

Small Cell Carcinoma of Rectum - An Infrequent Aggressive Tumor

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Abstract:- We report the case of a 63 year old man with small cell carcinoma of rectum. He presented with occasional rectal bleed, anal verge pain and generalized weakness. Colonoscopy revealed an ulcerated growth in proximal rectum. Endoscopic biopsy revealed poorly differentiated carcinoma. Laparoscopic anterior resection was done and histopathological examination along with IHC confirmed small cell carcinoma(SCC). Colorectal SCC is a rare and aggressive tumor.

Keywords:- Carcinoma Rectum, Small Cell Carcinoma, Rectal Bleed, Neuroendocrine Tumor, Small Cell Neuroendocrine Carcinoma.

I. INTRODUCTION

Small-cell colon carcinoma is a very rare disease among colonic neoplasms. Small-cell carcinomas (SCC) are malignancies derived from cells of neuroendocrine system. Gastrointestinal tract has the largest component of neuroendocrine cells, and neoplastic proliferation of these cells occurs primarily in the appendix, ileum and rectum^[1].

Incidence of colorectal SCC is less than 0.2% among all kinds of colorectal cancers^[2]. The prevalence is higher in Asian population. Patients with neuroendocrine neoplasm (NEN) often present in the sixth or seventh decade. A small proportion of these tumors are associated with adenomatous polyps ^[4,5].

Small cell neuroendocrine carcinoma (SCNEC) most frequently arises in the lung, outnumbering the extrapulmonary locations by 10:1. SCNEC can arise as a component of a non-neuro endocrine carcinoma reflecting clonal genomic evolution. Increased risk is associated with a family history of cancer, tobacco smoking, alcohol consumption and increased body mass index ^[12].

II. CASE REPORT

A 63 year old male who is a surgeon by profession, presented with history of colonic pseudopolyps 20 years ago and has been under follow up. He underwent screening colonoscopy every 2 years, which revealed no progression in number and size of polyps. He underwent MRI abdomen and pelvis as a part of workup of his recent abdominal pain and it revealed a flat focal sessile mildly irregular polypoidal thickening involving the lateral/posterior wall of rectosigmoid with focal wall thickening of 10 mm. Multiple small adjacent irregular pericolic lymph nodes were noted, largest measuring 1.4 x 1.2 cm. These findings were suggestive of proximal rectal neoplastic wall thickening with local pericolic lymphadenopathy. There was no abnormality on physical examination except for an abdominal scar due to previous appendectomy and his performance status was ECOG - 0. There were no abnormalities found on laboratory tests.

Colonoscopy showed ulcerated growth with slough in rectosigmoid junction. Microscopic examination revealed of poorly differentiated carcinoma. Immunohistochemical staining with INSM1, synaptophysin and chromogranin were strongly positive with Mib1 of 80% , which suggested neuroendocrine carcinoma (Fig 3). Tumor was negative for LCA, NKX 3.1 and CK5/6 ruling out differentials of lymphoma, prostate carcinoma and basaloid squamous cell carcinoma respectively. He underwent laparoscopic anterior resection and colorectal anastomosis. Macroscopically cut surface showed an ulcerated area measuring 2.4 x 2.0 cm with a depth of 1.0 cm. Histopathology was suggestive of small cell carcinoma, invading through muscularis propria into pericolic/subserosal tissue with mitoses of 60/2mm² and 2/13 lymphnodes were involved by tumor (Fig 1&2). The tumor stage was pT₃ N1b. Post surgery, he had an uneventful recovery and was initiated on adjuvant chemotherapeutic regimen constituting Cisplatin and Etoposide .

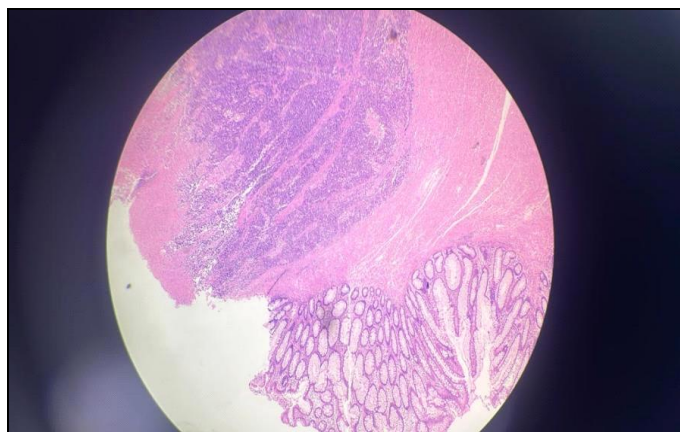


Fig:1 Small Cell Carcinoma Cells Filling up Muscularis Propria (Low power 10x)

