Colorectal Mucinous Adenocarcinoma in a 30-year-old Gentleman: A Rare Case Report

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ABSTRACT
Colorectal carcinoma is major cause of morbidity and mortality worldwide occurring in younger age group. Its histological subtype of mucinous adenocarcinoma (MC) is associated controversially with poor prognosis and rare regional lymph node metastasis. Family history of such malignancy is seen, if its microsatellite instability (MSI) associated. We present a rare case of a 30-year-old gentleman with recurrent episodes of dull aching pain in lower left abdomen, constipation, bleeding per rectum since last four years diagnosed as mucinous adenocarcinoma on histopathology and with family history of deaths due to mucinous colo-rectal carcinoma. On immunohistochemistry, CDX2 has been reported to be positive in 70% of colorectal mucinous adenocarcinoma, just like in our case.

KEYWORDS
Mucinous adenocarcinoma, CDX2

Introduction
Colorectal carcinoma is the fourth most commonly diagnosed malignant disease in the world occurring in younger patients.1,2,3 Mucinous adenocarcinoma (MC) accounts for 10%-20% of all colo-rectal carcinoma cases, mostly reported in western literature than in South Asia/India. MC is associated with controversially based poorer prognosis than non-mucinous adenocarcinoma (non-MC) of colon because most MC cases have been reported in advanced stages.4 MC should not be considered as an independent factor for poor prognosis because of controversial results.4 As the local mucin production is a major cause of morbidity and mortality worldwide occurring in younger age group. Its histological subtype of mucinous adenocarcinoma (MC) is associated controversially with poor prognosis and rare regional lymph node metastasis. Family history of such malignancy is seen, if its microsatellite instability (MSI) associated. We present a rare case of a 30-year-old gentleman with recurrent episodes of dull aching pain in lower left abdomen, constipation, bleeding per rectum since last four years diagnosed as mucinous adenocarcinoma on histopathology and with family history of deaths due to mucinous colo-rectal carcinoma. On immunohistochemistry, CDX2 has been reported to be positive in 70% of colorectal mucinous adenocarcinoma, just like in our case.

Case Report
A 30-year-old male patient came with complaints of lower abdominal pain, constipation, per rectal bleeds since last four years. Per rectal examination revealed a solid mass obscuring the lumen of the rectum. With clinical diagnosis of suspicious of malignancy, he was subjected to CT scan which confirmed it as a 8 x 5 cm annular growth with rectal wall thickening of 3 cm and obstruction which was unevenly concentric in recto-sigmoid region with heterogeneous enhancement pattern. Tumor showed low attenuation areas in more than 2/3rd of the tumor without intra-tumoral calcification or polypoidal growth or regional lymphadenopathy or peri-colic fat infiltration.

Further with CT guided biopsy, the diagnosis of adenocarcinoma was given. The surgeons subjected the patient for colono-
and 0.7x0.5x0.5 cm respectively without lymph nodes were separately sent.

Sections studied revealed an aggressive mucinous malignant tumor, arising from neoplastic, infiltrative mucinous glands. There were extra-cellular mucin pools with infiltration deep into the muscularis propria (80% tumor component) (Fig. 1). Also poorly formed/abortive neoplastic glands were seen (10% of tumor) (Fig. 2). The aggressive tumor was dissecting through the bowel wall.

The extracellular mucin – pools had neoplastic, malignant cells lining the pools and singly dispersed. These individual tumor cells were highly pleomorphic cells with very scanty cytoplasm, round to oval hyperchromatic nuclei and inconspicuous to prominent nucleoli at places. Areas of hyalinization of stroma were seen, adjoining the extra-cellular mucin pools. Atypical mitotic figures were seen at places.

Also focal necrosis and intra-vascular tumor emboli were noted. Signet–ring like cells were also seen infiltrating the stroma at places (<10% tumor) (Fig. 2). Lymphatic emboli were seen. There was mixed inflammation in stroma-few neutrophils, lymphocytes, plasma cells, histiocytes with foreign body giant cell reaction.

