Childhood Colon Carcinoma: PET-CT Insights in a rare case

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ABSTRACT

Background: Colorectal carcinoma (CRC) in childhood is extremely rare, usually diagnosed at an advanced stage, and often has a poor prognosis. The disease in those younger than 20 years of age has an annual incidence of 1 case per million individuals worldwide.

Case report: A 14-year-old female child underwent a left transverse and left descending colectomy for infiltrating adenocarcinoma of the descending colon, proven by the histopathology of the resected sample. Before that, the patient presented with abdominal pain, more on the left, and a history of chronic constipation. She had no history of ulcerative colitis or a family history of carcinoma of the colon or colonic polyposis. The tumor was MMR protein proficient and microsatellite stable on immunohistochemistry (IHC). Her serum Ca-19.9 was markedly raised, and pre-operative colonoscopy was also compatible with colonic growth. After surgery, a whole-body ^{18}F-FDG PET-CT baseline scan and a post-chemotherapeutic follow-up scan after 3 months were done, revealing a good prognosis for the patient.

Conclusion: Though CRC in childhood is an aggressive neoplasm with a poor prognosis, the recent case showed a good prognosis due to early diagnosis, correct treatment, and follow-up. The case also highlighted the usefulness of the ^{18}F-FDG PET-CT scan. A PET-CT scan provides anatomic, morphologic, and metabolic information about the tumor and patterns of spread. These are very crucial for proper treatment planning, therapy response assessment, and follow-up of patients with colorectal malignancy.

Keywords: Colon carcinoma, colorectal carcinoma, ^{18}F-FDG PET-CT scan, childhood.

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INTRODUCTION

Colorectal carcinoma (CRC) is the third most frequently diagnosed malignancy and the second most common cause of malignancy-related deaths worldwide (1). It accounts for about 10% of all carcinomas globally (2, 3). Colon carcinoma mainly occurs due to genetic alterations in the cells lining of the colon. Other contributing factors include a sedentary lifestyle, obesity and a positive family history of colorectal malignancy or polyposis syndromes. The risk of the malignancy is also influenced by some environmental factors such as consumption of a diet high in red meat and fat, low intake of dietary fiber, diabetes mellitus, smoking and alcohol consumption (4).

In recent years, a rapid increase in the incidence and mortality of colorectal cancer has been observed in several developing countries. Besides the hereditary predisposition, most of the cases of CRC are sporadic and typically develop slowly over years (5).

CRC in childhood is extremely rare, usually diagnosed at an advanced stage and often has poor prognosis (6, 7). The disease in those younger than 20 years of age has an annual incidence of 1 case per million individuals worldwide (6, 8).

CRC has an immunohistochemical background like DNA stability, whether it is microsatellite instable (MSI) or microsatellite stable (MSS). MSI has been shown to play a significant prognostic role in adult CRC. However, corresponding data regarding the prognostic significance of MSI in pediatric CRC are limited (2). The status of MMR (mismatch repair) genes on IHC has shown also importance to select a therapy modality (13, 15).

Positron emission tomography (PET) scanning improves the detection of recurrent colorectal cancer, thereby aiding in prognosis assessment and influencing patient management. Whole body 18F FDG PET-CT scans in colorectal cancer are primarily used for staging, detection of distant metastases, evaluation of recurrence and monitoring of treatment response rather than for the initial diagnosis. By detecting metabolic activity using a glucose-based tracer, PET-CT is highly effective in identifying metastatic disease and recurrent lesions, often leading to changes in clinical management in more than half of the patients (5, 9, 10).

CASE REPORT

A 14-year-old girl underwent a left transverse and left descending colectomy for infiltrating adenocarcinoma (grade II) of the descending colon, proven by the histopathology of the resected sample. Before that, the patient presented with abdominal pain, more on the left, and a history of chronic constipation. She had no history of ulcerative colitis or a family history of carcinoma of the colon or colonic polyposis. The tumor was MMR protein proficient and microsatellite stable on immunohistochemistry (IHC). Her serum Ca-19.9 was markedly raised, and pre-operative colonoscopy was also compatible with colonic growth. After surgery, she performed a whole-body ^{18}F-FDG PET-CT scan for baseline evaluation and revealed no hypermetabolic lesion in the post-operated colectomy site nor any hypermetabolic abdomino-pelvic lymph nodes. However, a small hypermetabolic activity (17.1 x 22.6 mm), SUVmax: 2.67, was noted in the post-operated left distal transverse colectomy site, and diffuse uptake of FDG in the incisional site of the anterior abdominal wall on the left side suggested post-surgical sequelae; recommended further follow-up. Then the patient received 10 cycles of adjuvant chemotherapy. A follow-up scan after three months was done and showed no definite residual/recurrent mass or abnormal FGD uptake at the anastomotic site. A small, intensely FDG-avid cyst (18 x 15 mm), SUVmax: 7.9, was seen in the left ovary, suggesting a follicular cyst. No ascites or lymphadenopathy could be seen. In that scan, brown fat uptake was also seen in bilateral cervical, supraclavicular, axillary, mediastinal, and para-spinal regions.

