Scholars Journal of Medical Case Reports (SJMCR) **ISSN 2347-6559 (Online)**
Abbreviated Key Title: Sch. J. Med. Case Rep. **ISSN 2347-9507 (Print)**

A United of Scholars Academic and Scientific Society, India

Composite Tumour of the Rectum: A Diagnostic Dilemma

Susanta Kumar Das[1*], Anadi Nath Acharya[2], Debasis Bhattacharya[3]
[1]Assistant Professor, Dept. Of General Surgery Burdwan Medical College and Hospital, Burdwan, West Bengal, India
[2]Professor, Dept. Of General Surgery R.G.Kar Medical College, Kolkata
[3]Professor, Dept. Of General Surgery North Bengal Medical College and Hospital, Kolkata

***Corresponding author**
Susanta Kumar Das

Article History
Received: 10.06.2018
Accepted: 18.06.2018
Published:30.06.2018

DOI:
10.36347/sjmcr.2018.v06i06.011

Abstract: Rectal adenocarcinoma with additional neuroendocrine component is an extremely rare entity with about a little more a dozen of cases being reported in the literature. Mostly the neuroendocrine component is that of well-differentiated type (carcinoid); sometimes can be the unusual variant of neuroendocrine carcinoma capable of distant metastasis. Here we report a very rare case rectal cancer which was a diagnostic dilemma for us with the final histopathological examination revealing it to be a composite tumour of the rectum having adenocarcinoma and a more aggressive neuroendocrine carcinoma component with a solitary liver metastasis from the neuroendocrine component.
Keywords: Composite tumour, adenocarcinoma, neuroendocrine tumour, rectum, liver metastasis.

INTRODUCTION

Composite tumour of the rectum is an uncommon tumour of the rectum often presenting often with diagnostic dilemma in the initial endoscopic biopsy. Clinical features are a constellation of the histological components. Here we present a case of such composite tumour of the tumour having both neuro-endocrine and adeno-carcinomatous components.

CASE REPORT

In July, 2012, a 42 year old man presented to the outpatient department with history of vague abdominal pain for 6 months duration. An ultrasound examination of the abdomen done elsewhere revealed a space occupying lesion in the left lobe of the liver suggestive of hydatid cyst for which he was treated with oral albendazole without any improvement.

On examination he had pallor and non tender hepatomegaly. His white blood count was 9.5 X 10^9/L with 78% neutrophils. A CECT scan of the upper abdomen revealed a space occupying lesion in the left lobe of the liver (Fig.1). After no improvement on albendazole therapy he was further worked up. USG guided FNAC revealed a metastatic neuroendocrine tumour (Fig.2).

Then a search for a primary neuroendocrine tumour was done. The upper GI endoscopy was normal. The lower GI endoscopy revealed a well defined, circumferential friable growth about seven centimetres from the anal verge with narrowing of the rectal lumen. The scope could be negotiated through and the rest of the colonic mucosa and the distal part of the ileum were normal. The upper GI endoscopy was normal. Colonoscopic biopsy was taken. Serum CEA was mildly elevated (6.3 ng/ml).

Colonoscopic biopsy was suggestive of squamous cell carcinoma. In view of a contradicting biopsy report a repeat biopsy of the lesion was done which turned out to be adenocarcinoma. In consultation with the pathologist a repeat biopsy along with the previous biopsy slides were reviewed. The pathologist was now of the opinion that it could be a composite tumour of the rectum with both components of adenocarcinoma and neuroendocrine carcinoma.

This had led to a diagnostic dilemma as to the original histology of the rectal tumour. There were three possibilities that were put forward:

- The metastatic neuroendocrine component of the liver was a secondary from the neuroendocrine component of the composite rectal tumour.
- The liver SOL was a second primary neuroendocrine tumour existing along with the adenocarcinoma of the rectum.
- The rectal carcinoma was a neuroendocrine tumour with a secondary metastatic lesion in the liver.

In view of this entire dilemma, it was decided to take him up for surgery after the initial preoperative work up. It was decided to undertake the left lobectomy of the liver first and send for frozen section and then proceed for the resection of the rectal cancer. At laparotomy there was a large growth that could be palpated in the rectum with extensive involvement of the adjacent structures. There was a space occupying lesion measuring about 5x6 cm in size occupying the left lobe of the liver. The rest of the colon and small bowel was normal. There was no peritoneal dissemination.

We proceeded with left lobectomy of the liver for resection of the metastatic tumour. The patient had significant blood loss during the same and the hemodynamic condition of the patient became unstable. Considering the extensive involvement of the rectal cancer the rest of the procedure was put on hold. An end sigmoid colostomy was performed to take care of any obstructive symptoms that might occur at a later date.

The histopathological examination of the left hepatectomy specimen revealed a 5.5 x 5.5 metastatic SOL from a neuroendocrine carcinoma (Fig.3) as suggested by the FNAC report. Post operatively he had a prolonged hospital stay because of sepsis which was managed conservatively with IV antibiotics.

