Case Report

Malignant Peripheral Nerve Sheath Tumor of the Colon in a Teenager: A Case Report and Review of the Literatures

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Key Words
Teenager;
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Purpose. Malignant peripheral nerve sheath tumors (MPNSTs) are uncommon in the colorectal tract, and only five such cases have been reported. We present a case of colon MPNST in a teenager who was treated with laparoscopic resection.
Case Report. The 17-year-old girl initially presented with right lower abdominal pain and watery diarrhea for two weeks. She had no family history or signs of neurofibromatosis type 1 (NF1; Von Recklinghausen Disease). Abdominal computed tomography revealed a 5 cm mass in the ascending colon causing intussusception, and a laparoscopic right hemicolectomy with primary anastomosis was performed. The microscopic examination showed some spindle cells, and the immunohistochemical staining was positive for S-100 and CD-34 but negative for SMA, desmin, CD-117 and CK. These results support the diagnosis of low-grade MPNST.
Conclusions. After laparoscopic surgery, the patient recovered uneventfully and has been disease free for 24 months.
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Malignant peripheral nerve sheath tumor (MPNST) is a very rare neoplasm. In the general population, the reported incidence of MPNST is only 0.001%.[1] Although the tumor can arise in any part of the body, MPNST occurs most frequently in the extremities followed by the trunk, head and neck, but it is very rare in the gastrointestinal tract.[1,2] Additionally, most patients with MPNST are older than 35 years of age. Here, we present a 17-year-old girl with MPNST of the colon who was treated by laparoscopy. This could be the first report of MPNST found in the colon of a teenager.

Case Report

A 17-year-old girl presented with intermittent right lower abdominal pain for two weeks. The patient had no family history or signs of neurofibromatosis type 1 (NF1; Von Recklinghausen Disease). She was diag-

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nosed with acute appendicitis by a local medical doctor, and subsequent abdominal computed tomography (CT) revealed a polypoid mass in the ascending colon causing intussusception (Fig. 1). She was subsequently referred to our hospital under the diagnosis of an ascending colon tumor with intussusception.

A double-contrast colon series showed a rounded mass that was approximately 5.1 × 5.5 cm^2 in the ascending colon with intussusception (Fig. 2). The subsequent colonoscopy revealed a large cauliflower tumor that was noted in the ascending colon near the cecum (Fig. 3), and a biopsy revealed ulcerative tissue. The laboratory data revealed an elevated C-reactive protein (CRP) level of 6.8 mg/dl (normal range = 0-0.3 mg/dl). Two tumor markers, carcinoembryonic antigen (CEA) and carbohydrate antigen 19-9 (CA19-9), were within normal limits. Under the diagnosis of ascending colon tumor with intussusception, the patient underwent laparoscopic approach. The operation revealed a large cauliflower colon tumor (4 × 3 × 2 cm^3) near the cecum with intussusception and some regional lymph nodes along the ileocolic artery (Fig. 4). And right hemicolectomy with wide excision of lymphadenopathy was done (Fig. 5).

The pathological report showed a MPNST without lymph node metastasis. The surgical margins were free of tumor tissue. The tissue section had a submucosal mass containing some spindle cells with alterna-

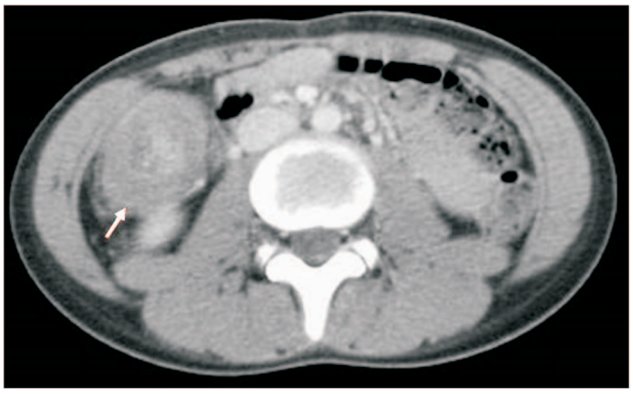

Fig. 1. Abdominal computed tomography (CT) revealed a polypoid mass in the ascending colon with intussusception.

