นิพนธ์ต้นฉบับ

Primary sarcoma of small bowel : retrospective study in 14 cases.

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Small bowel has more surface area than the remaining portions of the alimentary tract, but primary malignancy in this organ is rare. A retrospective study of primary sarcoma excluding malignant lymphoma was made between 1975 to 1984. Fourteen cases were recorded with a male to female sex ratio of 8:5, and an age range of 30 to 77 years old. Two to four cases were distributed at each decade from the third to the seventh decade. The major clinical manifestations were abdominal mass and pain. In most cases the lesion was located in the duodenum, jejunum and ileum in equal numbers, 3 at each site. All except one were histologically leiomyosarcoma. At the time of operation 2 of 14 cases disclosed evidence of matastasis. Problems in diagnosis and morphology that supported the diagnosis of leiomyosarcoma are reviewed.

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เสาวณีย์ เย็นฤดี, ชัชวาล ธรรมาภรณ์พิลาส. มะเร็งชนิดซาร์โคมาปฐมภูมิของลำไส้เล็ก : การศึกษาย้อนหลังจำนวน 14 ราย. จุฬาลงกรณ์เวชสาร 2533 กันยายน ; 34(9) : 675-680

มะเร็งของลำไส้เล็กพบน้อย เมื่อเทียบกับส่วนอื่น ๆ ของทางเดินอาหาร อย่างไรก็ดีมะเร็งที่เกิดจากเยื่อบุผิวลำไส้ยังคงพบมากเป็นลำดับหนึ่ง โดยเฉพาะที่บริเวณดูโอดินั่ม ได้มีการศึกษารวบรวมมะเร็งของลำไส้เล็กจาก series หนึ่งในช่วงหลัง ๆ พบว่ามะเร็งชนิดลิมโฟมาของลำไส้เล็กพบบ่อยที่สุดและพบที่ส่วนต้น ๆ ของเจจูนั่มและส่วนปลายของอิเลี่ยม ซึ่งในอดีตมักพบที่ส่วนปลายของอิเลี่ยม อย่างไรก็ดีที่กล่าวมาแล้วเป็นเพียงรายงานใน series ค่อนข้างใหญ่ series หนึ่งเท่านั้นเอง ในที่นี้ได้ศึกษาย้อนหลังเกี่ยวกับมะเร็งชนิดซาร์โคมาปฐมภูมิของลำไส้เล็กโดยไม่รวมลิมโฟมา ระหว่างปี 2518-2527 รวบรวมได้ 14 ราย เป็นอัตราส่วนชายต่อหญิงเท่ากับ 8:5 อายุของผู้ป่วยอยู่ระหว่าง 30 ปีถึง 77 ปี อาการที่สำคัญได้แก่มีก้อนในท้องและปวดท้อง ตำแหน่งที่แน่นอนของมะเร็งเท่าที่ทราบอยู่ที่ดูโอดินั่ม เจจูนั่มและอิเลี่ยมตำแหน่งละ 3 ราย และอีก 1 รายอยู่ที่รอยต่อระหว่างอิดูโอดินั่มและเจจูนั่ม จุลพยาธิของซาร์โคมาส่วนใหญ่เป็นมะเร็งที่เกิดจากเซลล์กล้ามเนื้อเรียบ ขณะที่ทำการผ่าตัดมีการแพร่กระจายแล้ว 2 ราย ได้กล่าวถึงปัญหาในการวินิจฉัยและจุลพยาธิสภาพที่ช่วยในการวินิจฉัยมะเร็งชนิดนี้.

In comparison with other portions of the gastrointestinal tract, primary malignancy of the small intestine is rare. However adenocarcinoma is the most common neoplasm in this region particilarly in the duodenum.[1]

However during the period of our study, malignant lymphoma out-numbered the remaining malignancies.[2] Recently a new pattern of distribution of malignancy in the small intestine was reported. Malignant lymphoma was the most common malignant lesion and primary carcinoma were frequently located at the diatal part of the small bowel.[3] Of the latter excluding lymphoma, leiomyosarcoma was out-numbered by other soft tissue sarcomas.[1,3,4] Liposarcoma, fibrosarcoma, malignant schwannoma and angiosarcoma were extremely rare.[5,6] The present paper concerns a retrospective study of 14 cases of primary soft tissue sarcoma during a 10 year period. The purpose is to evaluate clinical features. Moreover the pathology and extent of the lesion are reviewed for academic interest and predicion of prognosis.

