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Primary Squamous Cell Carcinoma of The Small Intestine- A Rare and Intriguing Case Report

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Abstract

Primary gastrointestinal malignancies arising from the small intestine constitute less than 2%. Primary small intestinal carcinomas are very less frequent than metastatic tumors of the small intestine, amongst which 30% to 50% are adenocarcinomas, 25% to 30% are carcinoids and 15% to 20% are lymphomas. Here, we report a 62 year old male male presented with abdominal pain, loss of appetite and weight for two months with histopathology showing features of primary squamous cell carcinoma of ileum, which is an extremely rare presentation with only a few cases reported in the literature.

Keywords: *Primary squamous cell carcinoma, small intestine, small bowel resection, keratin pearl, atypical mitosis.*

Introduction

Malignant tumors of the small intestine are uncommon compared to other malignant tumors of gastrointestinal region. Primary gastrointestinal malignancies arising from the small intestine constitute less than 2%. Primary small intestinal carcinomas are very less frequent than metastatic tumors of the small intestine, amongst which 30% to 50% are adenocarcinomas, 25% to 30% are carcinoids and 15% to 20% are lymphomas.**[**1**]** Squamous cell carcinomas of the small intestine, either primary or metastatic are extremely rare and only a handful of cases have been reported in the literature.**[**2**]** Here, we report a case of primary squamous cell carcinoma of ileum.

Case Report

A 62 year old male presented with abdominal pain, loss of appetite and weight for two months with few episodes of vomiting. Examination revealed a vague mass palpable in the left iliac fossa and suprapubic region. Computed tomography (CT) revealed an irregular circumferential and segmental wall thickening involving the mid jejunum and proximal ileum. Small bowel resection and anastomosis was performed following which the specimen was sent for histopathological examination. On gross, the segment of bowel had a nodular external surface. Cut section showed an irregular circumferential thickened growth measuring 8.5x6.0x1.5cm with a grey white, firm to soft cut surface which reached

upto the serosa. Tumor distance from proximal margin, distal margin and serosa were 11cm, 9.5cm and less than 0.1cm respectively.

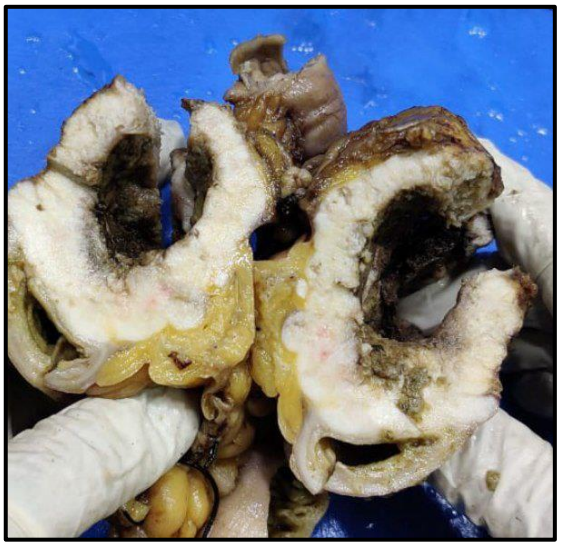

Figure 1: Circumferential tumor with the firm grey, white cut surface seen infiltrating the subserosal adipose tissue.

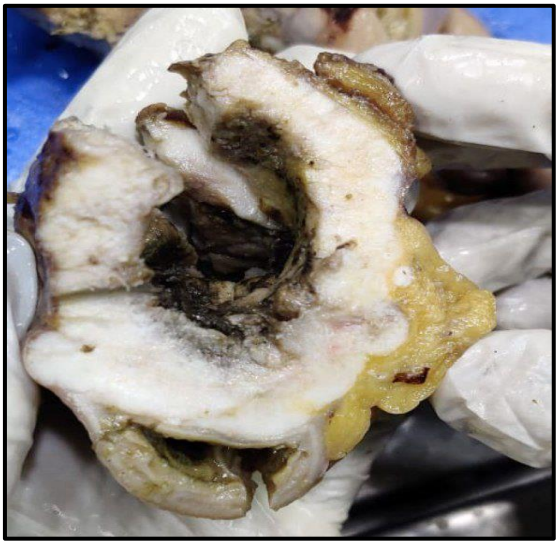

Figure 2: There is clear demarcation between overlying mucosa and underlying tumor.