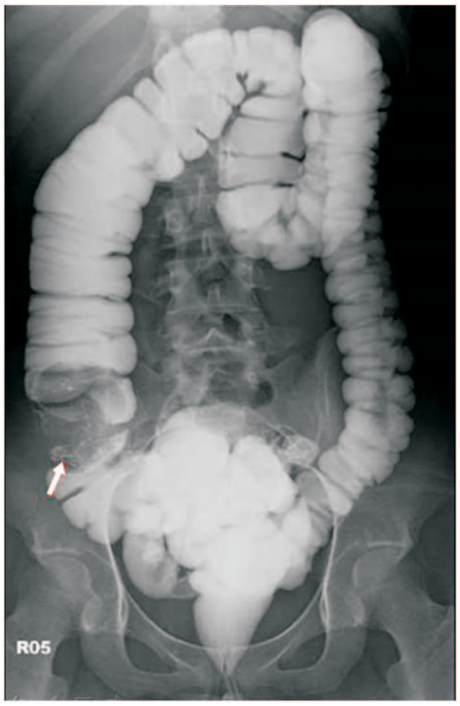

Fig. 2. Double-contrast colon series showed a rounded mass at the ascending colon with intussusception, measuring approximately 5.1 × 5.5 cm^2.

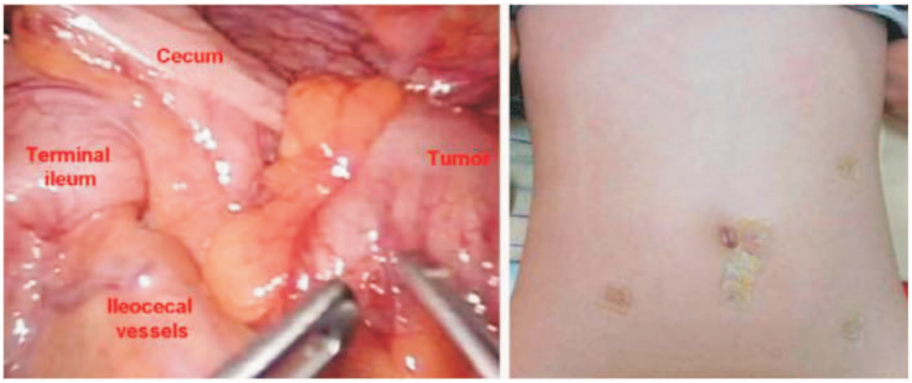

Fig. 3. Colonoscopy revealed a large cauliflower tumor in the ascending colon near the cecum with intussusception.

tive cellularity, intermixed with some delicate, branched small vessels and myxoid stroma. These spindle cells had wavy, hyperchromatic nuclei and indistinct cytoplasm, mixed with a few inflammatory cells. Among the spindle cells, mitosis was rarely observed. Based on immunohistochemical staining (IHC), these spindle cells were positive for S-100 and focally positive for CD-34, but they were negative for smooth muscle actin (SMA), desmin, CD-117 and pan-cytokeratin (CK). The wavy nuclear morphology and the S-100 protein expression indicated the neurogenic origin of these cells. These results support the diagnosis of low-grade MPNST without lymph node metastasis (Fig. 6 A; B; C).

The postoperative condition was uneventful. The patient was subsequently discharged one week after surgery. No adjuvant radiation or chemotherapy was given. She was followed-up regularly every three months at the outpatient department. The patient has been disease free for 24 months.

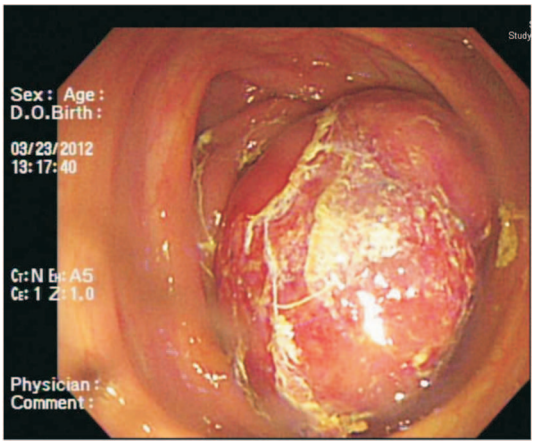

Fig. 4. Image of the large cauliflower tumor with intussusception located from the ascending colon near the cecum.

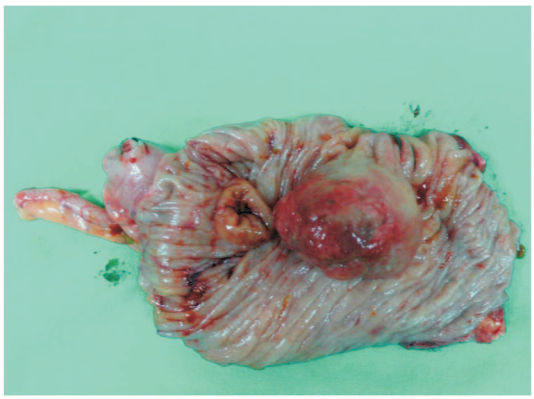

Fig. 5. Laparoscopic right hemicolectomy with wide excision of the lymphadenopathy.

