

# A rare cause of intestinal obstruction in children: signet-ring cell adenocarcinoma of the colon

Basak Erginel, M.D.,<sup>1</sup> Naila Mustafayeva, M.D.,<sup>1</sup> Çetin Ali Karadağ, M.D.,<sup>2</sup> Fatih Yanar, M.D.,<sup>3</sup> Rejin Kebudi, M.D.,<sup>4</sup> Hikmet Gülşah Tanyıldız, M.D.,<sup>5</sup> Deniz Tuğcu, M.D.,<sup>5</sup> Neslihan Berker, M.D.,<sup>6</sup> Burak İlhan, M.D.,<sup>3</sup> Feryal Gün Soysal, M.D.<sup>1</sup>

<sup>1</sup>Department of Pediatric Surgery, Istanbul University, Istanbul Medical Faculty, İstanbul-Türkiye

<sup>2</sup>Department of Pediatric Surgery, Sisli Hamidiye Etfal Training and Research Hospital, İstanbul-Türkiye

<sup>3</sup>Department of General Surgery, Istanbul University, Istanbul Medical Faculty, İstanbul-Türkiye

<sup>4</sup>Department of Pediatric Hematology-Oncology, Istanbul University, Oncology Institute, İstanbul-Türkiye

<sup>5</sup>Department of Pediatric Hematology-Oncology, Istanbul University, İstanbul-Türkiye

<sup>6</sup>Department of Pathology, Istanbul University, İstanbul Medical Faculty, İstanbul-Türkiye

## ABSTRACT

**BACKGROUND:** Signet-ring cell adenocarcinoma of the colon is well-recognized in adult patients who are extremely rare and not well-documented in children. Our study aims to raise awareness about this rare disease and its long-term outcomes.

**METHODS:** We retrospectively evaluated patients with signet-ring cell colon adenocarcinoma.

**RESULTS:** Six patients, three boys and three girls, with a mean age of 14.83 (range, 13–17 years), presented with signs of intestinal obstruction and were diagnosed with signet-ring cell colon adenocarcinoma. All patients had air-fluid levels on abdominal X-ray. Abdominal ultrasonography of all patients revealed subileus. Abdominal computed tomography was performed in five patients, and pre-operative colonoscopy was conducted in two patients before the emergency intervention. All of the patients underwent emergent exploratory laparotomy with the preliminary diagnosis of acute abdomen. In two patients, debulking surgery followed by a stoma was performed. The remaining four patients were treated with anastomosis following intestinal resection. All girls had metastases on the ovary. One of the patients died due to the burden of multiple metastases in the early period, and three died in the sixth post-operative year. We have been following the remaining two patients since then.

**CONCLUSION:** Although signet-ring cell carcinomas (SRCCs) are rare, they should be considered in the differential diagnosis of acute abdomen and intestinal obstruction in pediatric patients. Despite early diagnosis and treatment, SRCC has a poor prognosis in the pediatric population.

**Keywords:** Acute abdomen; pediatric surgery; signet-ring cell adenocarcinoma.

## INTRODUCTION

In contrast to its prevalence in the adult population, colorectal carcinoma (CRC) is a rare pathology among pediatric patients and accounts for almost 1% of all pediatric malignancies.<sup>[1,2]</sup> It usually presents with simple, non-specific gastrointestinal symptoms that mimic benign diseases.<sup>[3]</sup> Due to

its rarity, it is mostly diagnosed in advanced stages and has a worse prognosis in children than in adults.

The incidence of CRC in children is <0.1 cases per million. Signet-ring cell carcinoma (SRCC) is a rare subtype that accounts for 0.7% of all CRC cases.<sup>[4]</sup> It is less well-documented in children than in adults. SRCC of the colon is quite aggressive and has a very high malignancy potential; it is usually di-

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Address for correspondence: Basak Erginel, M.D.

Istanbul University, Istanbul Medical Faculty, İstanbul, Türkiye

E-mail: basakerginel@hotmail.com

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agnosed at advanced stages and has a poor prognosis.<sup>[5]</sup> It has been reported that the prognosis is worse in children.<sup>[6]</sup> Although the gender distribution is equal in adults, it is twice as common among male patients in the pediatric age group.<sup>[7]</sup>

The limited availability of the literature on this condition, which is due to the restricted number of cases, complicates the management of signet-ring cell adenocarcinoma in children. Therefore, this study was conducted to retrospectively evaluate pediatric patients diagnosed with SRCC who underwent surgery for acute abdomen; this investigation was aimed to increase the awareness of SRCC in the differential diagnosis of intestinal obstruction in children and to evaluate the long-term outcomes of this rare pathology.

