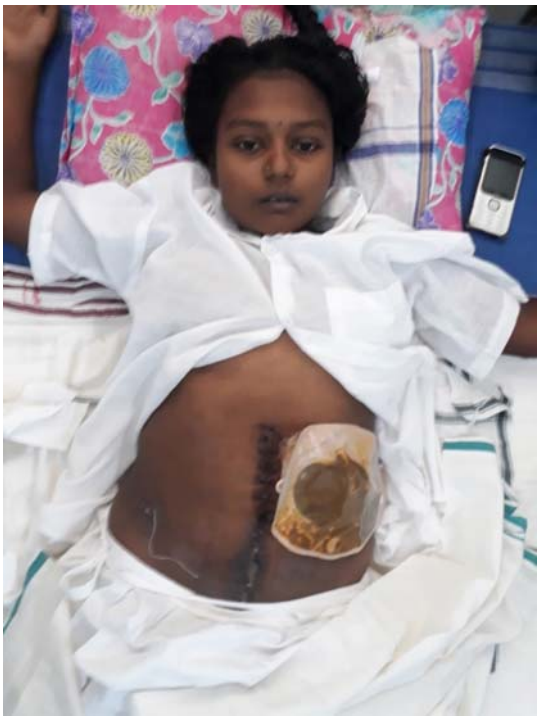
IMPRESSION:

A - HEMORRHAGIC NECROSIS OF INTESTINE
LYMPH NODES (4) -SINUS HISTIOCYTOSIS
B – SIGNET RING CELL ADENOCARCINOMA.
TRANSMURAL INFILTRATION SEEN. RESECTED MARGINS – FREE OF TUMOR.
4/4 LYMPH NODES – METASTATIC CARCINOMATOUS DEPOSITS

PROFESSOR OF PATHOLOGY
Dr. K. SWAMINATHAN, M.D
Professor of Pathology,
Regn. No. 50820

Histopathology

Figure no: 5



DISCUSSION

SRCC is a distinct entity based both on clinical presentation and pathology. SRCC occurs in younger population with female predominance, usually less than 40 yrs of age. More than 96% of cases arise in the stomach with the rest occurring in the colon, rectum, gallbladder, pancreas, urinary bladder and breast. SRCC usually presents at an advanced stage with node positive disease and metastatic spread which results in poor prognosis. A unique feature of SRCC is its fibrotic appearance and preference for peritoneal metastasis over the liver.

Pathologicallly SRCC demonstrates unique features that distinguish the cells from routine AC. The signet ring malignant cells are seen floating in abundant extracellular mucin pools either as clusterd or isolated cells. These cells have a slightly lower prevalence of KRAS mutation but a higher BRAF mutation rate as compared to classical AC, and the prognostic implication is unknown. Loss of E – cadherin expression has been reported and contributes to the high grade and invasive nature of SRCC as the cells acquire stem cell like characteristics.MSI tends to be more prevalent in SRCC than AC.

The overall prognosis of SRCC in general is similar to other cancers in that staging and grade are important determinants. The available literature suggests that the tumor staging is the best predictive factor for the prognosis of SRCC of the colon where higher tumor staging means poorer prognosis. The 5 yr survival rate in SRCC of the colon ranges from 0-12% and disease recurrence is more frequent in SRCC of the colon compared to AC. Absence of lymphovascular invasion, lack of lymph node metastasis and lower TNM stage has a favourable effect on the survival of SRCC patients. patients with SRCC can definitely benefit from closer follow up or even intensified adjuvant therapy due to their high rates of local and distant recurrence. In a study by Hugen et al adjuvant chemotherapy for SRCC stage III of the colon was associated with improved survival.

CONCLUSION

Primary SRCC is considered a distinct entity based on the clinical and pathological features. In this case report, we have shown a rare case of primary SRCC colon. Given the poorer prognosis of these patients, 5 FU based therapy was considered in an attempt to prevent recurrence.

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