Benign Esophageal Tumors

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KEYWORDS

• Leiomyoma • Gastrointestinal stromal tumor • Mediastinal cyst

KEY POINTS

- Endoscopic evaluation including endoscopic ultrasonography is foundational to the evaluation of benign and indeterminate esophageal pathology.
- Leiomyomas have distinctive distributions, behavior, and entailed therapeutic significance in pediatric patients.
- Immunohistochemical analysis is an important adjunctive diagnostic tool in distinguishing noncarcinomatous tumors of the esophagus.
- Symptomatic lesions and those with rapid change in size dictate surgical management.
- Endoscopic, thoracoscopic, and laparoscopic techniques including enucleation are widely used in the management of benign tumors of the esophagus.

INTRODUCTION

Unlike esophageal carcinoma, benign esophageal tumors and cysts are rare. Multiple autopsy series have been performed in the past, and although the specific results vary, the overall incidence is less than 1%. In addition, benign tumors account for less than 5% of all surgically resected esophageal tumors.[1] Nevertheless, the past century has shown an increasing trend in the incidence of these lesions, most likely a reflection of improving diagnostic methods,[2] and continued advancements in the understanding of their natural history and management. Benign esophageal tumors are often asymptomatic and typically require only close surveillance. If surgery is indicated because of symptoms or diagnostic uncertainty, many of these tumors can be successfully resected with excellent long-term outcomes. Because these lesions are rare, the general or gastrointestinal (GI) surgeon should have a strong foundation in their diagnosis and treatment.

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HISTORY

The first documented record of a benign esophageal tumor was in 1559 by Sussius. The tumor was discovered on autopsy, located in the distal esophagus, and has been cited as a leiomyoma, although histologic confirmation is lacking.[3] In 1763, Dallas-Monro performed one of the first treatments of a benign esophageal tumor when he excised a pedunculated esophageal mass using a snare from a 64-year-old man who had regurgitated the mass into his mouth. The first successful surgical treatment of a benign esophageal tumor is generally credited to Sauerbach, who performed a partial esophagectomy with esophagogastrostomy in 1932 for a myoma, most likely a leiomyoma. One year later, Oshawa performed the first open enucleation of an esophageal leiomyoma, and in 1937, Churchill performed the first open enucleation of a benign esophageal tumor in the United States for what was initially described as a neurofibroma but later reclassified as a leiomyoma.

According to Storey and Adams[4] in their case report and review of leiomyoma of the esophagus, only 16 documented surgical cases were found up until 1948, but between then and time of their publication in 1956, they found an additional 94 cases described, including 4 cases of their own. Since then, there have been many more recorded surgeries for benign esophageal tumors, and within the past 2 decades, there has been a shift toward minimally invasive approaches, specifically via thoracoscopy and endoscopy.

INCIDENCE

Several autopsy series and medical literature reviews have been performed in the past, searching for the true incidence of benign esophageal neoplasms. In 1932, Patterson[5] reported a total of 62 benign esophageal tumors during a 215-year period from 1717 to 1932. In 1944, Moersch[6] found 44 benign tumors and cysts in 7459 autopsy examinations, for an incidence of 0.59%. Plachta[7] in 1962 reviewed 19,982 postmortem examinations and found a total of 505 esophageal neoplasms, 90 of which were benign, resulting in an overall incidence of 0.45% with approximately 18% of all esophageal tumors being benign. In 1968, Attah and Hajdu[8] found 26 benign tumors among 15,454 autopsies during a 30-year period, for an incidence of 0.16%. Allowing for some variation among these studies, the overall incidence is cumulatively documented as less than 1%.[1] By way of comparison, malignant esophageal carcinoma is approximately 50 times more common.[9] The mean age of presentation for benign lesions is between the third and fifth decade of life, much younger than the mean age of presentation for esophageal carcinoma, and studies suggest a slight male predominance with an average ratio of 2:1.[1]

Unlike other benign tumors, esophageal duplications and cysts are more common in children. Accordingly, although such lesions are estimated to comprise only 0.5% to 3.3% of all benign esophageal masses in adults, they account for approximately 12% of all mediastinal tumors in the pediatric population. Between 25% and 35% of all esophageal duplications first become manifest in adults, and of these, most present in adults younger than 50 years.[10]

CLINICAL FEATURES

Benign esophageal tumors are generally slow-growing masses, and they may remain stable without any change in size for many years. At least 50% of benign esophageal masses are asymptomatic,[7] and they are frequently diagnosed incidentally on imaging or endoscopy performed for other reasons.[2] Choong and Meyers[1] broadly categorized