The fat deposits adherent to the tumor did not show evidence of tumor.

The sections from fibrofatty tissue, separate from main specimen did not show evidence of tumor. Lymph nodes were not identified grossly and microscopically within appendices epiploicae/fat deposits. It was thus diagnosed as colo-rectal mucinous carcinoma. The tissue sections containing mucinous tumor areas were mucicarmine/PAS positive and was CDX2 positive on IHC.

Discussion
Mucinous adenocarcinoma (MC) is a rare histological subtype of adenocarcinoma occurring in young people.2 The youngest reported MC case was a 9-months old baby.6 It usually involves the proximal colon as the primary site, unlike our case.2

It is characterized by large pools of extracellular mucin with malignant cells in cords, columns and dispersed singly. MC generally has no local inflammatory response around the mucinous deposits, scanty desmoplastic stroma and peri-colic tumor nodules with gelatinous components more than the primary colo-rectal tumor compared to non-MC malignancies.3 MC is often confused with signet ring cell carcinoma of colon which is rarer and more aggressive than the former because intra-cellular mucin can be confused with signet ring cells.4 The differentiation is possible by using histochemical staining like mucicarmine/PAS which highlight the mucin and thus differentiate from signet ring cells, like in our case.1

MC cases have been categorized into1: 1) pure mucinous type (PMA)-with extracellular mucin more than, that of tumoral volume, like in our case; 2) mixed type-50 to 80% of extracellular mucin (MMA); 3) mixed type with <50% of extracellular mucin (mMA). Pure mucinous category has advanced stage of presentation.1,3 mMA category cases were more in number followed respectively by PMA and MMA by one study.1

There were two subtypes reported of MC cases in histopathology:4 Mucocellular type which had poorer 3-year survival rate compared to papillotubular type. Our case was of mucocellular subtype of MC and was of PMA category and so, had poor prognosis accordingly1,4

Though microsatellite instability (MSI) is not an independent predictor for survival of MC cases, studies have shown that MSI associated MC cases had better prognosis than that of microsatellite stable MC cases.4 Family history of such colonic malignancy is seen, if its MSI associated. Hence there is importance of familial screening in such cases for bowel complaints.7 Our patient had family history of colonic malignancy, however this MSI tests were not carried out in our patient due to non-feasibility and non-affordability.

CDX2 has been reported to be positive in 70% of colorectal mucinous adenocarcinoma, just like in our case.7 Chemotherapy and radiotherapy are used as adjunct modes for control of local and distant metastasis or recurrence.8 Accordingly, our MC patient was subjected to APR surgery after completion of chemo-radiotherapy. Most studies of MC are retrospective studies2 and prospective study of cases is needed like in our case to understand this MC cases properly.

In conclusion, collaborative team-work between surgeons, physicians, radiologists, oncologists and pathologists is helpful in early diagnosis and control of disease. Effective family screening is important in such cases. A simple digital per rectal examination can help in diagnosis of colo-rectal malignancies with other radiological investigations. However even though histopathology remains the gold standard for MC diagnosis, immunohistochemistry-CDX2 positivity can help just like histochemical stains.

Figure 1a: Histopathological sections reveal extra-cellular mucin pools (80% tumor component) with infiltration deep into the muscularis propria (H&E, x40)

Figure 1b: Malignant cells are seen lining the mucin pools and singly dispersed. These individual tumor cells were highly...
pleomorphic cells with very scanty cytoplasm, round to oval hyperchromatic nuclei and inconspicuous to prominent nucleoli at places (H&E, x100).

Figure 2: Sections revealed poorly formed/abortive neoplastic glands (10% of tumor). Signet-ring like cells were also seen infiltrating the stroma at places (<10% tumor), which later stained positive for mucicarmine/PAS (H&E, x400).

References