## MATERIALS AND METHODS

Six children who presented with signs and symptoms of acute abdomen who underwent emergency surgery between January 2006 and October 2022 and whose pathological report was signet-ring cell adenocarcinoma were evaluated retrospectively. Informed consent was obtained from their parents. The study was approved by the Ethics Committee (decision year/no: 2023/661). The patients were assessed regarding age, sex, clinical findings at diagnosis, diagnostic methods, operation, histological type, and outcome.

Six patients with a histopathological diagnosis of signet-ring cell colon adenocarcinoma were reviewed retrospectively. Three of the patients were boys, and three were girls; the mean age of the patients was 14.83 (range, 13–17) years. One of the patients had ataxia-telangiectasia. All patients presented with signs and symptoms of acute abdomen. On physical examination, all patients were found to have abdominal distension and tenderness. Three had a palpable mass on the left side of the umbilicus on palpation. In addition to conducting routine blood tests, tumor markers were preoperatively checked in two patients, and their CA and CEA-125 values were high. Air-fluid levels were determined in the abdominal X-rays of all patients. Abdominal ultrasonography of all the patients revealed subileus. Abdominal computed tomography (CT) was performed on five patients before the emergency operation, and pre-operative colonoscopy was performed on two patients. In two patients, debulking surgery was performed, and a stoma was matured due to intestinal obstruction. In the remaining four patients, resection anastomosis was performed with safe surgical margins. Metastasis was detected in the biopsies of the ovaries of all three girls. One of the patients died due to multiple metastases in the early period, and three died in their 6th year of treatment. Two patients remained alive and were followed up for 10 years. The 5-year survival rate of the patients was 83.3% (SE: 15.2%). A post-operative chemotherapy FOLFOX regimen, including oxaliplatin in combination with 5-fluorouracil and leucovorin, was administered to all patients by the post-operative pediatric hematology and oncology unit. Subsequently, the treatment was changed to the FOLFIRI regimen combined

with anti-VEGF in the patients who developed metastatic disease during follow-up. Pathologically, microsatellite instability (MSI), a hypermutable phenotype caused by the loss of DNA mismatch repair (MMR) activity, could only be examined in two of the recently diagnosed patients by immunohistochemistry (third and fourth patients). No loss of nuclear MLH1, MSH2, MSH6, and PMS2 protein expression in tumor cells indicated the lack of MSI. Table 1 summarizes our patients, and the detailed information about the patients is as follows:

### Case 1

A 15-year-old male patient with no known disease was admitted to our clinic with complaints of abdominal pain and vomiting occurring for 1 month. Physical examination showed that the patient had diffuse abdominal tenderness. His acute phase reactants were high, and diffuse air-fluid levels were seen on abdominal X-ray. The patient underwent diagnostic laparotomy under emergency conditions and had multiple masses in his abdomen, with the largest in the descending colon; a stoma was opened with the diagnosis of intestinal obstruction. Histopathological evaluation revealed signet-ring cell colon adenocarcinoma. The patient was immediately started on FOLFOX therapy, but due to the development of metastatic disease during follow-up, the treatment was changed to FOLFIRI combined with anti-VEGF. The patient died due to brain metastasis in the 6th month after diagnosis during the pediatric hematology and oncology unit's follow-up.

### Case 2

A 15-year-old male patient was referred to us after a mass lesion in his splenic flexure was observed during a colonoscopy procedure following complaints of weight loss, abdominal pain, and bloody stools. On admission, the patient's abdomen was distended, and a mass lesion was palpated in the left lateral umbilicus. The patient had high CEA and CA19-9 levels in his laboratory reports. In the abdominal CT interpretation, an irregular 58 mm wall-thickness increase was seen in a segment of the descending colon. The patient underwent descending colon resection and colocolonic anastomosis. The pathology report revealed signet-ring cell adenocarcinoma. The patient, whose post-operative follow-up was performed by the pediatric hematology and oncology unit due to progressive disease related to multiple recurrences, died in the 6th post-operative year despite receiving FOLFIRI treatment.

