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Neoplasms of Gastric Smooth Muscle*

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위의 평활근에 생기는 종양

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정 기 용

위의 평활근에 생기는 종양인 평활근종, 평활근육종, 평활근육아세포종은 그 발생빈도가 매우 낮으며 주로 백인에 많고 남녀의 발생비율은 2 : 1로 남자에 많으며 주로 40—50대에서 호발한다.

주요 증상은 종양의 종류나 악성도에 따라 다르나 주로 위출혈, 복통, 조기포만감, 식욕부진 등이며 위내의 호발부위는 평활근종의 경우는 위중부, 평활근육종에서는 위체부, 평활근육아세포종에서는 위전정부와 위분문부이다. 이들 종양들은 대체로 단단하며 잘 피막화되어 있어 초기에는 양성종과 악성종의 감별이 어려우나 종양의 크기와 세포핵분별의 수가 악성도를 나타내는 지표가 된다. 여기에서 평활근종 1예와 평활근육아세포종 1예를 문헌고찰과 더불어 보고하고자 한다.

Introduction

Leiomyoma, leiomyosarcoma, and leiomyoblastoma are uncommonly seen in practice. While leiomyomas are benign lesions with an incidence of up to 50% in some autopsy series, they usually remain asymptomatic.[1] Leiomyosarcomas are distinctly less common, comprising from 0.25%[2] to 3%[3] of all primary malignant gastric neoplasms, but frequently cause abdominal pain, gastrointestinal hemorrhage, or weight loss. Leiomyoblaltoma is a rare tumor that appears to be of intermediate malignant potential.[4]

Representative examples of leiomyoma and leiomyoblastoma are presented and the pathophysiology and mangement of each type of gastric smooth muscle tumor is reviewed.

Case Reports

Case 1

This 51-year-old white male was admitted to the internal medicine department on July 15, 1978 with a long history of indigestion, abdominal discomfort, progressive weakness, and tarry stool. Past history was unremarkable. At the time of admission, the hemoglobin was 10.4gm% and the hematocrit was 28%. A diagnosis of anemia of chronic disease was made after barium enema and upper GI series failed to show a significant lesion. He was discharged with iron home medication and referred for further evaluation serveral months later. Two months later, he was readmitted because of melena. At that time, the hemoglobin was 8.6gm% and hematocrit was

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25vol%. After transfusion of 2 units of packed red cells, the hemoglobin was increased to 10.2gm %. Other laboratory findings were within normal limits. On physical examination, the abdomen was soft but mildly distended. No intraabdominal mass or organomegaly were palpable. A repeat upper GI series showed a mass lesion in the antrum of the stomach.

Barium enema was normal. Endoscopy revealed a 7cm rounded mass on the lesser curvature with a 2cm area of erythema and central ulceration. Biopsy and cytology were nondiagnostic. On Oct. 3, 1978, an operation was performed. The patient underwent a 50% gastrectomy with Billroth I anastomosis and wedge biopsy of liver. No gross lymph node or other organ involvement were noted.

Microscopically, the tumor composed as spindle shaped cells with abundant eosinophilic cytoplasm and mild nuclear pheomorphism, consistent with the diagnosis of leiomyoma. Only one mitotic figure was found in 75 HPF. The lymph node and liver tissue were normal. The patient's postoparative course was uneventful and there had been no evidence of recurrent disease.

Case 2

A 69-year-old black male complained of increasing abdominal girth, early epigastric fullness, swelling of lower extremity and 5Lb weight loss over 4 months preceding admission. He had had no significant prior medical problems. He was a heavy alcohol drinker as a young man but he denied taking alcohol for 40 years, and he was a nonsmoker.

Physical examination revealed a slender black man with tensely distended abdomen measuring 93cm in girth. No intraabdominal mass or organomegaly was palpable. There was central dullness to percussion with tympany in the flanks. A fluid was felt to be present but there were no evidence of liver disease. Moderate edema of lower extremities ware present to midcalf. Other examination was unremarkable. In laboratory study hemoglobin was 12.2gm%, the hematocrit was 35% and red cell indices were normal. The leucocyte count was 6200 with normal differential count. Urinalysis was normal. Total bilirubin, LDH, SGOT, serum amylase, alkaline phosphatase and uric acid were normal. Serum albumin was 3.4gm/100m*l* and total protein was 6gm/100m*l*. The prothromain time was prolonged but the partial thromboplastin time and thrombin clotting time were normal. His chest X-ray was normal. Plain flims of the abdomen revealed displacement of the small bowel to the left.

