Dysphagia 8:292–296 (1993)

Dysphagia

Malignancy-Induced Secondary Achalasia

Henry P. Parkman, MD and Sidney Cohen, MD
Department of Medicine, Temple University School of Medicine, Philadelphia, Pennsylvania, USA

Abstract. Secondary achalasia refers to the development of clinical, radiographic, and manometric findings of achalasia as a result of (i.e., secondary to) another underlying disorder. A variety of malignancies have been associated with secondary achalasia. Adenocarcinoma of the esophagogastric junction accounts for the majority of cases of malignancy-induced achalasia, however, noncontiguous tumors may also cause this disorder. Although rare, malignancy-induced achalasia will occasionally be encountered by gastroenterologists and gastrointestinal radiologists who see patients with dysphagia and/or achalasia. Since treatment is aimed at the underlying neoplasm, it is important to recognize this disorder. Three clinical features suggest the possibility of malignancy as a cause of achalasia: 1) short duration of dysphagia (<1 year); 2) significant weight loss (>15 pounds); and 3) age >55 years. The presence of any of these should at least raise a suspicion of malignancy. Diagnosis may not be evident on routine esophagrams and endoscopy, and requires clinical suspicion for further evaluation with thoraco-abdominal CT scanning and endoscopic ultrasonography.

Key words: Achalasia — Pseudoachalasia — Esophageal manometry — Esophageal aperistalsis — Deglutition — Deglutition disorders.

Achalasia is an esophageal motility disorder characterized manometrically by the complete absence of esophageal peristalsis, and an incompletely relaxing lower esophageal sphincter (LES). Radiographically, there is typically esophageal dilatation with tapering at the gastroesophageal junction to a characteristic narrowed "bird's beak" appearance (see Fig. 1). Patients present primarily with dysphagia. Secondary achalasia refers to the development of these same clinical, radiographic, and manometric findings of achalasia as a result of (i.e., secondary to) another underlying disorder [1].

Both benign systemic disorders (e.g., amyloidosis, Chagas' disease, or intestinal pseudo-obstruction) and malignant tumors may cause secondary achalasia. Malignancy-induced secondary achalasia is often referred to as pseudoachalasia [2,3]. A variety of malignancies have been associated with secondary achalasia (Table 1). Adenocarcinoma of the gastric cardia accounts for nearly ¾ of cases of malignancy-induced achalasia. Noncontiguous forms of cancer may also produce this syndrome, including lymphoma and primary neoplasms from the lung, pancreas, prostate, and liver.

Although well described in the medical literature, malignancy-induced secondary achalasia is rare and accounts for only 2–4% of patients with esophageal manometric findings of achalasia [2]. It is important to recognize secondary achalasia as the treatment is different than for idiopathic achalasia. In idiopathic achalasia, the treatment centers on reducing the LES pressure by forceful disruption of the circular muscle—either by pneumatic dilatation or surgical esophagomyotomy [1]. In secondary achalasia, treatment is aimed at the underlying neoplasm. Esophageal function may return to normal after the tumor is removed or treated by radiation therapy or chemotherapy [4,5].

Pathophysiology or Malignancy-Induced Achalasia

Several mechanisms have been proposed to explain how malignancies can cause secondary achalasia (Table 2) [1]. First, malignant strictures of the distal esophagus may produce an obstruction in the region of the LES and secondary degeneration of esophageal peristalsis. Adenocarcinoma of the gastric cardia may also infiltrate the

Address offprint requests to: Henry P. Parkman, M.D., Department of Medicine, Temple University Hospital, Parkinson Pavilion, 8th Floor, 3401 North Broad Street, Philadelphia, PA 19140, USA

and the patient was reported as T2N0M1. The patient recovered uneventfully, but died 10 months after surgery of mediastinal recurrence.

Case 4

A 78-year-old nonalcoholic, nonsmoker female was admitted with a 3-month history of dysphagia. Eleven years before she had been treated with mastectomy and radiotherapy for a left breast cancer. Radiation doses were unknown. She received no chemotherapy. Upper endoscopy with histological study showed an invasive esophageal squamous cell carcinoma, extending from 20 to 24 cm. No resection was carried out because of tracheal invasion and the patient died 1 month later.

Discussion

The relationship between ionizing radiation and upper cervical and esophageal cancer has been well established by different authors [1–6]. The concept of radiation-induced esophageal cancers can be established on experimental, clinical, and epidemiological bases, although they are infrequent, representing less than 1% of the esophageal cancers operated on in our department (personal observation).

Some experimental data suggest that radiation-induced esophagitis is dose dependent [8]. Experimental studies in the rat have demonstrated the effect of radiation on the esophageal mucosa and the high rate of radiation-induced esophageal cancer [9,10].

Acute and chronic radiation-induced esophagitis have been reported in humans [7,11–14]. Doll [3] and Goffman et al. [4] reported 10 cases of esophageal cancer among 14,000 patients who received radiotherapy for ankylosing spondylitis. The esophageal cancer risk was higher than the calculated risk for the population studied. Ueda et al. [6] reported a 0.33% rate of esophageal carcinoma in females irradiated for breast cancer.

In the present series, our patients have the following issues in common to support the origin of radiation-induced cancers: (1) esophageal cancer has developed in areas previously irradiated for other malignancies; (2) esophageal cancer was revealed after a long interval of from 8 to 11 years following radiotherapy; (3) in all of our resected cases, chronic postradiation fibrosis was present [11]; (4) none of our cases had a history of tobacco or alcohol abuse.