### Case 3

A 15-year-old female patient with ataxia-telangiectasia was admitted with intermittent abdominal pain and vomiting occurring for 1 month. She had tenderness in her abdomen. Her AFP level was found to be high, and air-fluid levels were present in the right lower quadrant of the abdomen X-ray (Fig. 1). Her intestinal loops were observed by abdominal ultrasonography to be widely dilated. CT revealed a mass lesion of approximately 4 cm obstructing the lumen in the ascend-

**Table 1.** Summary of patients with signet-ring cell carcinoma

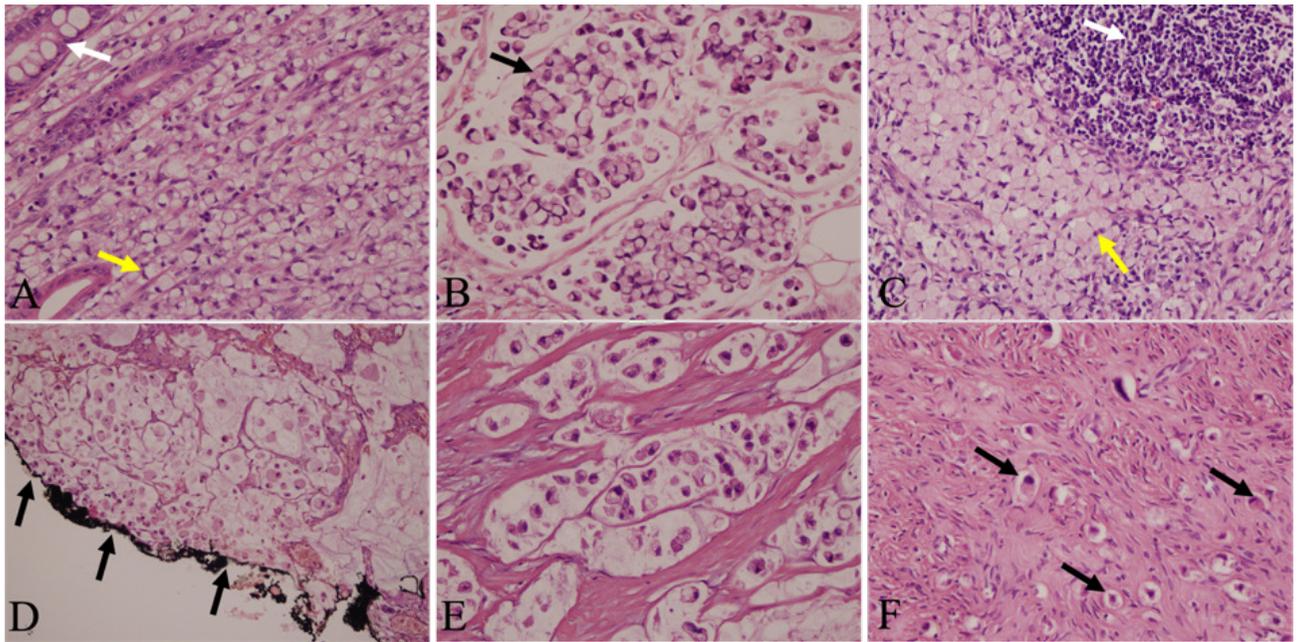
Patient no.	Age, sex	Symptoms	Examination	Imaging	Localization	Surgery	Outcome
1	15, M	Abdominal pain, vomiting	Abdominal distention, tenderness, decreased bowel sounds	X-ray (air-fluid levels) USG	Descending colon (left)	Stoma	Exitus, 6 <sup>th</sup> month
2	15, M	Abdominal pain, bloody stools, weight loss	Abdominal distention, tenderness, decreased bowel sounds	X-ray (air-fluid levels) USG Colonoscopy CT	Descending colon (left)	Descending colonic resection, colocolonic anastomosis	Exitus, 6 <sup>th</sup> year
3	15, F	Abdominal pain, vomiting	Abdominal distention, tenderness, decreased bowel sounds	X-ray (air-fluid levels), USG CT	Ascending colon (right), ovarian metastasis	Colonic resection, colocolonic anastomosis	Exitus, 6 <sup>th</sup> year
4	17, F	Abdominal pain, vomiting	Abdominal distention, tenderness, decreased bowel sounds	X-ray (air-fluid levels), USG CT	Descending colon (left), ovarian metastasis	Colonic resection, colocolonic anastomosis, oophorectomy	Alive, (3 years)
5	13, M	Abdominal pain, vomiting, nausea,	Abdominal distention, tenderness, decreased bowel sounds, rectal examination—mass.	X-ray (air-fluid levels), USG CT	The sigmoid colon (left) omental, peritoneal metastasis	Sigmoid resection, end-to-end anastomosis	Exitus, 6 <sup>th</sup> year
6	14, F	Abdominal pain	Left lower abdominal tenderness Abdominal distention Colonoscopy	X-ray (air-fluid levels), USG CT—misdiagnosis as ovarian torsion	Sigmoid (left) ovarian metastasis	Oophorectomy sigmoid resection stoma	Alive

