Abstract:
35 yr old female a case of generalized neurofibromatosis was admitted with history of bleeding per rectum and Difficulty in passing stools for 3 months was found to have Palpable fungating mass on per rectal examination, the lower border is felt 5 cm from the anal verge upper border could not be felt. Biopsy done showed adenocarcinoma with carcinoid tumour. Immunohistochemistry done to support the diagnosis and is diagnosed a case of adenocarcinoid. CT scan taken showed Rectal wall growth with no perirectal invasion. Abdomino perineal resection with permanent colostomy was planned and executed. Post op period was uneventful. Patient was treated with adjuvant chemotherapy. This case is presented for its rarity since adenocarcinoid is common in appendix about 0.6 percent, but rectal adenocarcinoid is very rare, only seven cases have been reported in literature and for its management based on biological behaviour of the tumour. 

Keyword: Composite tumour of Rectum, Adenocarcinoid, Rectum

INTRODUCTION:
Adenocarcinoid tumour is a rare entity that possesses histological features of both carcinoid and adenocarcinoma. The term 'adenocarcinoid' was first coined by Warkel et al., in 1978(1). It has been reported that a majority of adenocarcinoids occur in appendix. Adenocarcinoid arising in rectum is extremely rare and only seven cases have been reported in literature(2-8). We present a case of adenocarcinoid of rectum with generalised neurofibromatosis.

CASE REPORT:
35 year female a known case of Generalised Neurofibromatosis, presented with complaints of bleeding per rectum and difficulty in defaecation for 3 months. Digital rectal examination showed palpable fungating mass. The lower border is felt 5 cm from the anal verge upper border could not be felt. Per abdomen and other systemic examinations no abnormality detected. Haematological examination was unremarkable except for anaemia (Haemoglobin-8.2gm%). ECG and Chest Xray were normal. USG done showed thickening of anterior
wall of rectum suggestive of carcinoma of rectum. CT abdomen and pelvis showed mass involving anterolateral aspect of rectum. Perirectal fat planes maintained, no evidence of uterus or Bladder invasion.

Colonoscopy done showed an ulceroproliferative growth extending about 5 cm from anal verge covering one third circumference of rectum, scope could be negotiated with difficulty. No other lesions seen up to caecum. Per rectal biopsy done showed Biphasic tumour population with adenocarcinomatous areas and neuroendocrine areas (Fig a and b). Immunohistochemistry done showed tumour were focally positive for chromogranin. The tumour also contained adenocarcinoma component (Fig c and d).

Planned and proceeded with Abdomino-perineal resection of Rectum with permanent colostomy after obtaining an informed written consent. Rectum with mesorectum excised into to, intraoperatively no carcinoid crisis occurred. Post operative period was uneventful. Patient was given adjuvant chemotherapy with 5-FU (Flourouracil) and Leucovarin. Patient got 4 cycles of adjuvant chemotherapy after which patient did not turn up for review.

DISCUSSION:
Composite tumours with histologic features of both carcinoid and adenocarcinoma are rare and their biologic behaviour is still unclear. Such tumours with combined characteristics have been found in various locations in gastrointestinal tract including stomach, small intestine, appendix, colon and gall bladder. Most cases reported in appendix which is also the most common location of carcinoid tumour in GIT. The aggressive clinical behaviour of these tumours were recognised with serosal extension, perineural invasion and metastasis to ovary and liver (9).

The histogenesis of composite tumours has not been fully characterised. Intimate arrangement with a sizable glandular and carcinoid component suggests a common histogenesis (composite), either occurrence of coincidental malignant changes in two mature cell types (collision) or a neoplastic change involving a common precursor cell (Amplcliffe). Clinically they behave more like an adenocarcinoma.

Edmonds et al in their review of 86 cases of composite tumour arising from appendix, found that 51 were disease free, 3 had persistent disease and 11 died of disease following treatment by colectomy or appendicectomy alone(10). Contrary to appendiceal composite tumours prognosis of colon composite tumours could not be determined due to less number of cases reported in literature and concentration was given more on histological evaluation of removed tissue and omitted information on clinical picture of these patients. Of seven cases of rectal adenocarcinoid tumour reported so far, four were male and three were female. Grossly two had ulcerated lesion, two had a sessile polypoid lesion and one was reported to have intact mucosa. Lymphnode metastasis were confirmed in five cases and liver metastasis in three. Three patients with lymphnode metastasis were found to be alive for more than 3 years after surgery. One large population based study demonstrated that a biological behaviour of adenocarcinoid of appendix is between malignant carcinoid and adenocarcinoma in terms of age at diagnosis and proportion of lymphnode involvement(11). Patients diagnosed with adenocarcinoid are less likely to have tumour spread beyond the colon; however they have an unusual propensity for ovarian metastasis(12). As for prognosis survival in adenocarcinoid tumour tends to be better than that in adenocarcinoma and worse than that in malignant carcinoid; however, the difference was not statistically significant(11). There is no consensus on the surgical treatment for adenocarcinoid tumour and there is debate on how aggressive the operation should be even in a case of appendiceal adenocarcinoid tumour that constitutes the majority of the entity.

Large clinical trials have suggested that adjuvant chemotherapy with 5-FU and leucovarin provides a significant survival benefit in patients with stage III colon cancer(13), and it was the standard adjuvant chemotherapy regimen in Japan at that time; however there is no consensus on adjuvant chemotherapy for adenocarcinoid tumour. Mandai et al reported a patient with appendiceal adenocarcinoid with bilateral ovarian metastasis who survived for 2 years after receiving cisplatin based adjuvant chemotherapy, which is standard for ovarian carcinoma (14). Hirschfield et al., reported a similar patient with bilateral ovarian metastasis and peritoneal seeding who benefited from chemotherapy with 5-FU and streptozotocin(15). Tomoyuki and colleagues from National cancer center hospital,
Tokyo showed that locally advanced rectal adenocarcinoid successfully treated by APR with total pelvic exenteration resulting in long term survival. Moreover groin lymph node recurrences were successfully removed twice, and it is possible that extended surgery is beneficial for patients with locally advanced rectal adenocarcinoid when resectable.

CONCLUSION:
We presented this case because of its rarity and because there is no strict guidelines regarding the treatment of these tumours and the radicality of resection to be done. But recent concepts regarding biological behaviour of the tumour gives us the idea of management and depending on the major cell type present. Majority of these tumours behave similar to pure adenocarcinoma and the management being targetted towards the same recently.

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