Case Report

Signet Ring Cell Carcinoma Colon in A Twelve Year Old Child: A Case Report with Review of Literature

Amita Jain Gupta[1], Jenna B Bhattacharya[1]*, S Vijaya[1], Richa Gupta[1], Vivek Manchanda[2]

[1]Department of Pathology, Maulana Azad Medical College, New Delhi, India
[2]Department of Paediatrics, Maulana Azad Medical College, New Delhi, India

Keywords: *Signet Ring Cell Carcinoma, Colon, Paediatric Colon Carcinoma*

ABSTRACT

A twelve year old male patient presented with pain abdomen and vomiting on and off since 4 months. Patient had abdominal distension and a lump was palpable in abdomen on examination. The gross examination of the resected sigmoid colon revealed a stricture which on microscopy revealed the features of signet ring carcinoma colon. This case is being presented here due to rarity of this type of carcinoma in children.

***Corresponding author:**
Dr Jenna. B. Bhattacharya, C-2102, Amrapali Eden Park , Appts, Block-F, Sector-50, Noida. U.P. 201301 India
Phone: +91 9899840023
E-mail: jennamamc@gmail.com

Introduction

Signet ring cell carcinoma colon is aggressive tumour [1] with an overall prognosis worse than conventional adenocarcinoma due to high rate of metastases and local spread. This tumour constitutes 0.01 to 0.9% of patients of colon and rectal carcinoma. [1] The occurrence of these tumours is even rare with very few cases reported till date. We present a case of signet ring cell carcinoma in a twelve year old child.

Case Report

A 12 year old male patient presented with pain abdomen and vomiting on and off since four months. Patient had abdominal distension and constipation since three days. There was history of weight loss and weakness. Patient had pallor and was cachexic. An abdominal mass was palpable on examination. There was no palpable lymphadenopathy. Clinically tuberculosis was suggested.

X ray abdomen showed dilated bowel loops. The patient had to be operated in emergency as he had sudden severe pain abdomen along with obstipation and features of acute obstruction. Operative findings revealed sigmoid stricture with peritoneal white deposit in pelvis and extensive dilated loops of colon. A provisional diagnosis of tubercular stricture with caseous material and tubercles in peritoneum was thought of in view of the age and presentation of the patient.

Gross Examination: Colonic segment measured 40 cm in length comprising of sigmoid colon and proximal large intestine. No growth was seen grossly. On cutting open a stricture was indentified 4 cm from the smaller resected end comprising of sigmoid colon with dilation of the proximal colon. The mucosa over the stricture was completely denuded and ulcerated and the bowel wall was thickened over the entire length of the stricture to 0.8 cm. The stricture measured 6 cm in length (Fig.1A) and both the resected ends were uninvolved. The colon proximal to the stricture was completely dilated and the bowel wall had become thin however no area of perforation was seen. The mucosa over the dilated segment was maintained.

Externally intestine was congested. Mucosa was predominantly flattened out and partially ulcerated. Lymph nodes were isolated in mesocolon were firm and homogenous.

Microscopic Examination: Sections from the stricture revealed complete mucosal ulceration and presence of sheets of atypical cells spread transmurally in a dyscohesive pattern. The cells were signet ring type with peripherally placed hyperchromatic nucleus. The presence of intracellular mucin was confirmed by mucicarmine stain. Also present were focal areas (comprising approximately 10%) with extracellular mucin deposits and malignant glands.

The tumor cells reached the serosa. A diagnosis of signet ring cell carcinoma was given. Fourteen lymph nodes were isolated out of which one showed tumor deposit. Resected ends and intervening mucosa were uninvolved. Extensive sampling of the peritoneum revealed a peritoneal deposit. The colon cancer was classified as Duke stage C2 in this patient. There was no lymphovascular invasion.

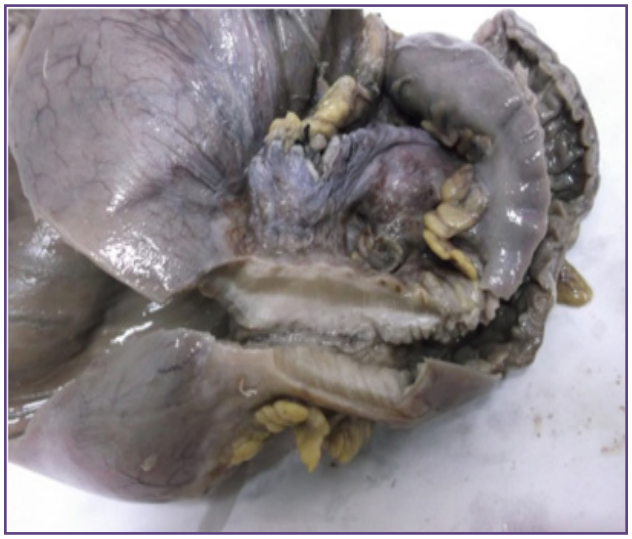

Fig. 1A: Gross appearance of intestine showing presence of stricture.