**Figure 1.** Air-fluid levels seen in the right lower quadrant of the abdomen X-ray in Case 3.

ing colon. Since the patient presented ileus, the necessary consultations with pediatric hematological oncology were urgently conducted, and then an emergency operation involving colonic resection and colocolic anastomosis was performed by creating a clean surgical margin. The patient's histopathological report revealed signet-ring cell adenocarcinoma (Fig. 2). Immunohistochemical examination (MSH1, MSH2, MSH6, and PMS2) for MSI and PDL-1 immunohistochemistry were performed in the tumor tissue. No loss of nuclear expression was observed with MLH1, MSH2, MSH6, and PMS2 in the tumor cells, indicating the absence of MSI. PDL-1 was negative in the tumor cells. The pediatric hematology unit started an appropriate dose-modified FOLFOX chemotherapy regimen to prevent drug instability in the patient. In the follow-up, an MRI revealed a mass in her left ovary (Fig. 3). and she was operated on again. The pathology report of this patient, who underwent left salpingo-oophorectomy, was found to be compatible with metastasis. Unfortunately, the patient died due to progressive disease after multiple recurrences in the 6th year of follow-up.

#### Case 4

A 17-year-old female patient with no known disease was evaluated due to complaints of abdominal pain and vomiting that had continued for 4–5 days. It was reported that the patient had diffuse air-fluid levels in the abdominal X-ray, and an image consistent with a mass lesion in the transverse and descending colon was seen in the CT (Fig. 4). After a consul-



**Figure 2.** Histopathologic images of the tumors in Case 3 (a-c) and Case 4 (d-f). (a) Signet-ring cells (yellow arrow) adjacent to the crypts (white arrow) of the colon mucosa. (b) Signet-ring cells (arrow) dispersed in the mucinous stroma. (c) Lymph node metastasis of the tumor. Signet-ring cells (yellow arrow) adjacent the lymphocytes (white arrow) of the lymph node parenchyma. (d) Peritoneal surface invasion of the tumor. Signet-ring cells adjacent to the black ink (arrows). (e) Signet-ring cells dispersed in the mucinous stroma. (f) Ovarian metastasis. Signet-ring cells (arrows) scattered within the ovarian stroma.

tation with the pediatric hematology oncology department, the patient underwent an emergency operation. During the exploration, a colonic mass of approximately 10 cm in length was found; it started at the level of the splenic flexure and involved the descending colon, completely occluded the lumen, and protruded beyond the lumen (Fig. 5). First, resection anastomosis was performed to create a safe border. Next, we

