PRINCIPLES AND PRACTICE OF SURGICAL PATHOLOGY

VOLUME 2

Edited by

Steven G. Silverberg, M.D.

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Preface

Why another textbook of surgical pathology? The raison d'être of this project is twofold: first, to provide a thorough and authoritative survey of the field; second, to present information in a practical format that is useful to the pathologist in training or practice.

To accomplish the first goal, the authors chosen represent an international array of experts, most of whom have written recent monographs or major review articles on the subjects that they have covered here. The subject assigned to each author or team of authors is narrow enough to be covered in depth. Toward the second goal, most chapters include introductory sections on specimen handling, clinical correlations, and normal gross and microscopic anatomy. The chapters are in part specimen-oriented rather than disease-oriented: thus, for example, lesions likely to be encountered as medical biopsy specimens of lung, liver, and kidney are discussed in separate chapters from those encountered primarily in surgical resection specimens of the same organs. Organ site chapters, with the exception of Chapters 24 and 26, are arranged so that Volume 2 contains all topographic sites from SNOMED code 56000 (SNOP code 56) on, and Volume 1 contains sites with lower code numbers. Finally, common problems, particularly of differential diagnosis, are emphasized over rare ones.

Several chapters at the beginning of the first volume are provided as introductory material for the student of surgical pathology. These include discussions of some problems (e.g., infections and metastatic tumors) that are not unique to any one organ or organ system and are thus applicable to many of the specific site chapters. I recommend that these chapters be read once in their entirety rather than merely referred to when a specific problem arises in the laboratory.

The main credit for this work must go to the individual authors, many of whom must have regretted ever having agreed to work with such an unreasonable and arbitrary editor as myself. It is a tribute to the development of the field of surgical pathology that there are so many pathologists expert in so many fields. Only a few years ago, the majority of authors of a work of this sort would have been clinicians who doubled as pathologists in their own limited fields but had no broad background in general pathology.

My thanks also go to a few people who provided special assistance with respect to the conceptual development and physical production of the entire work. Drs. Richard J. Reed, William J. Frable, Robert W. McDivitt, Andrew G. Huvos, and Richard J. Kempson served as editorial consultants and provided valuable suggestions in the planning phase. John de Carville, Robert Hurley, Scott Klein, and Charles Kyreakou at John Wiley & Sons provided constant encouragement, advice, and hard work from beginning to end. LaVonne King functioned diligently and uncomplainingly as my editorial assistant, and Luann Bergquist typed some of my chapters and pertinent correspondence. Howard Mitchell provided his usual excellent artistic and photographic assistance. Many of my colleagues and especially residents at the University of Colorado offered invaluable criticism during the process of writing and editing and certainly always showed patience and understanding when I was unavailable for other tasks because of an editing deadline.

Finally, two special words of thanks. The inspiration to undertake a task of this magnitude developed over many years largely as a result of my association with many talented surgical pathologists, first as a trainee, later as a colleague, and most recently as a mentor and consultant. I could hardly hope to list all those colleagues whose ability, enthusiasm, and dedication have promoted my own development over the years; but I should certainly single out Drs. Fred Stewart, Frank Foote, Saul Kay, and my good friend and partner in yet another opus, Claude Gompel. The late Drs. Abou Pollack, William Barriss McAllister, Jr., Harry Greene, and Averill Liebow also deserve the thanks that I never conveyed adequately when they were still with me. Ultimately, however, the main burden of an undertaking such as this falls on the loved ones who make personal sacrifices to free time from other activities for the many hours of work involved. For this and all other support, I offer my deepest gratitude to my wife, Kiyoe.

S.G.S.

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The Esophagus

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John Jones Thompson

EMBRYOLOGY

The esophagus is recognizable at about the 2.5-mm stage. In contrast to some reports based on other species, it maintains a lumen throughout development. The earliest epithelial lining consists of two or thee layers of stratified epithelium, which subsequently becomes vacuolated and is then replaced by ciliated epithelium by the 60-mm stage. Ciliated epithelium, in turn, is replaced by mucin-secreting cells. At about the 90-mm stage, stratified squamous epithelium reappears at the midesophagus and proceeds to line the entire organ by the 250-mm stage (approximately a fetal age of 5 months) (1).

ANATOMY AND HISTOLOGY

Surprisingly, there is still debate about important details of the histology and anatomy of the esophagus. All the usual layers of the gastrointestinal tract are represented. The mucosa is composed of stratified squamous epithelium. Keratinization is not a normal feature in humans. A basal zone is present and normally is not more than two or three cells thick or more than 15% of the total mucosal thickness. Mitoses are infrequent and confined to the basal zone. Papillae of the lamina propria extend through about two-thirds of the total mucosal thickness. Lymphocytes and plasma cells are present in small numbers in the lamina propria (2).