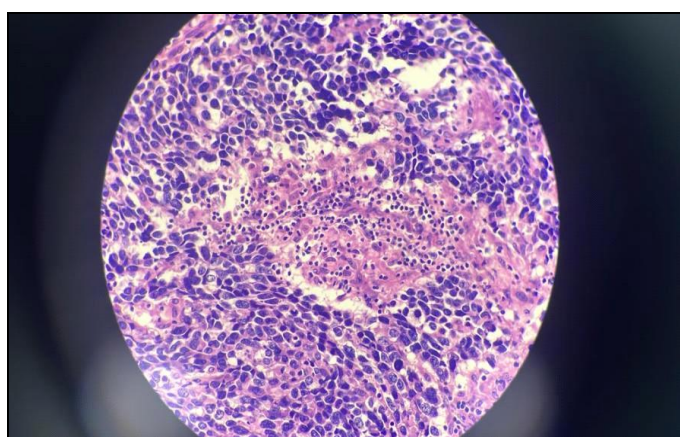


Fig:2 Tumor Composed of Small Cells Arranged in Sheets with Brisk Mitoses and Necrosis (High power 40x)

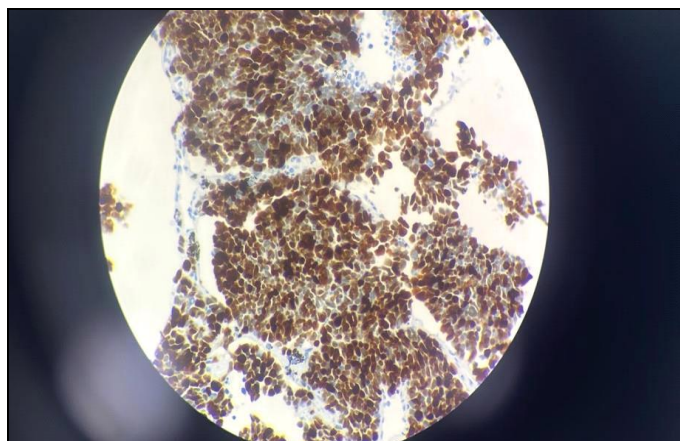


Fig:3 INSM1 Diffuse and Strong Nuclear Positivity (High power 40x)

III. DISCUSSION

Small cell carcinoma of the colon is a rare and aggressive type of cancer which originates from neuroendocrine cells in the lining of large intestine. It is also known as neuroendocrine carcinoma of the colon (NECS) or colorectal small cell carcinoma. NECS show a male predominance, displays an ominous outcome which is directly related to ki 67 proliferation, with ki 67 < 55% having a median overall survival of 25.4 months; where as

patients with higher Ki 67 proliferation have a median survival of 5.3 months ^[12].

The exact cause of small cell carcinoma of the colon is not known, but some risk factors may include smoking, family history of colorectal cancer, inflammatory bowel disease, and genetic mutations ^[9]. The symptoms of small cell carcinoma of the colon may vary depending on the location and size of the tumor; they may include abdominal pain, weight loss, blood in the stool, diarrhea, constipation, and bowel obstruction⁹.

The diagnosis of small cell carcinoma of the colon is based on histological and immunohistochemical features of tumor cells. The tumor cells are usually small, have high mitotic rate, are positive for cytokeratin, INSM1 chromogranin, synaptophysin, and CD56. TTF1, CDX2 may also be positive. Biallelic inactivation of TP53 and RB1 is by far the most common genomic alteration in SCNEC irrespective of the primary site^[12]. Small cell neuroendocrine carcinoma should be distinguished from other “small cell” neoplasms, including lymphomas, melanomas, or basaloid squamous cell carcinoma by immunohistochemistry.

The treatment of small cell carcinoma of the colon depends on the stage and extent of the disease, but it usually involves surgery to remove the tumor and the surrounding lymph nodes, followed by chemotherapy and/or radiation therapy. Some targeted therapies, such as epidermal growth factor receptor (EGFR) inhibitors, may also be used in some cases⁸. The prognosis of small cell carcinoma of the colon is poor, with a 5-year survival rate of less than 10%^[9]. Main factors that affect the survival are tumor stage, presence of distant metastases, and response to treatment.

IV. CONCLUSION

Small Cell Carcinoma of rectum is a rarely seen tumor and has a rapid progression. Diagnosis requires immunohistochemistry and treatment is surgical resection followed by adjuvant chemotherapy in advanced cases. Identification needs surveillance strategy in high risk cases and early identification prolong survival.

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