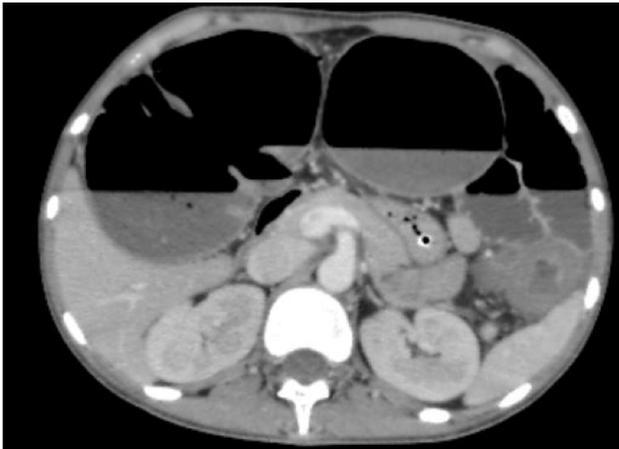
explored the abdomen, and the mass was excised from the right paraovarian region with a suspicious lesion. The patient had no problems during the post-operative follow-up, and her pathology report was interpreted as signet-ring cell mucinous colon adenocarcinoma with ovarian metastasis. The pediatric hematology unit followed up with the patient, and an MRI showed that the mass lesion in her right ovary had progressed. Right salpingo-oophorectomy was performed, and metastasis was found in the pathology report. The patient, then with a diagnosis of metastatic colon cancer, was transferred to the oncology department for the continuation of treatment. Chemotherapy was arranged for the patient, and immunohistochemical examination (MSH1, MSH2, MSH6, and PMS2) for MSI and PDL-1 immunohistochemistry were performed in the tumor tissue. No loss of nuclear expression was observed with MLH1, MSH2, MSH6, and PMS2 in the tumor cells, indicating the absence of MSI. PDL-1 was negative in the tumor cells. A post-operative chemotherapy FOLFOX regimen, including oxaliplatin in combination with 5-fluorouracil and leucovorin, was applied, and this treatment was effective and tolerable in the patient. The patient has been under follow-up for 3 years.



**Figure 3.** MRI showing a mass in the left ovary in Case 3.

### Case 5

A 13-year-old male patient presented with abdominal pain, nausea, and bilious vomiting. He had a palpable mass in the left lower quadrant that was within finger distance on the rectal examination. The patient's bowel sounds were metallic. A mass extended from the beginning of the rectosigmoid



**Figure 4.** A CT image consistent with a mass lesion in the transverse and descending colon in Case 4.

colon to the descending colon in the CT. Laparotomy, sigmoid resection, and end-to-end anastomosis were performed. Histopathological evaluation interpreted the lesion as a signet-ring cell adenocarcinoma with omental and peritoneal metastases. The patient, whose treatment was continued by the post-operative pediatric hematology and oncology unit, died in the 6th year of follow-up due to progressive disease. As initial treatment with FOLFOX and FOLFIRI was started on the onset of progressive disease, but unfortunately, we lost the patient while he was undergoing chemotherapy.

### Case 6

A 14-year-old female patient was admitted with left abdominal pain. CT revealed an 88 × 54 mm mass that was preoperatively misdiagnosed as ovarian torsion. On laparoscopic exploration, many millimetric lesions were found to be scattered on the left ovary and pelvic peritoneum. Due



**Figure 5.** A colonic mass of approximately 10 cm in length that started at the level of the splenic flexure and involved the descending colon, completely occluding the lumen and protruding beyond the lumen, in Case 4.

to the abnormal appearance, the surgery was converted to laparotomy. No adnexal torsion was found. Due to the tumor-like appearance of the lesion, a left oophorectomy was performed, and a biopsy was taken from adherent organs. The pathology specimens were reported to show signet-ring cell adenocarcinoma. A colonoscopy revealed a tumoral mass that filled the lumen. The biopsies from the colonic group also revealed signet-ring cell adenocarcinoma. Therefore, sigmoid resection and colostomy were performed. Systemic chemotherapy FOLFOX was administered according to the stage. The patient continues to be followed up and treated 3 years after diagnosis.

## DISCUSSION

CRC is rare in children and occurs at fewer than 0.1 cases per million.<sup>[8]</sup> Signet-ring cell adenocarcinoma is a rare subtype of CRC. It is sporadic in pediatric patients, and the exact number of such cases is not available in the literature. Signet-ring cell adenocarcinoma is a poorly differentiated subtype that behaves more aggressively than classical adenocarcinoma.<sup>[9]</sup> Late detection of this tumor, which already has a poor prognosis, shortens the patient's survival time. Considering that this tumor is seen in adolescence, the importance of increasing patient survival is evident. The present study was conducted to shed light on this rare and insidious disease by retrospectively evaluating patients with signet-ring cell adenocarcinoma. The aim is to increase awareness among pediatricians and surgeons of the different presentations of the condition.

CRC can be seen in children of any age; the youngest patient described in the literature was 9 months old.<sup>[10]</sup> Most cases reported in the literature were observed in late childhood or adolescence. The age range of the patients in the present study was 13–17 years, and the mean age was 15 years, which is entirely consistent with the literature.<sup>[11]</sup>

Although the gender distribution of CRC is equal among adults, the disease is twice as common among male patients in the pediatric age group. However, our study group comprised three boys and three girls equally. While CRC is seen early in adult patients with familial polyposis coli, ulcerative colitis, or colorectal cancer, it can develop sporadically in pediatric patients.<sup>[1]</sup> All cases in this study were sporadic.