Small mucus-secreting glands, similar to those of the gastric cardia and referred to as cardiac glands, occur in the lamina propria (Fig. 1). These communicate to the surface via a channel lined by a layer of mucus-secreting cells, which also may replace small areas of stratified squamous epithelium at these foci. They are inconstant, most common in the distal esophagus and less so in the upper esophagus (3).

Ham (4) states that there are only a few scattered glands in the submucosa. However, a study by Umlas and Sakhuja (5) of three normal esophagi sectioned transversely at intervals of 0.5 cm showed an average of four submucosal glands in each level (Fig. 2). These glands may occasionally show serous cells as well as the usual mucinous cells, thus resembling certain salivary glands (3).

The muscularis propria is of striated type in the upper third, smooth muscle in the lower third, and of mixed type in the middle third. Innervation is by parasympathetic fibers from the vagus and from the sympathetic trunks and coeliac ganglia. The usual myenteric ganglionic plexi are present (4).

The arterial supply is multiple, consisting of branches of the inferior thyroid, aorta, intercostals, inferior phrenic, and left gastric arteries. Veins accompany the arteries and form a plexus communicating with the gastric coronary and azygous veins. This creates a connection between the portal and systemic venous systems—the well-known reason for esophageal varicosities in portal hypertension.

Lymphatics

The esophagus contains a rich plexus of mucosal lymphatics connecting with a less-rich submucosal plexus that, in turn, communicates with widely spaced longitudinal channels in the muscle coats. Lymph (and tumor) spreads more freely in a longitudinal direction than circumferentially. There is normally little connection between mucosal lymphatics of the esophagus and stomach. In general, lymph from the esophagus below the tracheal bifurcation passes to the para-aortic nodes or the celiac axis region. Lymph from above this point drains upward, in turn, to paraesophageal nodes, upper paratracheal nodes, and finally to nodes in the region of the inferior thyroid artery (6).

Physiology

The esophagus is said to begin with the cricopharyngeus muscle, which connects anteriorly with the cricoid cartilage and spreads in a fan-shaped fashion to surround the upper esophagus. It variably contributes to the outer longitudinal fibers of the esophagus (7) and constitutes the upper sphincter. While a lower physiologic sphincter is agreed on, its anatomic counterpart is much debated (4,8,9). The gastroesophageal junction

Figure 1. Esophageal cardiac glands. Note the presence of the mucus-secreting glands in the lamina propria and their connection to the surface. Small areas of the squamous epithelium on the surface may be replaced by mucous cells from the duct orifice.

perhaps should be defined physiologically (8), since it does not necessarily exactly coincide with the squamocolumnar junction.

The upper and lower sphincters are normally closed. Swallowing consists of a coordinated opening of the upper sphincter, a rapid distally moving ring of contraction beginning in the upper esophagus, and relaxation of the lower sphincter (10). Pope (10) states that the lowest segment (sphincteric muscle) is selectively affected by various drugs and hormones. There is normally a distinct manometric pressure differential between the distal esophagus and stomach.

HETEROTOPIAS

Foci of columnar mucin-secreting cells have been reported (11) in 6.3% of esophagi of infants and children at autopsy. These foci may represent inadequate squamous replacement rather than true heterotopia. In the same study, foci of gastric fundal-type of epithelium with parietal cells were about half as common. These were distributed in the upper two-thirds of the esophagus and often accompanied by an inflammatory infiltrate, suggesting the possibility that they might in some circumstances be responsible for esophageal peptic ulceration in the upper esophagus. How frequently these foci of glandular mucosal epithelium persist into adult life is not clear.

ESOPHAGEAL AND GASTROENTERIC CYSTS

Cysts of the esophagus and paraesophagus are rare lesions. Nonetheless, cysts (type unspecified) are said to be the second most common benign "tumor" in the Mayo Clinic experience (12). They may be divided into retention cysts and developmental cysts. Within the latter group, gastroenteric (gastroenterogenic) cysts are best separated from other cysts more closely akin to bronchogenic cysts.

Mucoceles or retention cysts represent dilatations of the ducts of submucosal glands. They are usually small and more common in the distal esophagus (3). Developmental cysts (13–18) are discussed in detail in Chapter 24.