Microscopy showed an invasive neoplasm composed of malignant squamous cells arranged in nests, islands and sheets **[**Fig 1**]**. The tumor cells were polygonal, with moderate nuclear pleomorphism and moderate amounts of eosinophilic cytoplasm. Occasional multinucleated tumor cells were seen **[**Fig 2**]**. Mitosis was brisk (approximately 10 per 10 high power fields). Tumor was seen to infiltrate upto the subserosal adipose tissue **[**Fig 3**]**. Multiple foci of lymphovascular invasion seen **[**Fig 4**]**. Keratin pearls were distributed throughout the tumor **[**Fig 5**]**. Also seen were large areas of tumor necrosis **[**Fig 6**]**. We obtained four lymph nodes which were free of tumor. Based on the above findings, a diagnosis of well-differentiated squamous cell carcinoma, histological grade I was reported. Pathologic stage was given according to pTNM, AJCC eighth edition as pT3 and pN2.

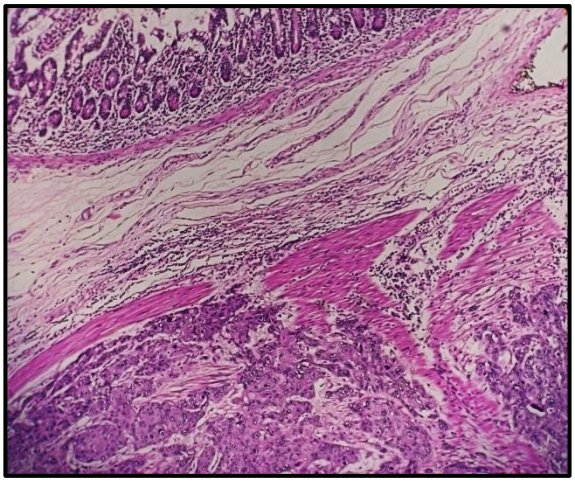

Figure 3: Small intestinal mucosa with underlying nests of squamous epithelial cells (H & E) - 100X.

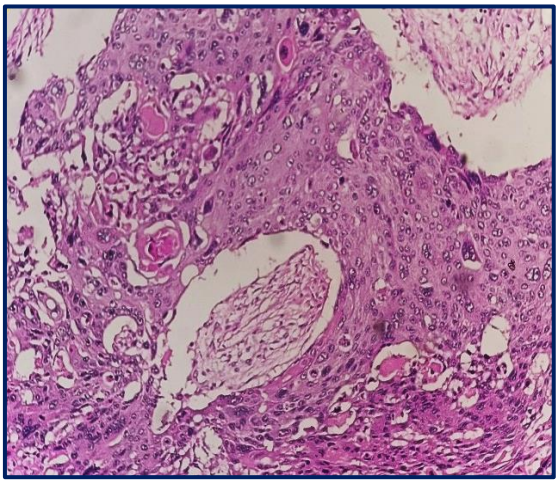

Figure 4: Several bizarre and multinucleated tumor cells (H & E) – 200X.

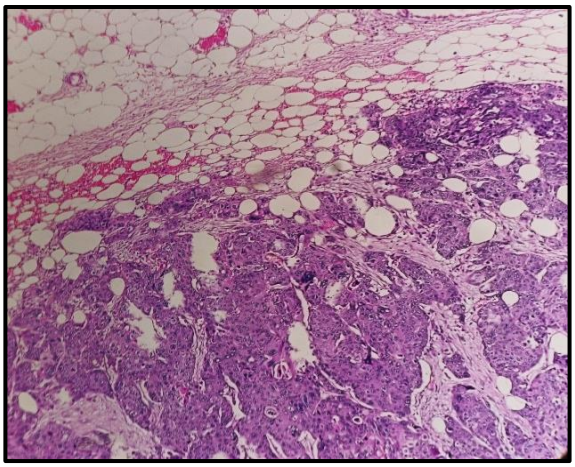

Figure 5: Tumor seen infiltrating the subserosal adipose tissue (H & E) – 100X.

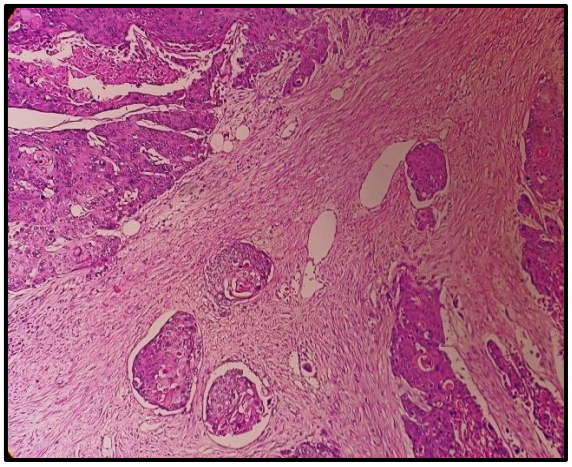

Figure 6: Several foci of lymphovascular invasion (H & E) – 100X.