In consultation with the radiation oncologists he was offered a long course of chemo radiation because of extensive involvement of the rectal cancer. After the completion of the chemo-radiation the rectal tumour clinically had shown good response. A repeat CT pelvis was done and the tumour was deemed operable. He was again taken up for definitive treatment for his rectal lesion. An abdomino-perineal resection for the rectal tumour was done. The final histopathology report confirmed it to be a rectal cancer having areas of neuroendocrine (Fig.4) and focal areas of adenocarcinomatous components (Fig.5). Finally it cleared the entire dilemma regarding the diagnosis. It was a composite tumour of the rectum with the hepatic metastasis from the neuroendocrine component of the primary lesion. He responded well with the neoadjuvant chemo-radiation with significant reduction in the non-neuroendocrine components in the final biopsy.

He was discharged with advice to attend the Medical Oncology outpatient department for further adjuvant therapy. However he refused any adjuvant therapy.

He was on regular follow up for the last five years. His recent liver function test was normal. USG abdomen revealed regenerated left lobe of liver with no evidence of any new lesion. CT volumetric study of the liver showed residual liver volume of 1080 cubic centimetres (Fig.6). Serial three monthly serum CEA values were all within normal limits. Check colonoscopy through the stoma was found to be normal without any evidence of metachronous lesion. He is currently asymptomatic with good weight gain and managing his end sigmoid colostomy well.

DISCUSSION

Composite tumour of the rectum is a very rare with little knowledge about their origin and basic biology[1]. The term composite tumour is used to describe neoplasm that contains histologic features of both a malignant adenocarcinomatous and carcinoid component in juxtaposition to each other [2]. In some cases the neuroendocrine component is composed of more malignant variant such as the neuroendocrine carcinoma.[3] According to the recent WHO classification of endocrine tumours, they have been termed mixed exocrine-endocrine carcinoma[4].

In most cases the neuroendocrine component of these mixed tumours is made up of a well-differentiated endocrine tumour (carcinoid). The term mixed or composite carcinoma-carcinoid has been introduced to describe these neoplasms.

Lewin had proposed a classification of these tumours who had distinguished *composite* tumour, where endocrine and exocrine cells are intermixed within the same tumour, and *collision* tumour where two different histologic patterns are in close proximity [5]. While collision tumours are thought to arise next to one another coincidentally, mixed adenocarcinoma and neuroendocrine tumours are hypothesized to be true single neoplasm which differentiated bidirectional [6].

Here we reported a case of composite tumour of the rectum with metastatic lesion in the liver from the more aggressive neuroendocrine component. Only 14 cases of mixed carcinoma-carcinoid of the colon and rectum have been reported so far as isolated case reports. Only four cases have been reported where the neuroendocrine component had developed within a benign adenomatous lesion [7,8]. According to the recent WHO classification (2000), endocrine tumours of the colon and rectum are divided into well-differentiated endocrine tumours (carcinoid) and carcinomas (malignant carcinoid), poorly differentiated endocrine carcinomas (small cell carcinomas) and mixed exocrine-endocrine carcinomas. While carcinoids are characterized by their benign behaviour, malignant carcinoids are known to metastasize in up to 34.4% of cases. Atypical varieties, including small cell carcinomas, mixed exocrine-endocrine carcinomas and amphicrine tumours show an even higher rate of metastasis of up to 55.6% [9]. The metastatic potential from these neuroendocrine carcinomas depend on the primary site with jejunal and rectal malignant carcinoids

having rates of metastasis as high as 60%, while appendiceal carcinoids having the lowest rates[10].

In view of very little evidence in the literature regarding such composite tumours of the rectum and that too most of them being isolated case reports; no consensus regarding correct protocol of management of these tumours is achieved.

In our case it was more of a diagnostic dilemma regarding the proper histopathological diagnosis of the rectal tumour with the final HPE proving it to be a composite tumour of the rectum.

