

Primary squamous cell carcinoma of the sigmoid colon

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ABSTRACT

Primary squamous cell carcinoma (SCC) of the colon and rectum is a rare malignancy. Overall there are less than 150 cases which have been reported and the first case was reported in 1919. We report a case of pure SCC involving the sigmoid colon. A 55 year old woman presented with chief complaints of bleeding per rectum for 5 months, anorexia and abdominal pain. Histological diagnosis of SCC was made by colonoscopy biopsy. Later it was confirmed by immunohistochemistry (IHC). Any other possible primary sites of malignancy were excluded. Complete surgical resection was performed. The prognosis of this disease seems to be worse than that of adenocarcinoma.

Keywords: *Abdominoperineal resection, Colon carcinoma, Squamous cell carcinoma.*

Adenocarcinoma represents the vast majority of colorectal cancers (CRCs). Non-epithelial malignancies such as neuroendocrine tumors, sarcomas, and lymphoid tumors account for majority of the remaining cases. Squamous cell carcinoma (SCC) is of epithelial origin and the incidence of SCC of the colon and rectum has been reported to be 0.25–0.1/1000 colorectal carcinomas [1]. Though neuroendocrine or squamous cell differentiation may be seen in colorectal adenocarcinoma, a pure colorectal SCC without anal canal involvement is an extremely rare tumor entity [2]. Because of the relative rarity of the disease, evidence based approach is not possible. Herein, we report a case of primary SCC of the sigmoid colon with local infiltration.

CASE REPORT

A 55 year old woman presented with complaints of painless bleeding per rectum for 5 months with anorexia and abdominal pain. She was referred to us after failed conservative treatments by a local physician. The patient had a history of abdominal hysterectomy for uterine fibroid 15 years back. There was no significant addiction or family history but was a known diabetic which was controlled with medications.

She was hemodynamically stable and her systemic examination was normal. The patient had pallor and a mass palpable in the right lower abdomen, on examination. Colonoscopy revealed a large ulcero-proliferative growth in sigmoid colon compromising lumen [Fig. 1]. A biopsy was taken from the growth during colonoscopy, considering the diagnosis of malignancy like adenocarcinoma. However, histopathological

examination proved this to be SCC [Fig. 2]. IHC (p63) [Fig. 3] of the specimen was also done for confirmation. Contrast enhanced abdominal computed tomography (CT) showed [Figure 4] an oval shaped heterogeneously enhancing soft tissue mass in right side of pelvis, measuring 4.5 cm x 3.5 cm. The tumor had encased the right ureter partially and right sided hydronephro-ureteric changes were seen. Few pelvic nodes were also evident in scan.



Figure 1: Colonoscopy showing Ulcero-proliferative growth at sigmoid colon.

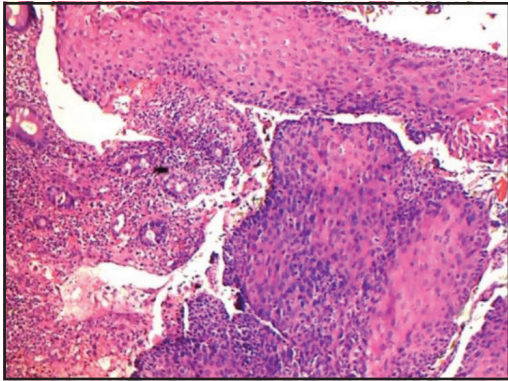


Figure 2: Histopathological examination showing keratin formation in SCC rectum specimen (in 100x hematoxylin and eosin stain).

CT scan of neck and chest did not reveal any abnormality. Both upper gastrointestinal endoscopy and laryngoscopy were within normal limit. Patient's ECOG performance status was 01. On routine investigations, we found Hb%-7.3 gm%, urea-60 mg/dl (normal 10-50 mg/dl), creatinine -1.6 mg/dl (normal 0.7-1.3 mg/dl). Carcinoembryonic antigen (CEA) was 5.75ng/ml (normal \leq 4ng/ml). Clinical Stage III withT4N1M0 (as per AJCC 8th edition) was established. Preoperatively the anemia was corrected with blood transfusions.

The primary surgery was decided after multidisciplinary tumor board consultation. Intra-operatively, tumor was found in sigmoid colon adhered to posterior bladder wall encasing the lower part of right ureter. En-block resection of sigmoid colon with part of bladder and distal part of ureter was done. Mucosa of bladder was free. Primary closure of bladder with reimplantation of ureter over D-J stent was performed. Bowel continuity was maintained by stapled anastomosis between descending colon and rectum. Patient was discharged on 7th postoperative day in stable condition. Final pathological staging was pT4N0M0, Grade-2 SCC with all margins free. Patient received adjuvant chemotherapy of 8 cycles with – oxaliplatin (180 mg on 1st day) and capecitabine (1000 mg twice daily for next 14 days). Now patient is on follow up for last 18 months.