Material and methods.

Surgical records of primary malingancies fo the small bowel were reviewed between 1975 to 1984. There were 14 cases of soft tissue sarcoma. The clinical features including age, sex and leading signs and symptoms were tabulated. Definite location in the small intestine with histological types were also recorded. The pathological study based on H & E sections in all cases. Special staining with Masson's trichromes stain for muscle and collagen and Gridley modification for reticular fibres was performed.

Result

Of 14 cases of soft tissue sarcoma of the small bowel, 8 cases were male and 5 cases were female. Two to three cases were regularly distributed in each decade, from the third to the seventh decade. Abdominal mass was the most frequent clinical presentation. In 7 cases the location of the mass was in mid abdomen, epigastric area or the history was not available. The frequent manifestation was abdominal pain or discomfort in 5 patients. Two patients each presented with suprapubic pain and midabdominal pain, table 1. Anemia with melena and jaundice was the chief complaint in the remaining 2 patients. Three cases each was found in the ileum, the jejunem and the duodenum. Whereas the lesion was not labeled in 4 cases and the remaining one at duodenojejunal junction. All cases were leiomyosarcoma. figure 1 except one which was pleomorphic sarcoma, figure 2.

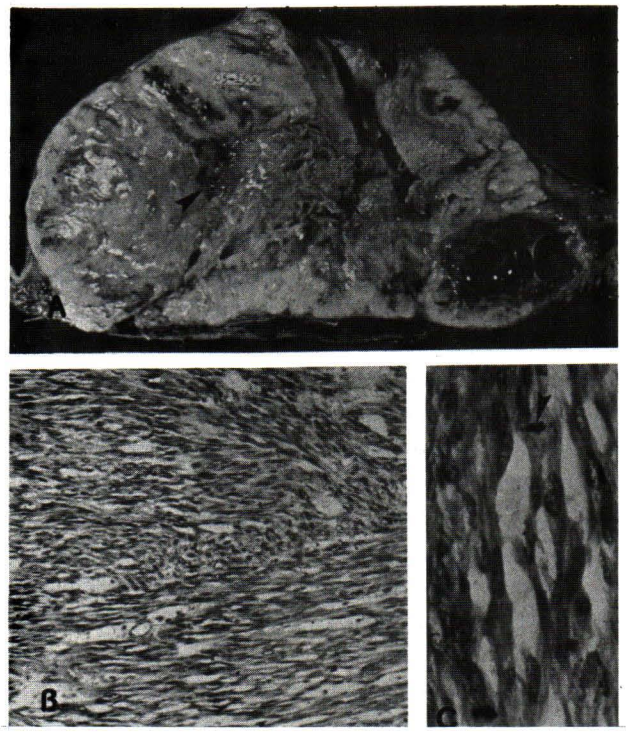

Figure 1. A. Large bulky mass of leiomyosarcoma showing hemarrhege, arrow head and necrosis, arrow.
B. Light microscopy showing interlacing bundles of cellular smooth muscle cell. H & E × 200.
C. Mitosis in smooth muscle cell, arrow head. H & E × 400.