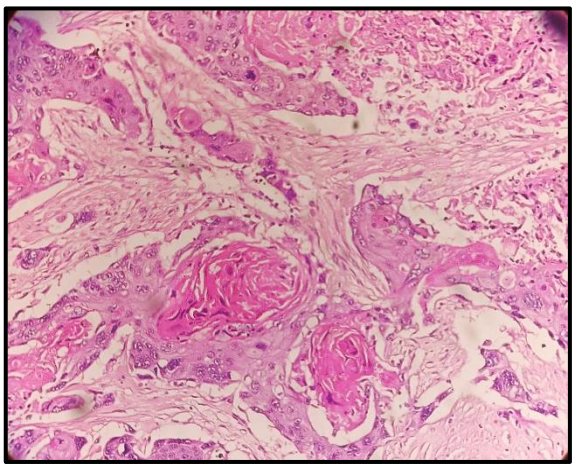

Figure 7: Keratin pearl formation with atypical mitosis (H & E) – 200X.

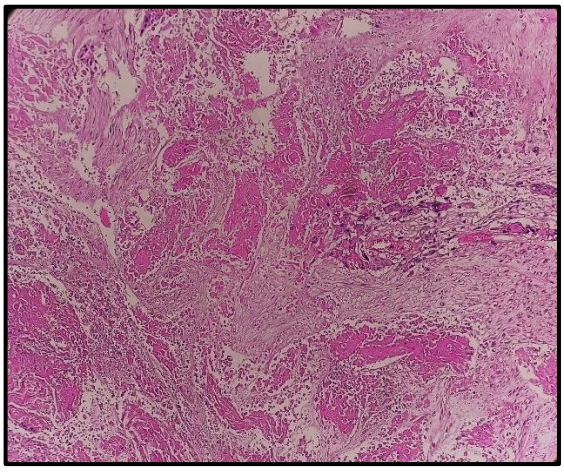

Figure 8: Large areas of tumor necrosis were seen (H & E) – 100X.

Discussion

Being an extremely rare malignant tumor, the first case of primary squamous cell carcinoma of the small intestine was reported in a 65-year-old man by Adair and Trowell in the year 1981.[3] So far, there are only very few cases reported in the English literature with primary squamous cell carcinoma of the small intestine confirmed either by biopsy or surgery, in addition to the case reported in our present study. According to the previous reports, these tumors were located in the duodenum, jejunum, ileum and diverticula of the small intestine with the duodenum being the most affected, followed by jejunum which is more common than the ileum.[3,4] In the duodenum, it is mostly located at the peripapillary area in the second part, but also cases in third part of duodenum also reported.[5] In most cases of small intestinal tumors, metastases remains the most common cause of squamous cell carcinoma, as secondary tumors of the small intestine are 2.5 times more frequent than the primary tumors. Those metastases most commonly arise from carcinomas of lung, breast, kidney and colon.[1]

Pathologically, it is difficult to determine whether it is a primary or metastatic squamous cell carcinoma when found in the small intestine as squamous cells are not seen there normally. There are four possible mechanisms proposed: (1) malignant transformation of heterotropic squamous epithelium, (2) pleuripotent stem cells differentiate to malignant squamous cells, (3) squamous metaplasia due to chronic mucosal damage undergoes malignant change, (4) adenocarcinoma transformed into adenosquamous carcinoma and eventually to SCC.[3] Though these mechanisms lack solid evidence, previous cases reported in the literature may provide a clue for the possibilty. In our case, typical squamous differentiation such as dysplastic squamous cells, keratinization with atypical mitosis and intercellular bridges without glandular differentiation were prominently seen. Though infiltrating squamous cell carcinoma upon histopathological examination is classic, it was very difficult to determine whether it is primary or secondary. Our case had no history suggestive of a primary squamous cell carcinoma elsewhere in the body. In microscopy, squamous metaplasia were seen in several foci in the epithelium. There was no underlying adenoma, inflammatory disease or duplication. With this background, we can further substantiate the fact that the squamous cell carcinoma may be of primary origin rather than a metastatic possibility. Most metastatic tumors are seen in submucosal region whereas in primary squamous cell carcinomas, the mucosa is always involved.[3]

Immunohistochemistry has limited value as it does not reflect the primary tissue expression in case of metastatic tumors. So, PET/CT is an effective tool to distinguish between primary and secondary tumors. Role of imaging in primary squamous cell carcinoma of small intestine is useful in locating, staging and assisting the surgeons to determine the resectable tumor.[3] Clinical features of squamous

cell carcinoma of the ileum are similar to that of other ileal neoplasms. Appropriate treatment and the prognosis are deceptive due to the rarity of this disease. Surgery remains the mainstay in the management of the condition. Possibility of recurrence and effects of radiotherapy and chemotherapy are still unclear as very few cases are available in the literature.**[**2**]**

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