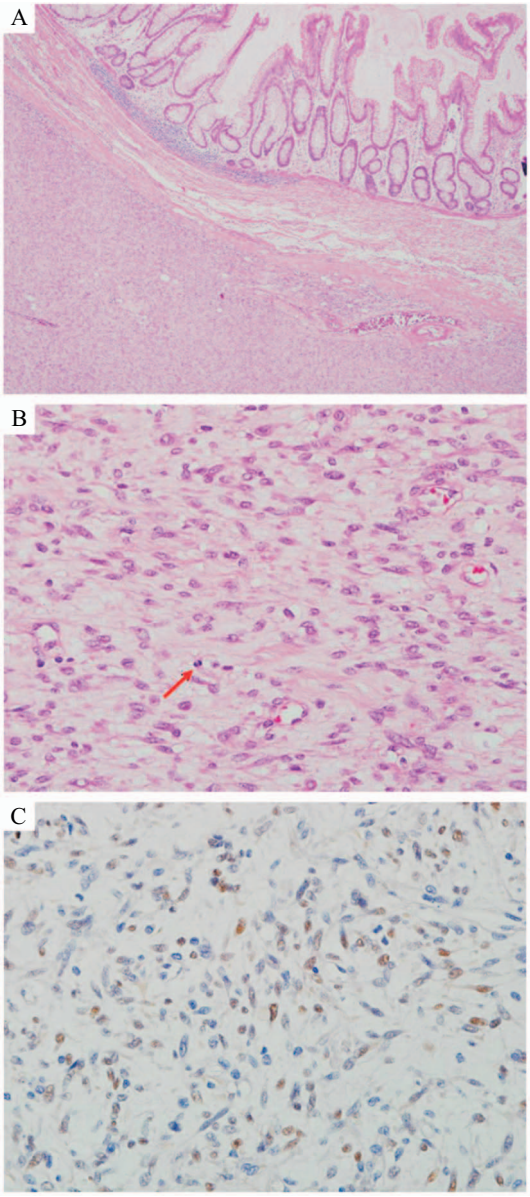

Fig. 6. Microscopy showed (A) a submucosal tumor containing spindle cells with myxoid stroma and varying vascularity (H & E, × 40); (B) spindle cells with wavy, mildly hyperchromatic nuclei within a myxoid matrix in the tumor, displaying mitotic features (arrow) (H & E, × 400); (C) frequent, positive staining for S-100 in the nuclei of the tumor cells (immunohistochemical staining, DAB as a chromogen) (H & E, × 400).

Discussion

MPNST is a soft tissue neoplasm that usually arises from the peripheral nerves, showing variable differentiation toward cellular components of the nerve sheath.[2] This malignancy is very aggressive, comprising approximately 5-10% of all soft tissue sarcomas, but it rarely occurs in the colorectal tract.[3] MPNST in children and adolescents may be sporadic or arise as a secondary malignancy after radiotherapy.[3] When it occurs at a younger age, it is always associated with NF1.[2] However, our case was a teenager who lacked a history of NF1. Based on the finding of double-contrast colon series and the colonoscopy, an ascending colon tumor complicated with intussusception was identified. After performing the laparoscopy, the pathological report showed a MPNST without lymph node metastasis, which was then confirmed with immunohistochemical staining.

The term MPNST, as defined by the World Health Organization (WHO), refers to any malignant tumor arising from a peripheral nerve or showing nerve sheath differentiation, designated MPNST in 2002 to replace previous terminology, such as 'malignant schwannoma', 'malignant neurilemmoma', 'neurogenic sarcoma', and 'neurofibrosarcoma'.[4] We used the terms 'malignant peripheral nerve sheath tumor', 'malignant schwannoma', 'malignant neurilemmoma', 'neurogenic sarcoma', 'neurofibrosarcoma' and 'colorectal' to search the Medline and PubMed database from 1965 to 2014. There are only five reported cases of MPNST in the colorectal tract, which are listed in Table 1.[1,5-8] Neither the previously published cases nor this case were associated with NF1. We have carefully reviewed the patient's past and family history, and no signs of NF1 were observed. Therefore, the association between MPNST and NF1 should be examined further.