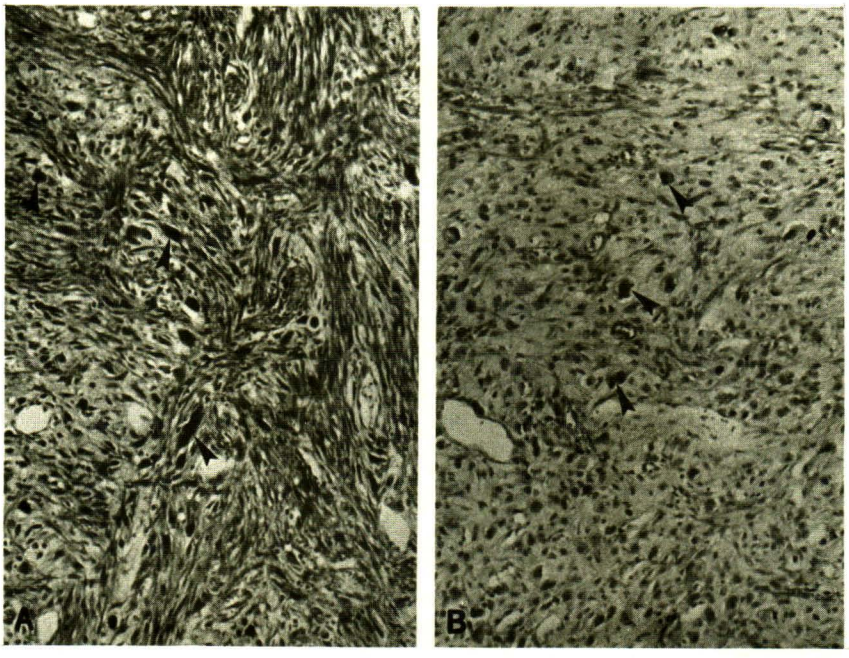

Figure 2. A. Pleomorphic sarcoma; pleomorphic spindle cells, (arrow head) arranging in haphazardous pattern. H & E × 200
B. Same case as in A showing bizarre cells, arrow head. H & E × 200.

Discussion

Although soft tissue sarcoma of the small bowel is a rare malignancy, leiomuosarcoma is the most common in this region.[7,8] Sarcomas of the other tissues that are components of normal structure of the intestinal wall are scantily observed.[5] Leiomyosarcoma is usually seen in the fifth and sixth decades with a male preponderance.[5,9] In the present series the male to female sex ratio was also 8: 5 but the age distribution was regularly distributed from the third through to the seventh decades. It may have been because of the small number of patients in the series analysed. Bleeding, pain and palpable mass are commom presentations, in that order.[5,9] Abdominal mass was the most frequent leading manifestation in the series studied, whereas abdominal pain or discomfort was the 2nd most frequently encountered complaint. In fact bleeding and pain are the chief clinical features of small bowel malignancies. The patient may ignor mild degrees or transient episodes of bleeding as well as discomfort. They come get treatment when the malignancy has advanced so that detectable mass is the leading feature. Soft tissue sarcoma is commonest in the jejunum and in descending order of frequency in the ileum and duodenum respectively.[1,9] Equal number of cases was seen in the ileum, jejunum and duodenum but 4 cases were not labelled. As previously mentioned most cases of soft tissue sarcoma in this area are leiomyosarcoma. All cases except one in the present series were also leiomyosarcoma. Two cases showed metastasis into the messentery and peritoneum. The latter was an index of advanced malignancy. In general leiomyosarcoma of the gastrointestinal tract are larger than leiomyoma. The malignant smooth mucle tumor is expressed as intraluminal, serosal or tranmural mass.[9] Microscopically it is not difficult to diagnose the smooth muscle tumor, but to say whether the lesion is malignancy or not, particularly when the mass is small or has histological variation. Well differentiated leiomyosarcoma is an example especially when an incisional biopsy or a small tumor mass is submitted. Mistosis is a useful histologic criterion in diagnosis of leiomyosarcoma,[7] however other microscopic features are also adjunct to diagnosis.[7] Cellularity is more frequent in the leiomyosarcoma than the benign one. Cytologic atypia is usually present in malignancy as well as necrosis. Hyalinization and calcification are benign features. Myxoid change is noted in both forms of tumors but rather ferquently in the leiomyoma whereas mucosal ulceration is present in both but preponderantly in malignant form.[7] Leiomyosarcoma arises from smooth muscle of the muscular layer or muscularis mucosae of the intestinal wall. At the time of deagnosis it is difficult to determine its origin. However the muscular

layer of intestinal wall is primarily considered.