The main complaint of all six patients was abdominal pain, which was consistent with the literature. Other complaints, such as vomiting, inability to pass gas, and nausea, were related to intestinal obstruction. In addition, in the literature, symptoms such as disturbances in bowel habits, weight loss, and anemia have been reported.<sup>[3]</sup> Unfortunately, these complaints are widespread and can occur in many benign diseases, resulting in a delay in prognosis.

More than 96% of SRCCs arise from the stomach, and colonic SRCC is rare. Primary SRCCs of the colon were first described by Laufman and Saphir.<sup>[12]</sup> However, only a limited number of cases involving children have been published thus

far. All pediatric patients detected in the past 20 years are presented in Table 2.<sup>[4,5,13-19]</sup> Even the most extensive series consisted of only four cases.<sup>[16]</sup> In 2017, Li et al. provided a case report and reviewed all the literature in this area.<sup>[17]</sup> Thibodeau et al. reported that a 15-year-old boy was admitted with abdominal pain and diagnosed with ascending and successfully treated colon SRCC with colectomy.<sup>[18]</sup> The patient had been alive for 6 months. Recently, Mahajan et al. published a report on seven cases of CRC, two of which were signet-ring cell adenocarcinoma and were located in the right and left colon, respectively; both of these patients died.<sup>[19]</sup>

MSI is a hypermutable phenotype caused by the loss of function of the DNA MMR system. This repair system is mainly composed of four proteins (MLH1, MSH2, MSH6, and PMS2) interacting together to detect mismatches and correct them during DNA replication.<sup>[20]</sup> MSI is found in approximately 15% of all colorectal cancers; 3% of these are associated with Lynch syndrome, which is related to germline mutations in one of the MMR genes (usually MLH1 or MSH2), and the remaining 12% are sporadic and usually related to epigenetic inactivation of the MLH1 gene. MSI is detected by PCR amplification of specific microsatellite markers or by immunohistochemical loss of expression of one of the abovementioned proteins.<sup>[21]</sup> To follow the family members of patients with Lynch syndrome before cancer arises and to choose the right treatment for sporadic tumors, it is important to recognize MSI. MLH1, MSH2, MSH6, and PMS2 protein expression could only be examined in two of our recently diagnosed patients by immunohistochemistry, and no loss of nuclear expression was found in tumor cells, indicating the absence of MSI.

The rarity of SRCC in pediatric patients poses challenges in diagnosis, treatment, and management. The present study retrospectively evaluated six pediatric patients diagnosed with SRCC who underwent surgery for acute abdomen. The aim was to increase awareness of SRCC in the differential diagnosis of signs of intestinal obstruction in children and to evaluate the long-term outcomes of this rare pathology. Early diagnosis of SRCC in pediatric patients is challenging due to its rarity and non-specific symptoms that mimic benign diseases. Abdominal pain, vomiting, and signs of intestinal obstruction were the main presenting complaints in the studied patients, which are consistent with previous reports. However, these symptoms are common and can be attributed to various benign conditions, leading to delays in diagnosis and treatment. Therefore, it is crucial for pediatricians and surgeons to maintain a high index of suspicion for SRCC in children presenting with gastrointestinal symptoms, especially when there is no improvement with conservative management or when alarm features are present.

Imaging modalities such as abdominal X-rays, abdominal ultrasound, and CT play an essential role in the diagnostic workup of SRCC. In the present study, all patients underwent abdominal X-rays, which showed air-fluid levels suggestive of intestinal obstruction. Abdominal ultrasound revealed subileus in all