ATRESIA, STENOSIS, AND TRACHEOESOPHAGEAL FISTULAS

Atresia of a portion of the esophagus, with or without a tracheoesophageal fistula, and a related phenomenon, congenital stenosis, are well-known pediatric and neonatal problems. Fistula may also occur independently of atresia or stenosis. In the most common situation (90%–95% of the cases), the proximal esophagus ends in a blind pouch, and a tracheoesophageal fistula connects with the lower segment of the esophagus. In addition to regurgitation and signs of aspiration, children with this type show contained air in the stomach and intestine. Excellent surveys of these anomalies are provided by Kissane (18) and (more clinically oriented) by Girdany (19).

RINGS AND WEBS

The terms *rings* and *webs* have been used more or less interchangeably for annular membranes with usually eccentric lumina composed of mucosa and lamina propria. Their pathogenesis is obscure for the most part, although a few are clearly postinflammatory adhesions, such as those reported in benign mucous membrane pemphigus (20,21). The most common site clinically recognized is the upper esophagus, and nearly half of these cases were in women with Plummer-Vinson syndrome. Associated carcinoma is frequently observed in the buccal area or esophagus (21). The histology is that of a fold of mucosa with a mild chronic inflammatory infiltrate in the lamina propria.

They may also occur in the mid- and lower esophagus. Those lesions at the squamocolumnar junction are referred to as Schatski rings. These lower esophageal rings (webs) are surfaced on one side with stratified squamous epithelium and on the other

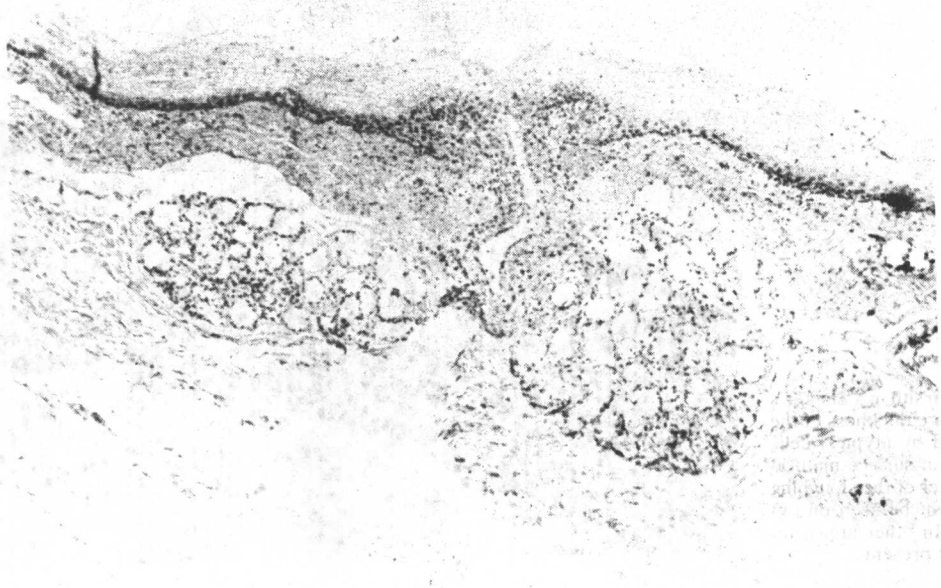

Figure 2. Esophageal submucosal glands. There are several glands seen in the submucosa that are connected to the surface by a central duct. The gland cells are largely mucinous.

with columnar cells. They lack significant inflammatory changes in this site (22,23). Goyal and coworkers (22,23) state that a lower (junctional) ring can be demonstrated at autopsy in 10% of the population when properly searched for and may be the most common cause of esophageal dysphagia. They recognize a less common muscular ring, always proximal to the site of the mucosal ring when both are present. Their origin is debatable.

Annular fibrous strictures of the lower esophagus are probably secondary to esophagitis (22,23).

ACHALASIA, CHALASIA, AND ASSOCIATED CONDITIONS

As indicated in the section on physiology, the smooth coordination of the sphincteric and peristaltic function of the esophagus is important. A number of conditions are caused or complicated by hyperfunction, hypofunction, and incoordination of esophageal muscular activity. These include diffuse spasm of the esophagus (hypertonic esophagus), achalasia, chalasia, Chagas' disease, and esophageal diverticula.

Diffuse Spasm of the Esophagus (Hypertonic Esophagus)

With increased use of manometric studies, a group of patients has been found who have simultaneous, repetitive, and nonperistaltic esophageal contractions that may be abnormally powerful. Although such contractions may be associated with various forms of organic disease, the term *diffuse spasm of the esophagus* or *hypertonic esophagus* has been used when no obvious underlying cause is found. The importance of these uncoordinated, nonperistaltic high-pressure spikes is their ability to mimic anginal pain. These tertiary contractions may give the esophagus a curled or corkscrew appearance on radiologic study.

