CASE REPORT

Diffuse Leiomyomatosis of the Esophagus

A Case Report and Review of the Literature

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Although leiomyoma is the most common solid benign tumor of the esophagus (1), it is a very rare condition and the vast majority of the leiomyomas of the alimentary tract (more than 90%) occur outside the esophagus (1).

In 1961 Gray et al (2) collected 345 reports of leiomyoma of the esophagus, published between 1559 and 1959, and recently Dillow et al (3), reviewing the literature, found 180 cases since 1959; they added 11 cases of their own.

Leiomyomatous neoplasms of the gastrointestinal tract usually form solitary, rounded or lobulated, well defined, although not encapsulated, masses. Other forms of leiomyomas, like multiple tumors or a more diffuse form without visible nodules, are very unusual.

CASE REPORT

FGN, A 21-year-old white woman was admitted to Santa Maria Hospital, Lisbon, on August 7, 1969, because of a 5-year history of difficulty in swallowing.

She had been in excellent health until 1964 when she noticed a sensation of food sticking at the level of the lower sternum. The difficulty in swallowing was persistent, and it included liquids and solids. Some months later she developed regurgitation, usually immediately after eating. Her complaints became progressively more severe, particularly 5 or 6 months prior to her admittance to the hospital, and occasionally she was unable to eat for 1 or 2 days. In the 3 years prior to hospitalization her general physical condition had deteriorated and she had lost 11.5 kg. She never had epigastric or retrosternal pain, heartburn, hematemesis or melena, or symptoms of pulmonary infection or Raynaud's phenomenon. There was no history of previous ingestion of corrosive fluids. In June, 1967, she was examined by her private physician and a barium swallow was accomplished. The radiological examination of the esophagus showed minimal diffuse dilatation and narrowing of the distal end of the esophagus, asymmetrical (not funnel shaped), and smooth. There was also a filling defect of the inner zone of the gastric bubble.

Physical examination showed nothing unusual except marked emaciation (weight of 45 kg). Laboratory data were within normal limits and the guaiac test was negative. The barium swallow performed in Santa Maria Hospital showed the same configuration of the distal end of the esophagus seen in previous examination 2 years before, but now the esophagus above the stricture was more dilated. In addition, the body of the stomach showed a filling defect with an image suggestive of an ulcer on the vertical part of the lesser curve (Figure 1). Esophagoscopy showed a stricture of the lower end of the esophagus; the esophagoscope could not be introduced beyond this level. Above the narrowed area the mucosa was normal and there was no sign of esophagitis or infiltration. Esophageal exfoliative cytology failed to reveal evidenced malignant cells.

During the operation, a firm indefinable mass was found arising in the lower end of the esophagus, extending to the body and lesser curve of the stomach, and to the body and tail of the pancreas. At the lesser curve, there was a large ulcer. To the surgeon the lesion appeared to be an invasive malignant tumor. There was no evidence of metastases or enlarged lymph nodes. A total gastrectomy plus terminal esophagectomy, splenectomy, and partial pancreatectomy, with a Roux-en-Y reconstruction was performed.

On microscopic examination, the tumor proved to be a leiomyoma of a diffuse type, which may be termed a diffuse leiomyomatosis of the esophagus, with invasion of the stomach. The pancreas was involved only by fibrosis. The postoperative course was uneventful and the patient was discharged on the 20th hospital day.

In February, 1970, 6 months later, she was readmitted to Santa Maria Hospital for evaluation. In the interim, she had been relatively well, although she did not gain any weight. With small feedings of bland foods every 2 hours, the patient complained only of minimal epigastric discomfort after meals, sometimes characterized as pain. She

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LEIOMYOMATOSIS OF ESOPHAGUS

had no difficulty in swallowing. The first 2 months after the operation she had some fluid regurgitation. She never had diarrhea or symptoms suggestive of dumping syndrome.

There was no anemia; the total protein value was 6.9 g/100 ml; albumin, 3.5 g/100 ml; calcium, 9.2 mg/100 ml; phosphorus, 3.8 mg/100 ml. The fasting blood sugar level was 58 mg/100 ml, and a glucose tolerance test showed 192 mg/100 ml at 30 min, 186 mg/100 ml at 60 min, 80 mg/100 ml at 120 min, and 45 mg/100 ml at 180 min. The fecal fat was measured for 3 days and averaged 7.7 g/day (2.9 g; 7.5 g; 12.8 g). Xylose excretion was 7.9 g/5 hr. Esophagoscopy showed minimal esophagitis. The roentgenogram of the esophagus revealed resection of the lower end of the esophagus and of the stomach, and the esophagojejunal anastomosis was functioning freely. Since February, 1970, the patient has been seen regularly at the out-patient clinic. She is well, with no dyspepsia, and weighs 48 kg at this time.

PATHOLOGY

The lower third of the esophagus and the upper portion of the stomach, especially on the lesser curve, were hard and thickened. On the serosal surface of the cardia, there was a firm, well-defined nodule, 2 cm in diameter. Just below the cardia, on the lesser curve, the mucosa showed an ulcer with an area of 4 × 2.5 cm.

Histologically abnormalities were present in all layers of the esophagus. They included particularly the following:

1. Marked diffuse hypertrophy of both muscular layers and muscularis mucosae with multiple areas where the normal fiber pattern of the musculature was replaced by irregular and plexiform fibers with whorl formation (Figures 2 and 3).

2. Extensive fibrous tissue formation, especially in both muscular layers and in the adventitia, leading to a dissociation of the muscular fibers.

3. In some areas of the submucosa, muscular coat, and adventitia there was a nervous and vascular hyperplasia, sometimes with a myomatous appearance of the muscular layer of the blood vessels with calcification in the internal elastic lamina.