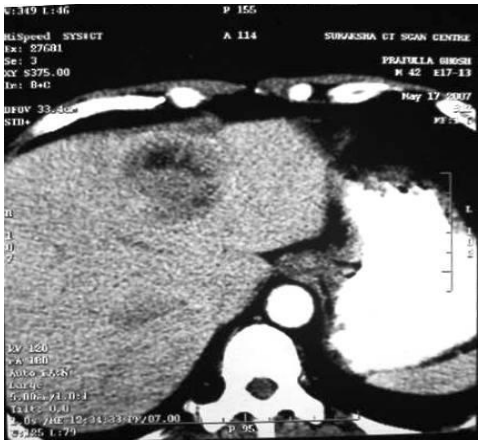

Fig-1: Space occupying lesion corresponding to the left lobe of the liver

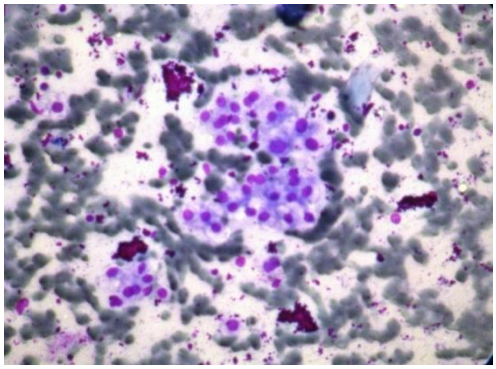

Fig-2: FNAC from the Liver SOL suggestive of metastatic neuroendocrine tumor

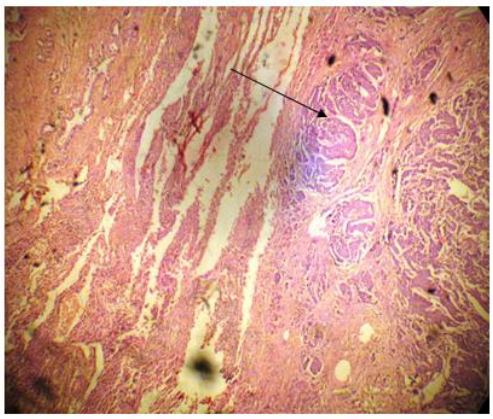

Fig-3: Metastatic deposits of carcinoid tumour in the HPE of left hepatectomy specimen

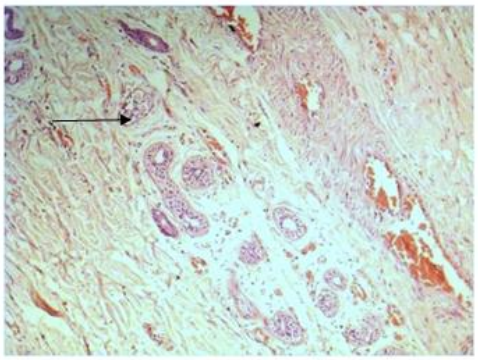

Fig-4: Final HPE: - Adenomatous glands within the muscle layer of rectum

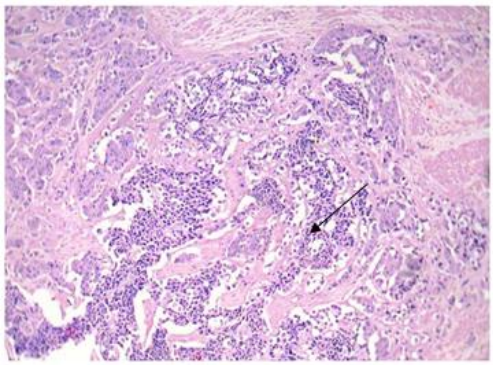

Fig 5: Final HPE: - Neuroendocrine component invading the muscle layer of the rectum

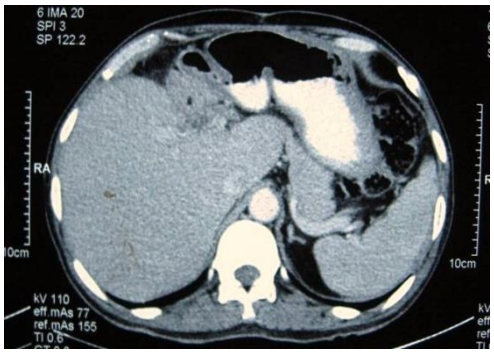

Fig-6: Recent abdomen with regenerated left lobe and no evidence of new lesion

REFERRENCES

1. Scott Klappenbach R, Kurman RJ, Sinclair CF, Patrick James L. Composite carcinoma–carcinoid tumors of the gastrointestinal tract: a morphologic, histochemical, and immunocytochemical study. American journal of clinical pathology. 1985 Aug 1;84(2):137-43.
2. Levendoglu H, Cox CA, Nadimpalli V. Composite (adenocarcinoid) tumors of the gastrointestinal tract. Digestive diseases and sciences. 1990 Apr 1;35(4):519-25.
3. Case records of the MGH. Case 28-2000. N Engl J Med 2000; 343:794-800
4. Endocrine Tumors of the Gastrointestinal Tract. In: Solcia E, Klöppel G, Sobin LH (eds) Histological Typing of Endocrine Tumors, 2nd edn. WHO Distribution and Sales, Geneva. 2000. p.61-68.
5. Lewin K. Carcinoid tumors and the mixed (composite) glandular-endocrine cell carcinomas. The American journal of surgical pathology. 1987;11:71-86.
6. Gibbs NM. The histogenesis of carcinoid tumours of the rectum. Journal of clinical pathology. 1963 May 1;16(3):206-14.
7. Mori K, Shinya H, Kalisman M. A composite tumor in tubulovillous adenoma of the rectum. Diseases of the Colon & Rectum. 1978 Oct 1;21(7):506-9.
8. Moyana TN, Qizilbash AH, Murphy F. Composite glandular-carcinoid tumors of the colon and rectum. Report of two cases. The American journal of surgical pathology. 1988 Aug;12(8):607-11.
9. Soga J. Statistical evaluation of 2001 carcinoid cases with metastases, collected from literature: a comparative study between ordinary carcinoids and

atypical varieties. Journal of experimental & clinical cancer research: CR. 1998 Mar;17(1):3-12.

10. Habal N, Sims C, Bilchik AJ. Gastrointestinal carcinoid tumors and second primary malignancies. Journal of surgical oncology. 2000 Dec;75(4):306-0.