**Table 2.** Pediatric cases with signet-cell carcinoma of the colon reported in the last 20 years

Author	Year	Age/sex	Symptom	Site	Surgery	Survival
Marone et al. <sup>[4]</sup>	2012	17/M	Right abdominal pain swelling	Ascending colon	Right colonic resection	Exitus
Singh et al. <sup>[5]</sup>	2014	10/M	Abdominal distension vomiting	Distal sigmoid colon	Exitus	Exitus
Agrawal et al. <sup>[13]</sup>	2021	11/M		Sigmoid colectomy peritoneal seeding		
Yang et al. <sup>[14]</sup>	2015	19/M	Abdominal pain vomiting	Transverse colon proximal to the splenic flexure	Stoma	Exitus
Chattopadhyay et al. <sup>[15]</sup>	2012	10/M	Abdominal pain	Descending colon	Left hemicolectomy	Alive for eight months
Pandey et al. <sup>[16]</sup>	2008	10/M	Abdominal pain rectal bleeding in all patients	Rectosigmoid	Resection/anastomosis	Alive after 11 months
Li et al. <sup>[17]</sup>	2017	11/M				
Thibodeau et al. <sup>[18]</sup>	2021	11/M	Abdominal pain melena	Mid-transverse colon	Resection-anastomosis	Alive after 6 months
Mahajan et al. <sup>[19]</sup>	2022	15/M	Abdominal pain	Ascending colon	Right hemicolectomy	Exitus
		9/M	Intestinal obstruction	Left	Laparotomy, biopsy	Exitus
		9/M	Abdominal pain fever	Right	re-exploration, ileostomy, biopsy	Exitus

patients, supporting the diagnosis. CT scans were performed in most cases and provided detailed information about the location, size, and extent of the tumor. These imaging techniques help guide surgical intervention and assess the presence of metastases, as observed in the ovarian metastases detected in the three female patients. Colonoscopy was performed in two patients and aided in visualizing the tumor and obtaining biopsy samples.

Surgical intervention is the primary treatment modality for SRCC in pediatric patients. The type of surgery depends on the extent of the disease, tumor location, and presence of metastases. In the presented cases, three patients underwent resection and anastomosis with safe surgical margins, while two patients required debulking surgery and colostomy due to intestinal obstruction. The choice of surgical approach should aim for complete resection of the tumor and regional lymph nodes whenever feasible. The involvement of a multidisciplinary team, including pediatric surgeons, pediatric oncologists, and pathologists, is crucial to ensure proper management and treatment planning.

Histopathological examination remains the gold standard for confirming the diagnosis of SRCC. In the present study, all patients had histopathological reports confirming signet-ring cell adenocarcinoma. Notably, immunohistochemical examination was performed in two recently diagnosed patients to assess MSI. The absence of loss of nuclear expression in MLH1, MSH2, MSH6, and PMS2 indicated the absence of MSI. This information is relevant, as MSI can guide treatment decisions, particularly in the context of potential immunotherapy options.

The prognosis of SRCC in pediatric patients is generally poor due to the late diagnosis, advanced stage at presentation, and aggressive nature of the tumor. In the present study, the 5-year survival rate was 83.3%, but the follow-up period varied among patients. Three patients died during the 6th year of treatment due to progressive disease and multiple recurrences. Two patients were alive at the last follow-up, with a follow-up period of 10 years. The importance of long-term follow-up and close monitoring cannot be overstated, as the risk of disease recurrence and metastasis persists.

The lessons learned from these cases include the need for increased awareness among health-care providers about the possibility of SRCC in pediatric patients presenting with gastrointestinal symptoms. Prompt recognition and referral for further investigation, including imaging and histopathological examinations, are crucial.

The limitation of the present study is the low number of patients due to the rarity of the disease. However, to the best of our knowledge, this is the most extensive series of SRCCs of the colon in children.

## CONCLUSION

Awareness and early intervention remain the main challenges

affecting the early diagnosis and prognosis of childhood signet-ring cell colorectal adenocarcinoma. Colon cancer should be considered in the differential diagnosis of acute abdomen and obstruction findings in patients. Therefore, more emphasis should be placed on colonoscopy at this stage. In the surgical management of signet-ring cell adenocarcinomas, if possible, surgical resection with a safe margin is preferred, but debulking surgery or only a biopsy or stoma can be performed in necessary cases. Despite advanced chemotherapy and surgical treatment methods, these tumors have very poor outcomes.

**Ethics Committee Approval:** This study was approved by the Istanbul University, Istanbul Medical Faculty Clinical Research Ethics Committee (Date: 14/04/2023, Decision No: 1725098).