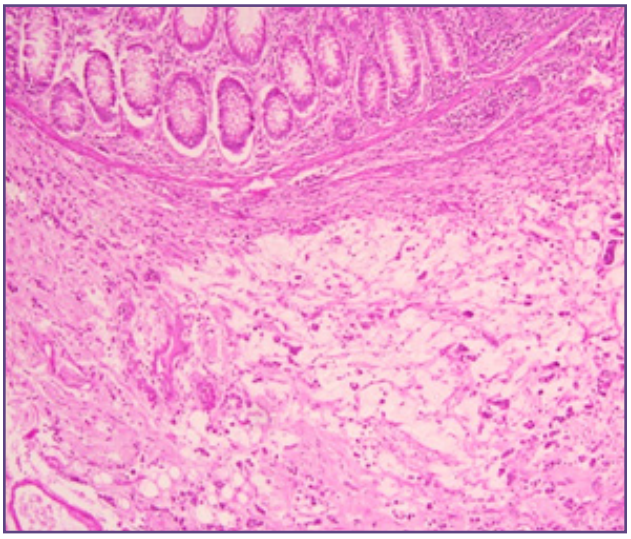

Fig. 1B: H&E section showing microscopic appearance at low power view (10x) of signet cells in the submucosa.

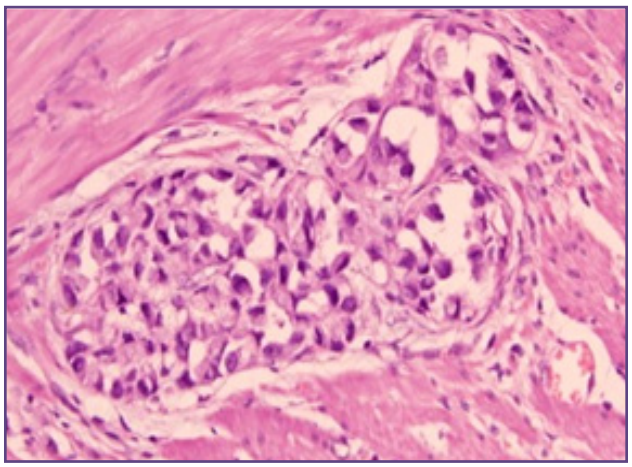

Fig. 2A: H&E section showing presence of signet cells at high power view (40x).

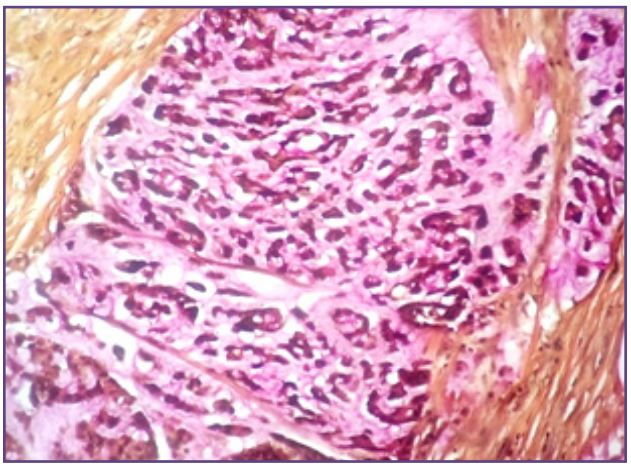

Fig. 2B: Mucicarmine stain showing positivity in both the signet ring cells and extracellular mucin.

Discussion

Signet ring cell carcinoma can be defined by the presence of signet cells comprising more than 50 percent of total tumour cells with prominent intracytoplasmic mucin. This tumour occurs across all age groups suffering from colon and rectal carcinoma.[1] The median age of presentation is 57.1 years.[1] This tumour is rarely reported in children and adolescent age group.[2,3,4,5] Right hemicolon is more frequently involved followed by left colon and rectum.[1]

The presenting features of tumor are rapid history of weight loss, rectal bleeding, abdominal pain, abdominal mass, vomiting, constipation and abdominal fullness. [5] The obstructive symptoms are more common due to stricture formation similar to clinical presentation of our case. In young age group, this can be often confused with tuberculosis due to obstructive signs and symptoms and can lead to a delay in diagnosis.