Achalasia

Achalasia is defined as "failure of relaxation of smooth muscle fibers at any junction of one part of the gastrointestinal tract with another." Because the upper esophageal sphincter (cricopharyngeus) is largely composed of striated muscle fibers, the term should be confined to failure of relaxation of the lower sphincter. Unfortunately, the term has also been used to apply to the cricopharyngeus.

Unless otherwise stated, achalasia refers to a high resting lower sphincter pressure when measured manometrically and a failure of adequate sphincter relaxation on swallowing. The proximal segment usually demonstrates lack of coordination and decreased strength of contraction. However, in so-called "vigorous" achalasia, in addition to abnormal sphincter function, the proximal segment may respond in a manner similar to the hypertonic esophagus. The proximal esophagus in the first type usually becomes dilated, lengthens, and eventually resembles a gourd in shape. In the second type, the proximal esophagus may appear normal.

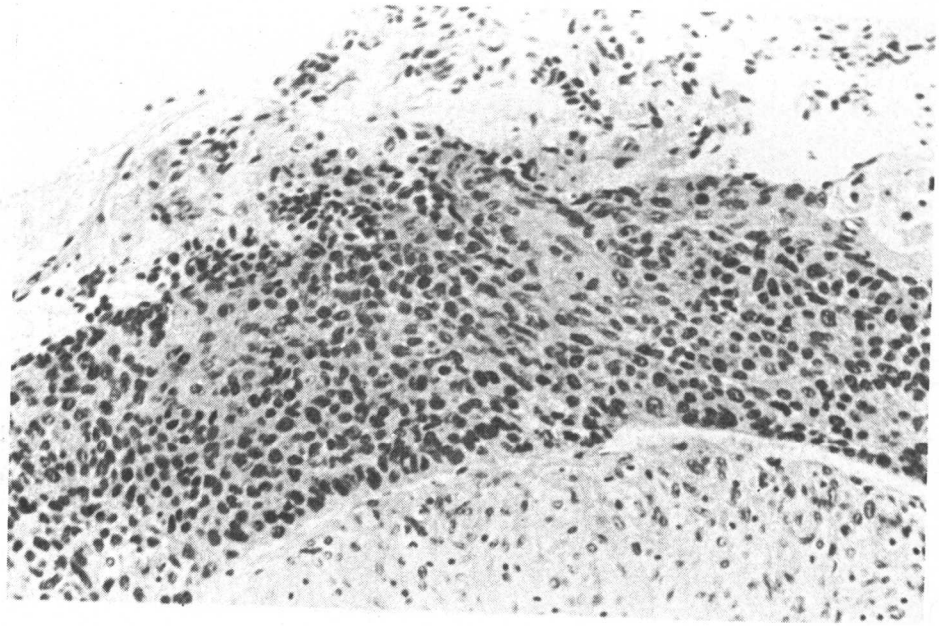

Figure 3. Carcinoma in situ in a Zenker's diverticulum. The entire thickness of the epithelium is replaced by atypical cells showing no evidence of surface maturation. Note the character of the sloughing surface cells that might be seen on cytologic examination. In other areas, invasive carcinoma was present.

Although controversial, considerable evidence suggests a neurogenic origin for achalasia. Casella and coworkers (24) state that the circular layer of muscle in the region of the cardia is inconstantly thickened. Above this point, they have found either hypertrophy or thinning of the circular layer. The adjacent mucosa may show evidence of inflammation. They have demonstrated loss or reduction of ganglion cells in the esophageal myenteric plexus in approximately 60% of the cases, with associated wallerian degeneration of vagal fibers, loss of nuclei in the vagal nucleus, and denervation atrophy of smooth muscle cells. In contrast, in 100% of the patients with Chagas' disease of the esophagus, a similar set of pathologic findings are noted, and an associated achalasia is also noted clinically. The trypanosomes responsible for the disease are known to elaborate a neurotoxin that destroys ganglion cells in the myenteric plexus (25). If the vagus nerves are cut in experimental animals, such as the dog or monkey, a few will also develop an achalasialike syndrome (26). Patients with achalasia usually have gradually progressive dysphagia, which is intermittent and is primarily to solid food. As the disease progresses, stasis of food is usual, and overflow into the mouth may occur as the esophagus becomes overfilled.

The incidence of carcinoma in these patients is increased.

Chalasia

Chalasia or hypotonicity of the lower physiologic sphincter may be a common temporary phenomenon in infants (27) and occasionally may be accompanied by bouts of aspiration pneumonitis, esophagitis, and stricture. It is probably secondary to a delay in maturation of the autonomic system. Chalasia (incompetence) of the lower sphincter in adults is probably the significant lesion of reflux esophagitis, esophageal peptic ulceration, and Barrett's esophagus (to be discussed) rather than hiatus hernia, which is commonly associated with and blamed for those conditions.