Dysphagia was the usual, initial symptom in all of our cases and none of them experienced preexisting functional symptoms, as previously reported [7]. Other causes of dysphagia, after a history of mediastinal irradiation, included mediastinal carcinomatosis secondary to breast cancer, benign postradiation stenosis, and esophageal location of Hodgkin's disease [7,11,15–17]. Accurate diagnosis can be performed using computed tomography scan, mediastinoscopy, and endoscopic ultrasonography [18].

In our cases, histological diagnosis was made with the first endoscopic biopsy samples, although some authors have pointed out difficulties due to inflammatory changes associated with radiation [19]. In such cases, stenosis dilatation followed by cytological studies and biopsies with vital coloration can be used [20].

Although none of our three surgical patients had radiation-induced dermatitis or pneumonitis, these conditions may influence the choice of thoracotomy side and the risk of respiratory complications [11].

Periesophageal fibrosis did not complicate mediastinal dissection in our resected cases; however, we do not advocate blunt dissection. Postoperative fistulae were not observed in this small series while anastomosis was being performed on irradiated esophagus. The use of stapler and the interposition of omentum between the anastomosis and the radiated aorta for that purpose may minimize the risk of fistulae [21].

Mediastinal lymph node dissection showed no metastasis in all of our cases; the only lymph node metastasis was a cardial one (Case 3) in a nonirradiated area. Mediastinal fibrosis involving lymphatic drainage of the esophagus could explain the absence of metastatic mediastinal lymph node. However, the absence of mediastinal lymph node involvement was not associated with a favorable survival. As reported by Ueda et al. [6], esophageal carcinoma developing after mediastinal irradiation is not different from common esophageal squamous cell carcinoma.

In conclusion, radiation-induced esophageal cancer is a rare entity that should be suspected in every patient with dysphagia and a previous history of cervicomediastinal radiotherapy. Radiation-induced esophageal cancer can be treated with an esophageal resection in most cases but is associated with a poor prognosis.

References

1. Goolden AWG: Radiation cancer. A review with special reference to radiation tumors in the pharynx, larynx and thyroid. *Br J Radiol 30*:620–640, 1957
2. Chudecki B: Radiation cancer of the thoracic esophagus. *Br J Radiol 45*:303–304, 1972
3. Doll R: Radiation's hazards: 25 years of collaborative research. *Br J Radiol 54*:179–186, 1981
4. Goffman TE, McKeen EA, Curtis RE, Schein PS: Esophageal carcinoma following irradiation for breast cancer. *Cancer 52*:1808–1809, 1983
5. Ito I, Miyaishi K, Mitsuhashi J, et al: Radiation-induced cancer of the esophagus after postoperative irradiation for breast cancer. *Nippon Acta Radiol 38*:985–991, 1978
6. Ueda M, Matsubara T, Kasum F, Nishi M, Kajitani T: Possible radiation-induced cancer of the thoracic esophagus after postoperative irradiation in breast cancer. *Nippon Kyobu Geka Gakkai Zasshi 39*:1852–1857, 1991
7. Rigaud A, Gimbergues H, Buffe P, Tardivel M, Legain J: Cancers radio-induits. *Ann Med Interne (Paris) 126*:837–845, 1975

8. Tochner ZA, Pass HI, Sindelar WF, De Luca AM, Grisell DL, Bacher JD, Kinsella TJ: Long-term tolerance of thoracic organs to intraoperative radiotherapy. *Int J Radiat Oncol Biol Phys 22:*65–69, 1992
9. Jennings FL: Acute radiation effects in the esophagus. *AMA Arc Pathol 69:*407–712, 1960
10. Gates O, Warrens S: Radiation-induced experimental cancer of the esophagus. *Am J Pathol 53:*667–679, 1968
11. Morichau-Beauchant M, Touchard G, Battandier D, Maire P, Fontanel JP, Daba A, Babin P, Matuchansky C: Oesophagites radiques chroniques après traitement des cancers ORL. *Gastroenterol Clin Biol 7:*843–850, 1983
12. Seaman WB, Ackerman LV: The effect of radiation on the esophagus. *Radiology 68:*534–540, 1957
13. Savary M: Cancer de l'oesophage: diagnostic endoscopique précoce. *Gastroenterol Clin Biol 9:*7–9, 1985
14. Sarrazin D, Fontaine F, Mouriesse H: Les complications de la radiothérapie des cancers du sein. *J Radiol 56:*805–812, 1975
15. Fékété F: Dysphagies par sténose extrinsèque de l'oesophage. *Med Chir Dig 3:*159–162, 1974
16. Lortat-Jacob JL, Fékété F, Garcia-Moran M: Dysphagie et cancer du sein. *Ann Chir Thorac Cardiovasc. 18:*975, 1964
17. Trotman BW, Glick JH, De Barros SGS, Atkinson BF: Dysphagia in a patient with Hodgkin's disease. *JAMA 244:*2552–2553, 1980
18. Murata Y, Muroi M, Yoshida M, Ide H, Hanyu F: Endoscopic ultrasonography in the diagnosis of esophageal carcinoma. *Surg Endosc 26:*123–127, 1987
19. Charles J, Fiasse R, Fingotj H: Les radio-cancers du pharynx et de l'oesophage. *Acta Gastroenterol Belg 42:*7–28, 1979
20. Monnier P, Savary M, Pasche R: Apport du bleu de toluidine en cancérologie bucco- pharyngo-oesophagienne. *Acta Endoscopy 11:*299–315, 1981
21. Fékété F, Breil P, Ronsse H, Langonnet F: EEA stapler and omental graft in esophagogastrectomy. *Ann Surg 193:*825–830, 1981