**Peer-review:** Externally peer-reviewed.

**Authorship Contributions:** Concept: B.E.; Design: B.E.; Supervision: B.E.; Materials: B.E., N.H., Ç.A.K., F.Y., R.K., C.T., D.T., W.B., B.I.; Data: B.E., N.M., Ç.A.K., R.K.; Analysis: B.E.; Literature search: B.E., F.Y., B.I.; Writing: B.E.; Critical revision: B.E., F.G.S.

**Conflict of Interest:** None declared.

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## ORİJİNAL ÇALIŞMA - ÖZ

### Çocuklarda nadir bir bağırsak tıkanıklığı nedeni: Kolon taşlı yüzük hücreli adenokarsinom

Dr. Başak Erginel,<sup>1</sup> Dr. Naila Mustafayeva,<sup>1</sup> Dr. Çetin Ali Karadağ,<sup>2</sup> Dr. Fatih Yanar,<sup>3</sup> Dr. Rejin Kebudi,<sup>4</sup> Dr. Hikmet Gülşah Tanyıldız,<sup>5</sup> Dr. Deniz Tuğcu,<sup>5</sup> Dr. Neslihan Berker,<sup>6</sup> Dr. Burak İlhan,<sup>3</sup> Dr. Feryal Gün Soysal<sup>1</sup>

<sup>1</sup>Istanbul Üniversitesi, İstanbul Tıp Fakültesi, Çocuk Cerrahisi Anabilim Dalı, İstanbul, Türkiye

<sup>2</sup>Şişli Hamidiye Etfal Eğitim ve Araştırma Hastanesi, Çocuk Cerrahisi Anabilim Dalı, İstanbul, Türkiye

<sup>3</sup>Istanbul Üniversitesi, İstanbul Tıp Fakültesi, Genel Cerrahi Anabilim Dalı, İstanbul, Türkiye

<sup>4</sup>Istanbul Üniversitesi, İstanbul Tıp Fakültesi Onkoloji Enstitüsü, Pediatrik Hematoloji-Onkoloji Bölümü, İstanbul, Türkiye

<sup>5</sup>Istanbul Üniversitesi, İstanbul Tıp Fakültesi, Pediatrik Hematoloji-Onkoloji Anabilim Dalı, İstanbul, Türkiye

<sup>6</sup>Istanbul Üniversitesi, İstanbul Tıp Fakültesi, Patoloji Anabilim Dalı, İstanbul, Türkiye

**AMAÇ:** Kolonun taşlı yüzük hücreli adenokarsinomu yetişkin hastalarda iyi tanınmaktadır, ancak son derece nadirdir ve çocuklarda o kadar iyi belgelenmemiştir. Çalışmamız bu nadir hastalık ve uzun vadeli sonuçları hakkında farkındalık yaratmayı amaçlamaktadır.

**GEREÇ VE YÖNTEM:** Taşlı yüzük hücreli kolon adenokarsinomlu hastalarımızı retrospektif olarak inceledik.

**BULGULAR:** Barsak obstrüksiyonu bulguları ile başvuran ve taşlı yüzük hücreli kolon adenokarsinomu tanısı alan ortalama yaşları 14.83 (dağılım 13-17) olan üç erkek ve üç kız olmak üzere altı hastamız vardı. Tüm hastaların karın grafisinde hava-sıvı seviyeleri mevcuttu. Abdominal ultrasonografide tüm hastaların subileus saptandı. Acil girişim öncesinde beş hastaya karın bilgisayarlı tomografisi, iki hastaya preoperatif kolonoskopi yapıldı. Hastaların tamamına akut karın ön tanısı ile acil eksploratuvar laparotomi uygulandı. İki hastada kitle küçültme ameliyatı ardından stoma açıldı. Kalan dört hasta intestinal rezeksiyon sonrası anastomoz ile tedavi edildi. Tüm kızların yumurtalıklarında metastaz vardı. Hastalardan biri erken dönemde multipl metastaz yükü nedeniyle, üçü ise postoperatif altıncı yılda kaybedildi. O zamandan beri kalan iki hastayı takip etmekteyiz.

**TARTIŞMA:** Taşlı yüzük hücreli karsinomlar nadir görülmekle birlikte akut karın ve barsak obstrüksiyonu olan pediatrik hastaların ayıncı tanısında akılda tutulmalıdır. Erken tanı ve tedaviye rağmen pediatrik popülasyonda prognozu kötüdür.

**Anahtar sözcükler:** Akut karın; çocuk cerrahisi; taşlı yüzük hücreli karsinom.

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