Pressure, Rupture, and Tear of the Esophagus (Boerhaave's Syndrome and Mallory-Weiss Syndrome)

Spontaneous rupture (Boerhaave's Syndrome) or tear (Mallory-Weiss Syndrome) of the esophagus is a well-known condition associated 90% of the time with an episode of vigorous vomiting. Sixty percent of the cases occur during an alcoholic episode, and the others have been described during episodes of coughing, hiccuping, and chest massage (28). The ruptures and partial tears are longitudinal, with most ruptures being supradiaphragmatic and most of the tears of the Mallory-Weiss syndrome being in the gastric cardia. In both, pathogenesis is thought to reflect a rapid rise of intraluminal pressure in the presence of a closed upper sphincter (25,28).

DIVERTICULA

Incoordination of the upper sphincter, that is, the cricopharyngeus, is now accepted as the basic cause of the relatively common Zenker's diverticulum, a posterior outpouching of the uppermost esophagus. The diverticula gradually enlarge and cause food retention and secondary aspiration. An increased incidence of carcinoma is recognized in Zenker's diverticula ranging from 0.31% to 0.7% (29,30), and excised examples

Figure 4. Traction diverticulum of esophagus. A small diverticulum is indicated by the arrow. The lesion is located at the tracheal bifurcation adjacent to the underlying carinal lymph nodes.

should be adequately sectioned to detect malignancy (Fig. 3). Myotomy has caused disappearance of early Zenker's diverticula (31).

Most individuals with Zenker's diverticula are middle-aged, although extremely rare examples in neonates have been reported (32).

Most midesophageal diverticula (33) in the region of the tracheal bifurcation have been reported as "traction" diverticula due to scarring of tubercular lymph nodes or similar conditions (Fig. 4). More recently, in cases with motility disorders, pulsion diverticula in this area as well as in the distal 10 cm of esophagus have come to be recognized (34).

ESOPHAGITIS

Esophagitis with its accompanying symptoms of dysphagia and pain is a very common problem. Its causes are diverse and include prolonged intubation, reflux of gastric contents, infections, alcohol, irradiation, and chemotherapy.

Reflux Esophagitis

Reflux esophagitis implies exposure of esophageal mucosa to gastric secretion products. Minimal reflux esophagtis may be recognized histologically by extension of papillae of the lamina propria almost to the mucosal surface, by thickening of the basal cell zone to greater than the normal 15% of the total mucosal thickness, and by the presence of neutrophils, particularly in the mucosal papillae. Lymphocytes or plasma cells per se in the lamina propria should not be considered evidence of esophagitis (2).

In more severe stages, hyperemia, obvious acute and chronic inflammatory infiltrates, and ulceration are seen (Fig. 5). The last may consist of shallow, linear, or, in more severe cases, confluent ulcerations. In continued reflux, frank peptic ulcers of various depths may develop (Fig. 6). Healing in these latter stages, of course, is accompanied by fibrosis, which may be irregular or take the form of an annular fibrotic "ring" (22,23). It has been demonstrated that re-epithelization may consist of glandular rather than squamous epithelium (see discussion of Barrett's esophagus).

Reflux esophagitis is frequently associated with hiatal hernias, particularly sliding hiatal hernia, and is often ascribed to the presence of the hernia. However, hiatal hernia is very commonly diagnosed by radiologists in patients without symptoms of reflux. It is more logical to assume incompetence of the lower sphincter as more immediately related.

Columnar-Lined Lower Esophagus (Barrett's Esophagus)

In 1950, Barrett (35) recognized a group of cases associated with hiatal hernia that he considered examples of congenital short esophagus with extension of the stomach into the mediastinum. As currently used, *Barrett's esophagus* may be defined as any esophagus in which a variable segment of esophagus above the physiologic sphincter is lined with columnar or glandular epithelium. The condition, with rare exceptions, occurs in adults. This age relationship plus good experimental evidence (36) has led to the current view that the lesion is acquired. In most cases, the condition is associated with hiatal hernia, esophageal peptic ulceration and stricture, and a low pressure differential between stomach and esophagus (37). The pathogenesis would appear to be that of acid reflux leading to ulceration with stricture and healing with glandular re-epithelization. Such changes may extend up to at least 10 cm from the

Figure 5. Reflux esophagitis. The presence of two small ulcers and an irregular white patch of thickened, hyperemic mucosa can be appreciated just above the lower sphincteric muscle.

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