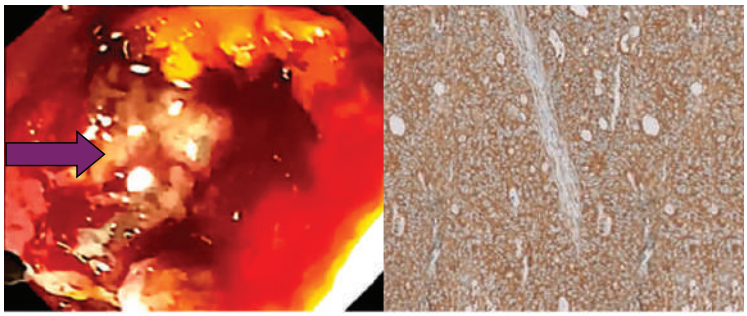

Figure 1: (A) pink arrow indicating colonoscopic evidence of circumferential ulceroproliferative growth within the descending colon causing complete luminal obstruction, (B) immunohistochemical picture of the resected sample of the carcinoma.

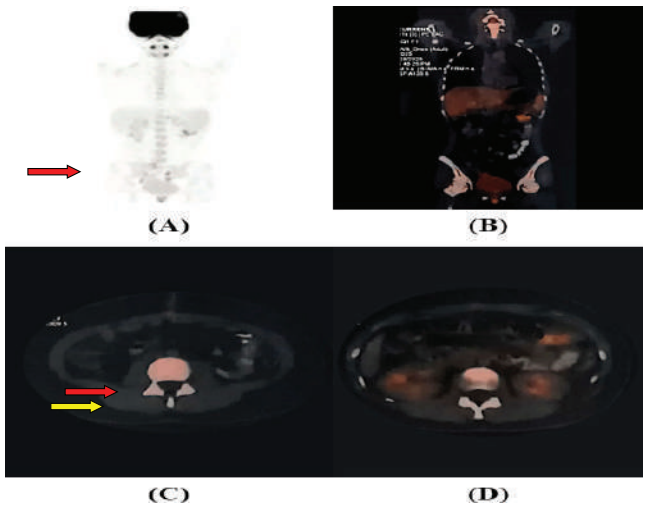

Figure 2: Post-surgical baseline 18F whole body PET-CT scan showing (A) MIP image, (C) axial CT image with evidence of suture at the anastomotic site, (B) coronal & (D) axial fusion images with red arrows revealing small hypermetabolic activity at the anastomotic site and yellow arrow indicating diffuse hypermetabolic activity in incisional area of anterior abdominal wall on left side, suggested post-surgical sequelae.