On upper GI series, the stomach was horizontal with the antrum and duodenal bulb being displaced to the left of the midline by an extrinsic mass. No gastric mucosal abnormalites were noted. The barium enema was unremarkable except for outward displacement of the colon. Intravenous pyelogram was consistent with extrinsic pressure on the distal ureters and bladder. An abdominal ultrasound confirmed the presence of a large cystic structure that appeared to contain multiple echogenic foci.

The possiblity of intraperitoneal carcinomatosis was entertained and a paracentesis was performed. Cytology of the peritoneal fluid revealed no evidence of malignant cells. The cell count was 6000 with 30% polymorphs, 55% lymphocytes and 15% tissue cells. The protein was 44mg%/dl. The amylase and LDH were normal.

An abdominal CT scan was done and revealed marked diaplacement of intra abdominal structures by a homogeneous mass filling the peritoneal cavity, The stomach, colon, kidney, liver and pancreas were normal appearing but displaced. The endoscopy revealed gastritis.

On April 10, 1980, a laparotomy was performed. At operation, a large cystic structure originating from a baseball size hard mass in the lesser omemtum was found. When the entire cystic mass was freed and isolated from all adhesions, it was found the mass was tenuously attached to the lesser curvature of

the stomach. When frozen section specimens were tentatively read as leiomyosarcoma, a hemigastrectomy and Billroth I anastomosis were performed. Other intraabdominal organs were normal and there was no evidence of metastasis.

The resected specimen measuring 25×27×14 cm in greatest demension weighed 7.0kg. Examination of the permanent sections revealed histology consistent with a gastric leiomyblastoma.

Postoperatively, the patient's recovery was uneventful and by five months after operation he was doing well and gaining weight.

Discussion

The age and sex distribution of all gastric smooth muscle neplasm is similar.[1,5,6] Males predominate over females two to one and whites far outnumber nonwhites. These lesions are found predominantly between the age of 40 and 70 with the largest number noted in the fifth decade. Malignant lesions tend to present at the younger age..

Leiomyomas are the most common benign gastric neoplasm. They are usually asymptomatic with the majority being found either at autopsy or incidentally at exploratory laparatomy for other diseases. However, almost all leiomyomas larger than 3cm are symptomatic, causing either gastrointestinal hemorrhage or abdominal pain,[1] which are also the prodominant symptomatic features of leiomyosorcoma and leiomyoblastoma. Bleeding with either hematemesis or melena occurs in about 50% of patients with these tumors.[2,4] Although the majority of patients experience chronic blood loss with iron deficiency anemia, sudden massive hemorrhage requiring emergency surgical intervention has been noted in several patients.[4] This hemorrhage is the result of mucosal ulceration overlying the tumor. Abdominal pain also frequently occurs.[1,2,4] Although it may be easily confused with ulcer pain, being intermittent and boring in nature and usually located in the epigastrium, the pain may also be relatively mild and generalized in location. Weight loss is more frequent with malignant tumors.[4] Anorexia, early satiety, dysphagia, nausea, vomiting and backache occur infrequently.

Examination is usually unrewarding although an intraabdominal mass may occasionally be palpable. While most palpable lesions will be malignant, size alone is not a reliable indicator of malignancy.[1,4] Obstruction is an unusual result of the tumors. Therefore, even when a large mass is present, findings of gastric distension or stasis are infrequent.

The primary diagnostic examination is the upper GI barium study that will demonstrate a lesion in 80—90% of patients.[2,4] Filling defects, with or without central ulceration, are typically seen. Extrinsic masses or polypoid lesions are less common. There are no distinguishing features of malignancy although malignant tumors are usually much larger.[2] The findings at endoscopy are variable with mass lesions, ulcerations, polyps, and infiltrating lesions having been seen.[5]

Malignant tumors tend to be locally aggressive and distant metastasis is unusual. Regional lymph node involvement is by direct extension only. In late cases, the liver may be involved and, rarely, metastasis may occur in the lung. Hepatic masses may be seen as filling defects on liver-spleen scan. Plain chest X-ray will demonstrate a noncalcified mass in the unusual case.