DISCUSSION

SCCs of the colon are an extremely rare clinical entity. The first case of a pure SCC of the colon was reported in the German literature by Schmidtman in 1919 [3]. Colorectal SCC had been found mostly in rectum (93.4%) followed by right colon. Females are affected in 2/3rd cases with mean age of presentation between 55-65 years [4]. The mechanism underlying the origin of SCC from the colorectal mucosa remains unclear, but several mechanisms have been suggested. A proposed hypothesis is that SCC results from the proliferation of basal cells following mucosal injury and squamous differentiation within an adenoma [5]. Chronic inflammation (inflammatory bowel disease, human papillomavirus, amoebiasis, and schistosomiasis), radiation therapy and neoplasm in an adjacent organ (ovary, endometrium, cervix and prostate) have also been reported to be associated with some colonic SCCs [6].

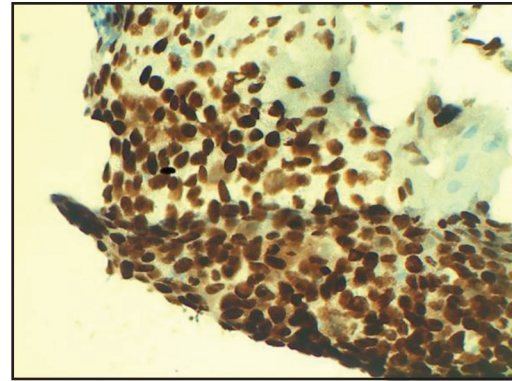


Figure 3: Immunohistochemistry showing p63 stain positivity in SCC rectum specimen.

Certain criteria must be satisfied before a diagnosis of primary SCC of the colon is made [7]. Metastasis from other sites to the bowel must be ruled out. A squamous - lined fistulous tract must not involve the affected bowel. SCCs of the anus with proximal extension must be excluded and its diagnosis must be confirmed by IHC analysis. Our case satisfied all these criteria.

Clinical presentation, diagnosis, and staging of colon and colorectal SCC are similar to primary colonic adenocarcinoma [8]. These tumors are usually aggressive and advanced at the time of presentation, leading to complications such as bleeding, bowel obstruction and urinary obstruction secondary to local spread [9]. In 1971 Comer *et al.* [10] suggested a poorer prognosis for patients with colorectal SCC than adenocarcinoma. Frizelle *et al* [6] had suggested nodal involvement, right sided lesions, ulcerated or annular carcinomas, grade 3 or 4 disease and stage IV disease to be associated with poor prognosis. Because of the rarity of these tumors, actual prognosis of patients with colorectal SCC is difficult to ascertain [10], but seems to be dismal.

The colorectal SCC seems to be more frequently locally invasive and more likely to involve regional lymphatics than adenocarcinoma, probably because of a delayed diagnosis. As a tumor marker, CEA may not be suitable as evident in our case. The proposed tumor marker for colorectal SCC is squamous cell carcinoma antigen (SCC Ag), which has already been used for SCC lung, uterus, cervix, esophagus, head and neck. Some literature highlighted its prognostic value as elevations of SCC Ag correlates with recurrence of this disease [11]. Unfortunately, we were not able to get SCC Ag levels for our patient. Staining with cytokeratin's AE1/AE3, CK 5/6 and p63 for cells of squamous origin, assists in differentiating from adenocarcinoma [4].

Historically the treatment is primarily surgical, depending upon the location and stage of the disease. Adjuvant treatments can include Cisplatin/5-Fluorouracil based chemotherapy as used for head and neck SCC [6]. Juturi *et al* [12] have also reported encouraging results, and even complete remission in one case using the same combination chemotherapy with addition of Leucovorin. Copur *et al* [11] reported that cisplatin, etoposide, and 5fluorouracil combination chemotherapy was effective and serum SCC antigen level was a useful marker of response to chemotherapy. Recent series have shown survival advantages of primary chemo-radiotherapy over conventional treatments [4]. However, the

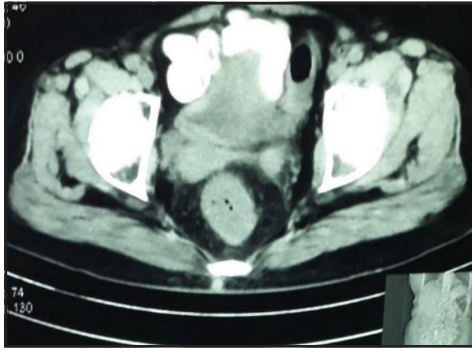


Figure 4: CT abdomen showing heterogeneously enhancing soft tissue mass in right side of pelvis.

condition has been encountered so rarely that adequate evaluation of adjuvant therapies has not been possible so far.

CONCLUSION

Primary SCC of the colon and rectum is a rare tumor; however, it is important that surgeons and pathologists dealing with colorectal malignancy keep its existence in mind. Perhaps with a greater awareness we will see more cases reported resulting in a better understanding of this variant of colorectal carcinoma leading to its recognition as a regular entity.

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