Colorectal signet cell carcinoma has a poor prognosis compared to conventional adenocarcinoma.[1,3] This is usually detected at advanced stages as the symptoms manifest in a very late stage with few cases detected at early stage.[1] Also there are higher chances of metastasis due to late manifestation, diagnosis and surgery. [6] The five year survival rate on review of various studies is 0% (median 15 months). [7] Thus the differential diagnosis of colorectal carcinoma especially signet ring cell carcinoma must be considered even in children and adolescent age group as delayed diagnosis will result in poorer survival of this disease which already has such a grave prognosis. In addition, in small biopsies, the diagnosis may be missed because the signet ring cancer cells may be misinterpreted as foamy macrophages. An important differential diagnosis is mucinous adenocarcinoma with 50% or more mucinous components. Signet ring cell carcinoma recurs more frequently compared to mucinous adenocarcinomas colon. [7]

It has been postulated in few articles that Signet ring carcinoma may also have precursor lesions [pre existing adenomatous polyp / denovo cancer] and may have multiple synchronous association.[8] Our patient did not have any family history of adenocarcinoma. On review of various studies in the literature in signet ring carcinoma K-ras mutation occurs at a lower frequency and B-RAF mutation has higher frequency compared to conventional adenocarcinomas. [9,10]

In conclusion in this case report, rare occurrence of primary signet ring cell carcinoma of the colon occurring in twelve year old child has been described as this is associated with poor survival and must be kept as differential diagnosis in children and adolescent age group. Earlier diagnosis with earlier surgery is necessary as the local and distant spread of this tumor is very rapid and responsible for the dismal prognosis.

Abbreviations

USG- ultrasonography

Acknowledgement

None

Funding

None

Competing interests

None Declared

References

1. Fu KI, Sano Y, Kato S, Saito H, Ochiai A, Fujimori T, Saito Y, Matsuda T,Fujii T, Yoshida S. Primary

signet-ring cell carcinoma of the colon at early stage: a case report and a review of the literature. World J Gastroenterol. 2006 Jun 7;12(21):3446-9.

2. Hamazaki M, Kono S, Mimaya J, Ishihara A. Signet ring cell carcinoma in a polyp of the colon. A case report of a six-year-old boy. Acta Pathol Jpn. 1987 Oct; 37(10):1679-84.
3. Thota R, Fang X, Subbiah S. Clinicopathological features and survival outcomes of primary signet ring cell and mucinous adenocarcinoma of colon: retrospective analysis of VACCR database. J Gastrointest Oncol. 2014 Feb;5(1):18-24.
4. Ko YS, Lin LH, Chen DF. Carcinoma of the colon in a child. Zhonghua Min Guo Xiao Er Ke Yi Xue Hui Za Zhi. 1995 May-Jun;36(3):227-30.
5. Marone J, Patel S, Page M, Cheriyath P. Signet ring cell carcinoma of the colon in a 17year old child. J Surg Case Rep. 2012 Sep; 2012(9): 3.
6. Tung SY, Wu CS, Chen PC. Primary signet ring cell carcinoma of colorectum: an age- and sex-matched controlled study. Am J Gastroenterol.1996 Oct;91(10):2195-9.
7. Secco GB, Fardelli R, Campora E, Lapertosa G, Gentile R, Zoli S, Prior C.Primary mucinous adenocarcinomas and signet-ring cell carcinomas of colon and rectum. Oncology. 1994 Jan-Feb;51(1):30-4.
8. Nakamura T, Nakano G, Sakamoto K. Adenoma of the rectum with multiple foci of signet-ring cell carcinoma. Report of a case. Dis Colon Rectum. 1983 Aug;26(8):529-32.
9. Sim HL, Tan KY, Poon PL, Cheng A. Primary rectal signet ring cell carcinoma with peritoneal dissemination and gastric secondaries. World J Gastroenterol. 2008 Apr 7;14(13):2118-20.
10. Ogino S, Brahmandam M, Cantor M, Namgyal C, Kawasaki T, Kirkner G, Meyerhardt JA, Loda M, Fuchs CS. Distinct molecular features of colorectal carcinoma with signet ring cell component and colorectal carcinoma with mucinous component. Mod Pathol. 2006 Jan; 19(1):59-68.