Primary squamous cell carcinoma of cecum- Case report and review of literature

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Abstract:
We report a rare case of primary squamous cell carcinoma of cecum. A 74 year old lady was operated for tumor of cecum involving adjacent ascending colon. We received right hemicolectomy specimen showing large, necrotic and friable mass arising from mucosal aspects of cecum. Microscopically tumor consisted of nests of atypical squamous epithelial cells, which was confirmed on immunohistochemistry. Keeping in mind the diagnosis of primary squamous cell carcinoma, additional examination was recommended to rule out possibility of metastasis from other sites. Extensive clinical examination including PET/CT scan showed no additional tumor. After exclusion of possible metastatic disease, the diagnosis of primary squamous cell carcinoma of cecum was confirmed. Primary squamous cell carcinoma of cecum is exceedingly rare malignancy. Before such a condition is accepted genuine care must be taken to exclude metastatic tumor presenting as primary growth, most likely from cervix in females.

Key words: Cecum; colon; Metastasis; Primary; Squamous cell carcinoma

Introduction:
The occurrence of primary squamous cell carcinoma in the gastrointestinal tract is rare phenomenon and its occurrence in the colorectum is extremely unusual [1]. The incidence of primary squamous cell carcinoma of colorectum has been reported to be 0.1 to 0.2 per 1000 colorectal carcinomas [1]. When they occur there is often suspicion that they are secondary to an occult primary present elsewhere. The presentation of a patient with this unusual lesion and review of previously reported cases of primary squamous cell carcinoma of colon form the basis of this report.

Case Report
We are reporting a case of 74 year female presented with abdominal pain, nausea and generalized weakness for last four month. Clinically the patient was anemic and had ill defined palpable pelvic mass was found on the right side of the abdomen. Per-rectal examination did not reveal any abnormality. Barium enema showed filling defect in the cecum and part of ascending colon. USG revealed large solid space occupying lesion measuring 14 x 13 cm in right lower abdomen and pelvis. Patient had a hysterectomy one year back for leiomyoma.
Pathological findings –
Right hemicolectomy was performed. There were no gross signs of metastasis to liver and lymph nodes. We received 30 cm long specimen of intestine comprising of ileum, cecum with tumor and ascending colon. On cut opening large necrotic friable growth measuring 14.8 x 13 x 6 cm. is seen in cecum and part of ascending colon. Serosa of ascending colon is adherent to cecal mass. Nine pericolic lymph nodes were resected along with tumor, were grossly unremarkable.

Microscopically multiple sections reveal nests of atypical squamous cells involving mucosa, sub mucosa and muscularis propria, however serosa was uninvolved. Intercellular bridges were evident. These nests of atypical squamous cell were positive for p63 and CK 5/6 and negative for CDX2, CK20, ER, PR. Mucicarmine and AB-PAS stains were negative indicating absence of mucin in the tumor cells and thus ruled out adenocarcinoma component. Keeping in mind the diagnosis of primary squamous cell carcinoma additional examination was done to rule out possibility of metastasis from other site, extensive clinical examination including PET/CT scan showed no additional tumour.

After exclusion of possible metastatic disease diagnosis of primary squamous cell carcinoma was done.

Discussion
Primary squamous cell carcinoma of colon was first reported by Schmidtmann in 1919 in 65 year old patient [2]. In India Bhatt et al reported first case of primary squamous cell carcinoma of colon in 1993 in 55 year old female [3]. Until now almost 150 cases of primary squamous cell carcinoma have been reported from all over world. Before diagnosis of squamous cell carcinoma of colorectum is made certain criteria must be satisfied as given by Williams et al 1979[4]. This criteria includes:
1. Absence of evidence of carcinoma of any part of body, ruling out any chance of possible metastasis from any organ to colorectum.
2. Exclusion of any proximal extension of anal carcinoma. By convention, tumor 7 cm proximal to dentate line usually has been excluded from discussion of squamous cell carcinoma of colon.
3. Absence of fistulous tract lined by squamous cell.
4. SCC must be confirmed by histological analysis [1].

Our case satisfied all these criteria.

A look at available literature reveals that squamous cell carcinoma of colorectum affect individual with mean age of 55-60 years. Women are more frequently predispose to SCC than men. Furthermore SCC occurs inconcomittence in advanced tumor stage (Dukes 5/6) [1, 5].

Clinical features of primary squamous cell carcinoma are exactly same as adenocarcinoma of colon. However there is significant risk of micro-metastatic disease in these patients. Therefore adjuvant therapy is given to eradicate any microscopic metastatic disease and to increase overall survival benefit [6].

Several pathogenic theories regarding origin of squamous cell carcinoma of colon has been proposed in literature [7, 8].
1. Proliferation of uncommitted basal cells in response to chronic mucosal injury.
2. Squamous metaplasia of glandular epithelium resulting from chronic irritation.
3. Origin from embryonal nests of ectodermal cells.
4. from stem cells.

Adenosquamous carcinoma of colorectum is considered variant of same tumor but agreement is lacking in terms of the amount of squamous epithelium needed for diagnosis [9].

Comer et al, suggested poorer prognosis for patients with colorectal SCC than adenocarcinoma [10]. Of patients who have recurrences after curative resection of colorectal carcinomas, 80% do so within three years. Therefore any follow up plan should have higher frequency of follow up during these three years and in decreasing frequency thereafter.

The present case is based on a one and half year follow up and is free from any symptoms/recurrence until now.

Conclusion
Primary SCC of cecum is exceedingly rare malignancy, before such a condition is accepted genuine care must be taken to exclude metastatic tumor presenting as primary growth.

SCC of cecum is rare and aggressive malignancy with worse prognosis, so it requires multimodality approach including surgery, chemotherapy and radiation.

References

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Figure 4: Squamoid cells with vague intercellular bridges

Figure 5: Atypical squamous cells positive for CK5/6

Figure 6: Atypical squamous cells positive for p63

Figure 7: Atypical squamous cells negative for mucicarmine stain