Adenocarcinoma of the Jejunum: A Pediatric Case Report

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The clinical history and surgical findings of an adenocarcinoma of the jejunum in a 15-year-old boy are reported. The diagnosis of small bowel carcinoma remains difficult, primarily because of the rarity of the disease and the ambiguity of its symptoms. We reviewed the literature on this subject and found a total of seven cases of adenocarcinima of the jejunum and ileum in patients under 20 years of age. We report this case for its rarity and the findings which should alert the pediatricians including intermittent abdominal pain, severe loss of body weight, and stools positive for guaiac. *(Chang Gung Med J 2003;26:204-7)*

Key words: adenocarcinoma, jejunum.

Small bowel malignancies are extremely rare in children. While this region of the bowel accounts for at least 75% the total length of the gastrointestinal tract and >90% of its mucosal surface area, <25% of all alimentary tract neoplasms and <2% of all malignant lesions occur here.[1-3] Adenocarcinoma is the most common type of small bowel cancer, constituting 32%-54% of all malignant enteric tumors, followed in the order of frequency by lymphoma and leiomyosarcoma.[4] The diagnosis of small bowel carcinoma remains difficult, primarily because of the rarity of the disease and the ambiguity of the associated symptoms.

Furthermore, the location is not conducive to visualization and biopsy. This usually results in a significant delay in diagnosis. Unlike gastrointestinal lymphoma, this disease cannot be cured by systemic chemotherapy thus, the outcome has generally been poor. A review of the literature provided only seven patients younger than 20 years with adenocarcinoma.[5] Adenocarcinoma of jejunum occurring at a young age are significantly rare to be worthy of reporting.

CASE REPORT

This 15-year-old boy had suffered from intermittent abdominal cramping pain, nausea and vomiting for 6 months. On admission, the patient had upper abdominal distention, poor appetite and a weight loss of 9 Kg within 6 months.

Physical examination revealed a thin, chronically ill-looking adolescent. There was no lymphadenopathy. The chest was clear and there was no heart murmur. The abdomen was flat and no definite mass was palpable. Palpation revealed tenderness over the left upper quadrant and a fullness above and to the left of the umbilicus. Rectal examination showed tarry stool. Admission laboratory studies revealed a hemoglobin of 12.7 g/dL and a hematocrit of 37.8%.

The patient reported an episode of anemia 6 months prior to admission. A hemogram at that time showed a hemoglobin of 8.3 g/dl, with a hematocrit of 28.3 % and he received blood transfusion therapy. Panendoscope and an upper gastrointestinal series were done and no definite abnormal findings were reported.

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After admission, signs of intestinal obstruction were obvious. The amount of nasogastric tube drainage was 1000 ml per day. Abdominal sonogram and computed tomography (CT) scan showed an irregular mass anterior to the left kidney with proximal bowel dilatation. Upper gastrointestinal (UGI) and small bowel series revealed a typical constriction with an apple-core appearance (Fig. 1).

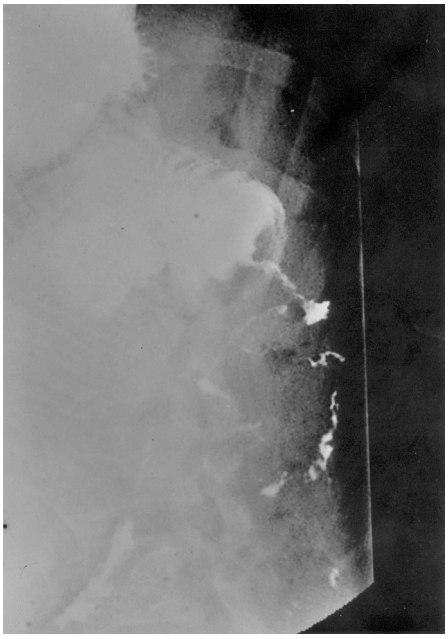

Fig. 1 Typical apple-core jejunal adenocarcinoma visualized in an upper GI series.