The prognosis is influenced by the advanced nature of the disease at the time of operation. Cellular atypia and number of mitosis are additional prognotic indices.[6] Majority of cases died from metastasis and the remaining cases exhibits low 5 years survival rate. Intraabdominal metastasis at the time of operation, delayed surgical management, failure of chemotherapy or radiotherapy contribute to poor prognosis.

Recently cytogenetic study of chromosome of leiomyosarcoma of the small bowel is performed. The previous chromosome study was rather characteristic for this malignancy that occure in this region of the alimentary tract.[10] This study may be helpful in the prediction or the early detection of this malignancy in the future.

Table 1. Age, Sex and Clinical History.

No.	Age	Sex	Clinical History
1.	57	M	Suprapubic pain for 3 months
2.	43	M	Suprapubic mass with pain for 5 days
3.	67	F	Abdominal pain with fever 2 days
4.	34	M	Left epigastric mass for 3 months
5.	63	F	History of anemia with 2 days melena
6.	38	M	Palpable midabdominal mass with abdominal discomfort
7.	NA	NA	Palpable abdominal mass with occasional pain for 3 month
8.	77	F	Weight loss, anorexia, suprapubic mass and mucous stool with abdominal pain 6 month
9.	64	M	History of hepatic trauma with postoperative abdominal distension and subsequent surgery showing mass in 1st part of duodenum and in mesentery
10.	50	M	Mass at lower abdomen for 2 months
11.	30	M	Jaundice for 2 months
12.	34	F	Abdominal mass for 2 months with subsequent gut obstruction
13.	76	M	NA
14.	43	F	NA

Abbreviation : NA = not available

Table 2. Location and Pathology.

No. of patients	Location	Patholog
4	Small bowel (definite location was not available)	Leiomyosarcoma : 3 cases (Metastasis to mesentery : 1 case Pleomorphic sarcoma : 1 case)
3	Ileum (Terminal ileum : 1 case)	leiomyosarcoma
3	Jejunum	Leiomyosarcoma
3	Duodenum (1^{st} part : 2 cases)	Leiomyosarcoma (Metastasis to peritoneum : 1 case)
1	Duodenojejunal junction	Leiomyosarcoma

References

1. Reyes EL, Talley RW. Primary malignant tumors of the small intestine. Am J Gastroenterol 1970 Jul : 54(1) : 30-34
2. Yenrudi S, Dharmmaponpilas J. Malignant lymphoma of the small intestine. Chula Med J 1987 Dec; 31(12) : 971-979
3. Taggart DP, Imrie CW. A new pattern of histologic predominance and distribution of malignant diseases of the small intestine. Surg Gynecol Obstet 1987 Dec; 165(6) : 515-8
4. Nance K, Reddick RL. Epithelioid leiomyosarcoma of the small intestine with oncocytic change. Arch Pathol LabMed 1987 Dec; 111(12) : 1181-2
5. Ashley SW, Wells SA Jr. Tumors of the small intestine. Semin Oncol 1988 Apr; 15(2) : 116-28
6. Taxy JB, Battifora H. Angiosarcoma of the gastrointestinal tract: a report of three cases. Cancer 1988 Jul; 62(1) : 210-6
7. Ranchod M, Kempson RL. Smooth muscle tumors of the gastrointestinal tract and retroperitoneum; a pathologic analysis of 100 cases. Cancer 1977 Jan; 39(1) : 255-62
8. Kieffer RW, McSwain B, Adkins RB Jr. Sarcoma of the gastrointestinal tract : a review of 40 cases. Am Surg 1982 Apr; 48(8) : 167-9
9. Akwari OE, Dozois RR, Weiland LH, Beahrs OH. Leimoyosarcoma of the small and large bowel. Cancer 1978 Sep; 42(3) : 1375-84
10. Dal Cin P, Boghosian I, Sandberg AA. Cytogenetic findings in leiomyosarcoma of the small bowel. Cancer Genet Cytogenet 1988 Feb; 30(2) : 285-8