For most MPNSTs, the tumor may resemble a colon mass; the lesion tends to be white, solid and fleshy and sometimes has myxoid changes with frequent necrosis and hemorrhage.[2] Most primary MPNSTs are composed of spindle cells.[9,10] An appropriate immunohistochemical panel is essential for the final diagnosis of MPNST. In this case, the immunohistochemical results of positive staining for S-100 and CD-34 and negative staining for SMA, desmin, CD-117 and CK suggest that the tumor originated from the nervous system. MPNSTs can be categorized as low grade (roughly 15%) or high grade (roughly 85%).[10] Most MPNSTs fall into the high-grade category, demonstrating cytologic atypia, brisk mitotic activity, and hypercellularity with or without necrosis.[10] Low-grade MPNSTs show more generalized nuclear atypia and increased cellularity with low levels of mitotic activity.[11,12]

MPNSTs are rare, and no laparoscopic techniques have previously been applied to this tumor type.[1] The feasibility of laparoscopic resection for colorectal tumors is well accepted, and the laparoscopic approach was chosen for a number of advantages, including the avoidance of the disability associated with a midline incision in the early postoperative recovery period, shorter hospital stay, and better cosmetic outcomes. Furthermore, laparoscopy facilitates excellent visualization with precise dissection in a limited space between the tumor and neighboring structures.[1,5-7] The case is a 17-year-old girl with only partial obstruction of the bowel. For the life quality, postoperative wound care and recovery, the laparoscopic surgery was ar-

Table 1. Reports of MPNST in the colorectal region

Site	Year	Authors	Sex	Age	NF-1	Tumor size	Treatment	Outcome
Rectum	1965	Bodner and De los Santos[6]	NA	NA	No	NA	NA	NA
Rectum	1996	Reinbold et al.[7]	M	72 yr	No	10 cm	Surgery	NA
Rectum	2001	Catania et al.[5]	NA	NA	No	NA	NA	NA
Descending colon	2002	Sung et al.[8]	M	43 yr	No	12 cm	Surgery	NA
Ascending colon	2006	Lee et al.[1]	F	2 days	No	5 cm	Surgery	Disease-free for 17 mo

NA: Not available.

ranged after the discussion with her family. This is the first report of successful laparoscopic treatment for MPNST.

Radiation therapy (RT) and chemotherapy treatment (CT) can improve local control of tumor margins if a tumor-free margin is difficult to attain by surgery alone.[14] Adjuvant radiotherapy is usually provided for high-grade tumors or microscopic residual diseases, but MPNSTs in the gastrointestinal tract appear to be resistant to chemotherapy because these tumors have limited sensitivity. Therefore, the applicability of adjuvant treatment for MPNSTs of the gastrointestinal tract is currently unclear.[1] Surgical resection remains the mainstay of treatment.[2] The tumor in our case was low-grade and had a resection margin free of tumor tissue. Therefore, post-operative adjuvant radiation therapy and chemotherapy treatment were not applied.

MPNST has a high local recurrence rate and a relatively poor prognosis.[3,13] The risk factors for a poor prognosis include a tumor greater than 5 cm, a high tumor grade, a history of NF1, elderly patients, distant metastases at the time of diagnosis, and an inability to attain tumor-free margins.[14-16] Accurate preoperative diagnosis for colon MPNST is rarely achieved because MPNST lacks specific symptoms or signs. Surgical management is mandatory, and radical resection can result in a favorable outcome. In this case, the patient was healthy and problem-free after surgery. This success uneventful outcome suggests that the laparoscopic approach may be is feasible for colon MPNST.

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病例報告

大腸惡性周邊神經鞘膜瘤青少年的罕見病例：案例報告及文獻回顧

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惡性周邊神經鞘膜瘤是一種少見的腫瘤，好發於四肢及軀幹，極少發生於大腸，治療以廣泛性手術切除為主，最後的確診則需靠病理檢查才能確定。我們報告一位 17 歲年輕女性患者，主訴腹痛伴隨著腹瀉兩個禮拜，而接受腹腔鏡右半大腸切除術，手術後證實為大腸低度惡性周邊神經鞘膜瘤，並無其它轉移情形，術後並未接受放射線治療或化學治療，門診追蹤兩年，無復發跡象。因屬罕見之惡性周邊神經鞘膜瘤病例，特就其細胞遺傳學提出報告。

關鍵詞 青少年、惡性周邊神經鞘膜瘤、大腸。