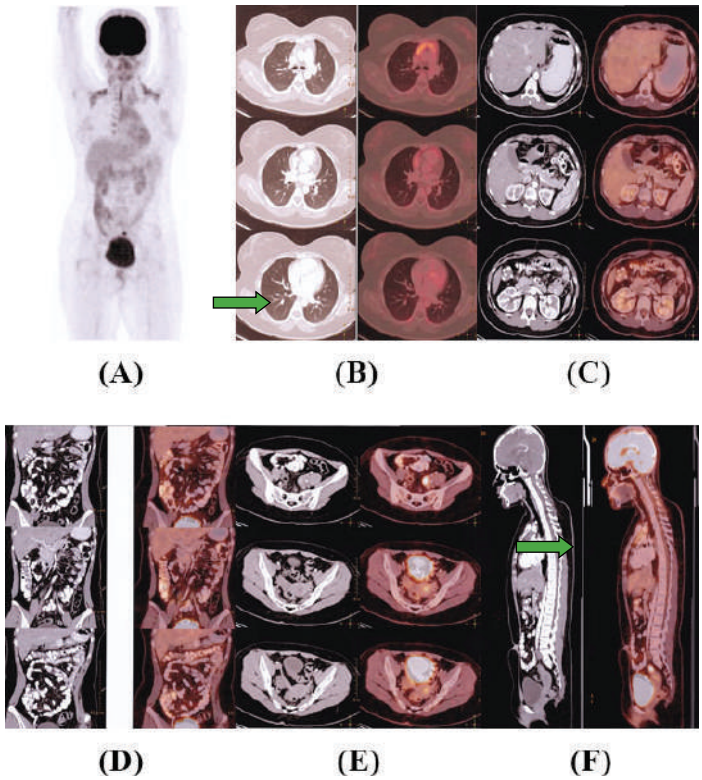

Figure 3: Follow-up 18F whole body PET-CT scan showing (A) MIP image, (B) axial CT & fusion images of both lungs reflecting clear lung fields, (C) axial CT & fusion images of abdominal organs with no definite abnormality, (D) coronal & (E) axial CT & fusion images of bowel loops with no definite abnormality or focal FDG uptake in the anastomotic site or anywhere rather, green arrow indicating a small hypermetabolic left ovarian follicular cyst, (F) left sagittal CT & fusion images with no definite spinal bony abnormality or abnormal FDG uptake.

DISCUSSION

Colorectal carcinoma (CRC) in the pediatric population is extremely rare. The present study reports a rare case of colon carcinoma in a child. Previous reports have also described small series of pediatric CRC, including cases in adolescents aged 12–16 years and others ranging from 11 to 14 years, highlighting the uncommon occurrence of this malignancy in younger age groups

The reported case presented with abdominal pain and a history of chronic constipation. In contrast, previously reported cases have described symptoms such as bloody stool, weight loss, and abdominal pain, while others have noted a broader range of presentations including abdominal pain, nausea, vomiting, diarrhea, constipation, and hematochezia (6, 7). CRC has well-recognized genetic predispositions. Hereditary Colorectal Cancer (HCRC) is broadly divided into two major categories with distinct clinical characteristics: Hereditary Nonpolyposis Colorectal Carcinoma (HNPCC) and Hereditary Polyposis Colorectal Carcinoma (HPCC). HNPCC accounts for approximately 3–5% of all CRC cases, with Lynch syndrome being the most common form. In contrast, HPCC syndromes constitute less than 1% of CRC cases and include adenomatous, hamartomatous, serrated, and mixed polyposis syndromes, classified according to the histological type of the polyps (12). On the other hand, the recent case had no family history of CRC or polyposis syndrome.

In the present case, microsatellite stability (MSS) was demonstrated on immunohistochemistry. In contrast, Wild H. et al. reported that microsatellite instability (MSI) in colorectal carcinoma is associated with improved survival and a lower incidence of distant metastases, indicating that MSI is linked with a more favorable prognosis (2).

Presence of an MMR-proficient gene on IHC is associated with a favorable prognosis when treated with adjuvant chemotherapy. Defective DNA mismatch repair (MMR) is found in approximately 15% of sporadic colorectal carcinomas. Several retrospective studies have shown that patients with MMR-deficient tumors tend to have a more favorable stage-adjusted prognosis compared to those with MMR-proficient disease. In addition, evidence indicates that MMR-deficient colorectal cancers derive limited benefit from adjuvant chemotherapy, whereas chemotherapy remains an important treatment modality for MMR-proficient tumors.

On the other hand, immune checkpoint inhibitors have significantly changed the management of MMR-deficient or microsatellite instability–high (MSI-H) colorectal cancers, demonstrating high response rates and durable clinical benefit. However, these agents have shown limited efficacy in MMR-proficient or microsatellite stable (MSS) tumors. Therefore, the MMR status and microsatellite stability of the tumor play a crucial role in guiding therapeutic decisions in colorectal carcinoma (13, 15).

In the present case, whole-body PET-CT revealed no evidence of residual or recurrent primary tumor, regional lymph node involvement, or distant metastasis, indicating a favorable prognosis. In contrast, Poles GC. et al. in a study of 918 colorectal carcinoma patients, reported that pediatric cases tend to demonstrate a more aggressive clinical course with poorer overall survival, particularly in rectal carcinoma. Similarly, Kim G. et al. observed disease progression in all four pediatric patients with colorectal carcinoma despite undergoing radical surgery followed by systemic chemotherapy.

Early detection, appropriate stage stratification, a multidisciplinary treatment approach, and prospective clinical trials are important for improving prognosis and outcomes in pediatric colorectal carcinoma patients (14).

CONCLUSION

Colorectal cancer (CRC) in childhood, though rare and aggressive with generally poor prognosis, can have a favorable outcome when detected early and treated appropriately, as demonstrated in a recent case. This case exhibited microsatellite stability and proficient mismatch repair proteins, contributing to its positive prognosis. The utility of whole body 18F FDG PET-CT scans in this context is underscored, particularly for staging and assessing treatment response rather than initial diagnosis. The article emphasizes the critical role of PET/CT in treatment planning and monitoring for CRC in pediatric patients, addressing challenges such as diagnostic dilemmas and late presentations that often lead to poor outcomes.

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