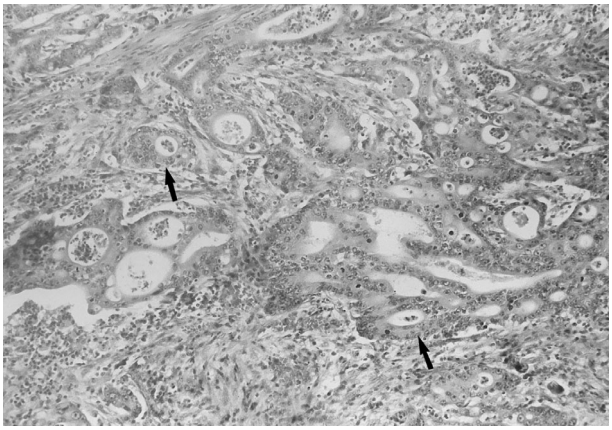

Fig. 2 Malignant neoplastic glands (arrows) infiltrating in a fibrous stroma of the intestinal wall (H & E, ×200).

Laparotomy disclosed a 6×4 cm polypoid mass arising from the proximal segment of the jejunum 50 cm distal to the ligament of Treitz involving the serosa and mesentry. A palliative resection was carried out with an end to end anastomosis. Pathology showed a poorly differentiated adenocarcinoma with mesentery lymph node metastasis (Fig. 2). The patient received chemotherapy 2 weeks after surgery. He is now being followed up in the pediatric oncology department.

DISCUSSION

Despite the introduction of new modalities such as endoscopy, CT scanning, and chemotherapy, the survival for small bowel adenocarcinoma has not improved during the past 30 years. Small intestine adenocarcinoma resembles adenocarcinoma of the colon. It arises from adenomatous polyp and grows silently at first and when vagus symptoms such as abdominal pain, anemia and chronic gastrointestinal bleeding occur, they are often attributed to other problems especially in young patients. As the small bowel contents are fluid, obstruction does not present until the disease is so advanced that stenosis is nearly complete. Clinical manifestations are similar between children and adults. Many researchers have noted the difficulty of making early diagnoses due to nonspecific symptoms compounded by the lack of physician vigilance; often the diagnosis is made only at time of surgery. The mean time to diagnosis in most series ranged from 6 to 8 months.[6] The time of delay from the onset of symptoms to diagnosis in our patient was 6 months.

Adenocarcinoma of the jejunum is most easily diagnosed using an upper GI (UGI) series with small bowel follow through. The overall accuracy of UGI series is 83%;[7] however, it depends on if the radiologist adequately visualizes the small bowel fluoroscopically. In our patient, the upper GI series was performed twice within 6 months before the lesion was demonstrated. Mucosal ulceration and annular constricting lesions are highly suggestive of adenocarcinoma. Abdominal sonogram and CT scanning were proved particularly useful in detecting mesenteric metastasis like the abdominal mass in our case.

Since early resection of nonmetastatic malignant neoplasms of the small intestine appears to be the

only chance for a cure, all investigators have stressed the need for early and radical resection.[2] Overall actuarial 5-year survival rate is 40 to 50%, and 0% after palliative resection.[8,9] The infrequency and possible lack of awareness usually result in a late diagnosis when only palliative bypasses or resections are possible, as in our patient. There is no current established role for any adjuvant or therapeutic chemotherapy, since small bowel adenocarcinoma appears to be as resistant to chemotherapy as other gastrointestinal adenocarcinoma.[3] Intra-abdominal recurrence is the usual cause of death.

Primary adenocarcinoma of the jejunum is an uncommon tumor, and the peak incidence is in 50 and 60 year old patients.[2] Nevertheless, we cannot eliminate this type of lesion from consideration in young patients. This case represents the eighth reported case of adenocarcinoma of the small intestine in a patient younger than 20 years. Dorman et al[10] stated that cramping abdominal pain, weight loss and a palpable abdominal mass were a significant triad of symptoms for small bowel malignant tumors; therefore, especially in patients with such long-standing manifestations, pediatricians should be highly suggested of the disease and perform more aggressive diagnostic examinations. With greater awareness of adenocarcinomas of the small intestine, it is possible that earlier diagnosis and treatment will lead to improved overall survival.

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小孩空腸腺癌的病例報告

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小腸癌症的診斷在臨床上仍舊非常困難，一方面是因病例稀少而另一方面是因症狀怪異。而在小孩身上更是罕見。回顧現有的文獻發現在小於20歲以下的病人僅有7例。在此我們報告一個15歲的罕見空腸腺癌的病例，並期待從其病史和臨床發現如長期的腹部陣痛，急遽的體重減輕和大便內有潛血反應等，讓小兒科醫師能對此病有所警覺。(長庚醫誌 2003;26:204-7)

關鍵字：腺癌，空腸。

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