Case Report

Signet Ring Cell-mucinous Adenocarcinona of The Descending Colon, A Case Report

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Abstract:

Colorectal carcinomas are quite rare in pediatric age group with a very poor prognosis and are usually beyond the scope of operative resection(1). The predominant histological type in children is the poorly differentiated mucinous adenocarcinoma which carrires a poor prognosis. Here. We present an interesting case report of a 14 years old male who presented to surgical emergency as acute intestinal obstruction and on laparatomy, a descending colon growth was identified for which resection and anastomosis with right transverse colostomy was done. On histopathological examination, the growth was found to be a signet ring cell mucinous adenocarcinoma and the patient responded well to chemotherapy. Key words: Signet ring cell, mucinous adenocarcinoma.

Introduction:

Gall Carcinoma when is a disease of the aged with majority of patient in the 6th, 7th decade of life. The age standardized rates of colorectal cancer in India have been estimated to be 4.2 and 3.2/ 1, 00,000 for males and females respectively as compared to 60.8 and 42.3 respectively in the USA (1). In children less than 17 years of age mucin producing adenocarcinoma is the most predominant type & 48% of these are of signet ring cell type. These rare tumors show more malignant biological behaviour as compared to the ordinary colorectal carcinomas and carry an extremely poor prognosis. (2)We report a rare case of signet ring cell variant of mucinous adenocarcinoma of the colon in a 14 years old boy presenting in emergency with acute intestinal obstruction. Surgical removal of the tumour as an emergency procedure with defunctioning right transverse colostomy and post operative chemotherapy has ensured survival of the patient without metastaticdisease even after six of the treatment. Despite the reported high malignant behavior and poor prognosis of such tumors (3), prompt of effective surgical & chemotherapeutic management may increase the chances of survival of such patients.

Case Report:

A 14 years old male, presented with generalized pain abdomen since 5 days bilious vomiting & constipation since 3 days, distension abdomen since 5 days. There was no history of such episodes in the past. There was no history of tuberculosis in the patient on in the family this investigation revealed following values.

Hb-9.8gm%	TLC- 15,050/mm3
BT- 1’ 15”	DLC – 88/10/1/1
CT-3’05”	S. Creatinine - 1.2 mg%
B. urea -44mg%	K+ -5.0 mg%

Plain X-ray abdomen shows multiple air fluid levels, while ultrasound of the abdomen showed free fluid in Morrison’spouch, sub hepatic regions, left paracolic gutter.

The patient was adequately resuscitated and was taken up for emergency laparotomy under general anesthesia. An annular stenosing growth encompassing the whole circumference of descending colon was detected. There was about 500 ml of serous fluid present in the peritoneal cavity and was sent for cytological examination peritoneal fluid cytology report revealsHemorrhagicexudative effusion, withmild lymphocytosis and mesothelial cell proliferation with no evidence of T.B. or

malignancy. There were no metaslatic deposits in the peritoneal cavity on the surfaces of solid viscous of the abdomen. There was no lymphadenopathy. An emergency resection &anastomosis of 8 cm of descending colon including growth with defunctioning transverse colostomy was done. The patienthad an uneventful postoperative period and was on oral feeding by the 7th post operative period. The histopathological report of the resected specimen showed signet ring cells & mucin adenocarcinoma. The malignant neoplastic tissue is infiltrating up to serosal layer. The patient was put on 5 fluorouracil 750mg daily for five days as a loading dose followed by weekly injection of 5fu.

The patient has been followed up to for 6 months. The general physical condition of the patient has remained stable, has appetite has improved. A remain ultrasound showed no metastaticdisease.

Discussion:

Colorectal malignancies are extremely rare inpediatric age group,the youngest recoded case is a 9 month old child. The reported incidenceis 1.3 million children4. In general, these malignancies in children have a very poor prognosisand are usually behind the scope of operative correction. The main reasons attributed for this are delay in the diagnosis, advanced stage of the disease at presentation and poor histological differentiation of the malignancy. The predominant histological type on children of adolescents is the poorly differentiated mucinous adenocarcinoma4. The prognosis with a mucinous case is very poor. The mucin absorbs water, swells and invades local tissues, thereby promoting spread of malignant cells. It also interferes with the immunerecognition of carcinoma cells due to muco-polysaccharides coating. This histopathology is known to be moreaggressive with predisposition to early metastasis.

The existence of predisposing conditions, which increase the risk of colorectal carcinoma developing, is well recognized. These include familialpolyposis coli, Gardner's syndrome, turcot's syndrome, peutz, Jegher's syndrome, Juvenile polyposis of clone and ulcerative colitis.

Signet ring cell variant of mucinous adenocarcinoma is seen equally distributed in the right & left colon. Most of them are ulcerated & involve full thickness of the bowel wall.The patient may present as a case of acute intestinal obstruction with no preceding signs & symptoms or rarely with a perforation. But more commonly thepatient presents with a long history of abdominal discomfort, diarrhea, constipation or a mass. Other complaints such as cachexia, weight loss, jaundice, ascitis, or amenorrhea, suggestive of metastatic disease are also commonly seen.

Depending on the mode of presentation, the patient needs to be investigated & because of the rarity of colorectal carcinoma in young, a high index ofsuspicion is required to asses the primary & the metastatic disease & to evaluate its operability & resectability. (5)

Nodal & metastatic disease can be present in 70% of patients at presentation. In signet ring cell carcinomas metastasis is more frequently to the peritoneal cavity, ovaries andlymph nodes. Most of these patients have advanced disease at presentation & only a few cases would be amenable to curative surgery. The surgery, curative or palliative should be followed by chemotherapy with levamisole, mitomycin-C, leucovorinand 5 fluorouracil combination of levamisole and 5fu is considered to be the best form of chemotherapy, even for advanced signet ring cell carcinoma. (6) These two agentsreducethe incidence of recurrence of carcinoma in a surgically cured patient and also debulk cancer that is surgically importable. The estimated reduction of overall death rate with use of these agents is 33% (7)

The overall prognosis of signet Ring cell carcinoma is extremely poor with most patients dying within the first year of diagnosis and peritoneal carcinomatosis. The 5 year survival rate is only 12%.

CONCLUSION:

The overall prognosis of the carcinoma of the colon & rectum in children will only improve with increased awareness leading to early diagnosis of the condition. A high level of suspicion coupled with a simple digital rectal examination followed by sigmoidoscopy and / or colonoscopy if required, can result in early diagnosis which well goes a long way in providing effective therapy. (8)

Resection even palliative is always preferable to the bypass, because it effectively relieves the obstruction & also decreases the tumour load. Surgery should be the first modality of treatment as the disease in children responds poorly to chemotherapy as well to radiotherapy. The value of

chemotherapy as a means of palliation has been controversial andresponses have been less than optimal. In rectal cancers, preoperative radiotherapy has been utilized extensively to convent unresectable lesions to resectable ones (9)

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