In the stomach, as in the uterus, the distinction between benign and malignant smooth muscle neoplasmms is not always clear cut.

The pars media of the stomach is the most frequent location for leiomyoma, but a significant number also arise in the fundus and antrum.[7,8] The majority of leiomyomas are submucosal with the remainder being either serosal or intramuscular. As a result, endos-

copic biopsy infrequently establishes a diagnosis. Leiomyomas infrequently exceed 10cm in diameter and most are less than 5cm in diameter.[1,7,9] However, benign neoplasms up to 40cm in diameter have been reported. Degenerative changes, such as liquefaction, hyalinization and calcification, are common.

Microscopically, the classic leiomyoma is composed of well-differedtiated, spindle-shaped cells organized into fascicles or bundles with variable amounts of connective tissue. Mitotic activity is low, generally less than one mitotic figure per 10 HPF.

In one series of 44 gastric sarcomas recorded by Appelman and Helwig,[10] 40% arose in the body of the stomach with the remainder evenly distributed among the cardia, fundus and antrum. Both gastric curvatures are equally involved. Intramural, submucosal and subserosal locations all occur commonly and both exogastric and endogastric growth are possible. Mucosal ulceration over the tumor also occurs more frequently. Microscopically mitotic activity is the most useful indicator of malignancy. Over 5 mitosis/10 HPF signifies aggressive behavior. Unfortunately, however, up to 40% of leiomyosarcomas proved by metastasis have fewer than this number.[11] Increased cellularity, cytologic atypia, and necrosis are all more common in leiomyosarcoma but may occur in benign gastric smooth muscle neoplasms.

Because leiomyoblastomas may be benign or malignant, some authors prefer to use the terms "epithelioid leiomyoma" and "epithelioid leiomyosarcoma."[4] The criteria for distinguishing benign from malignant leiomyoblastomas are the same as those previously described. Cystic degeneration may be more common in leiomyoblastoma than other smooth muscle neoplasm of the stomach. There may be greater tendency for occurrence in the antrum and cardia with a high incidence of malignancy in the latter site.[4]

Treatment and Results

The treatment of choice for these lesions is wide surgical excision.[1,2,3,5] Because the gross features of leiomyosarcoma may resemble those of the benign leiomyoma and frozen section diagnosis of malignancy in these tumors may be difficult, all smooth muscle tumors should be extirpated entirely. The highest survival rates are reported in those patients undergoing anatomical resections that include the appropriate lymphatic drainage.[2] For patients with unresectable intraabdominal disease, palliative gastric resection may be advisable for both debulking of tumor prior to postoperative radiation and for prevention of hemorrhage obstruction since some patients with extensive disease may survive for long periods.[1]

While postoperative radiation may bring about an occasional remission, statistically it has not increased survival.[2] No effective chemotherapeutic regimen is currently available and experience with those tumors is sparse. High-dose selective hepatic chemotherapeutic infusions have been suggested as a adjuvant therapy for patients at high risk of recurrence following curative resection,[5]

Leiomyomatous tumors when resected entirely with free margins of normal tissue have essentially 100% cure rate and recurrences are rare.[1] Unfortunately, leiomyosarcoma is not so successfully treated. Survival rates range from 25—50% at five years.[1] In patients treated with curative resection,[5] recurrence is primarily in the liver but occasional patient with locally recurrent disease may be benefitted by reresection. Patients with tumors larger than 8cm in diameter, extension of the tumor to the serosal surface, or low grade tumor differentiation have a significantly poorer outlook,[5] and are candidates for postoperative attempts at adjuvant therapy. Leiomyoblastoma uncommonly behaves in a malignant fashion (2%[6] to 12%[1]).

Conclusion

Smooth muscle tumors of the stomach are uncommonly encountered in clinical practice. Because of the difficulty of immediate pathologic identification, adequate extirpative resection and not enucleation of all these tumors is a necessary part of treatment.

Using these principles one may reasonably expect to cure 100% of leiomyomas resected. Leiomyosarcomas should be cured in about one half of cases with failures primarily the result of widespread disease at the time of diagnosis. Leiomyoblastoma usually follows a benign course. However, occasional variants with malignant behavior do occur. For this reason, as many as one in ten patients with leiomyoblastoma will die to their disease after resection.

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