Esophageal Leiomyoma

87

Definition

This is a benign tumor originating from smooth muscle cells of the esophagus. It belongs to the leiomyoma family (see section entitled "Leiomyoma").

Epidemiology and Presentation

Esophageal leiomyoma is a rare tumor but is the most frequent type of benign neoplasm of the esophagus. It most frequently occurs between the ages of 20–50 years, with a higher incidence in males (M:F = 2:1). The most common location is in the lower two thirds of the esophagus (only 10% of cases occur in the upper one third of the esophagus). Most cases are detected when they are less than 5 cm in size; rarely, this neoplasm grows larger than 10 cm (in this case it is called giant leiomyoma of the esophagus). Multiple tumors may be observed. The most common symptoms are dysphagia, chest pain, vague retrosternal discomfort, heartburn, and occasionally regurgitation; gastrointestinal bleeding is rarely observed. In the rare case of a giant leiomyoma of the esophagus, patients report persistent cough. Large esophageal leiomyomas usually grow outward, so dysphagia need not be present and does not reflect the size of such tumor. Many of these tumors are discovered incidentally during endoscopic procedures or radiological tests. With the increasing use of endoscopy and radiological investigation, the number of cases diagnosed is growing.

The typical appearance is a smooth concave space-occupying lesion underlying a normal mucosa. It is easy to recognize a sharp angle at the junction of the tumor and healthy tissue. When an endoscopy is performed, these tumors can be identified as relatively mobile submucosal swellings. An upper GI endoscopy will confirm the presence of a submucosal tumor by clearly visualizing a mass protruding into the lumen of the esophagus, with normal-looking mucosa covering the swelling. A computed tomography scan is a valuable investigation in confirming the diagnosis.

S. Mocellin, *Soft Tissue Tumors*, https://doi.org/10.1007/978-3-030-58710-9_87

Currently, endoscopic ultrasonography (EUS) plays a critical role to diagnose esophageal leiomyoma.

Pathology

The tumor presents as a circumscribed lesions composed of intersecting fascicles of bland spindle cells with abundant cytoplasm. These well-differentiated smooth muscle cells which are of the spindle type are arranged as braids. These bundles are demarcated by adjacent tissue or a definite connective tissue capsule. The spindle cells intersect with each other at varying angles. The tumor cells have blunt elongated nuclei and display minimal atypia and very few mitotic figures.

Biomarkers

Tumor cells stain positive for desmin and alpha-smooth muscle actin, while they stain negative for CD34, CD117 (KIT), and S100.

Prognosis

Esophageal leiomyoma is a benign neoplasm.

Therapy

All patients with symptomatic tumors are advised to undergo excision or enucleation of the tumor. The conventional surgical approach is an open thoracotomy; after this, an enucleation of the tumor with an esophageal myotomy or a resection of the tumor with the esophagus is performed. Surgical esophageal resection may be indicated in giant leiomyoma of the esophagus or tumors involving long segments of the esophagus. Tumors of the middle third can be approached using a right thoracic route, and tumors of the distal one third can be accessed through a left-sided approach. Video-assisted thoracoscopic surgery (VATS) has progressively gained acceptance in the last few years. For extra-mucosal excision or enucleation, the outer esophageal muscle is incised longitudinally, and then careful dissection is performed to separate and remove the leiomyoma from the underlying mucosa. Recently, endoscopic submucosal dissection and enucleation of esophageal leiomyomas are increasingly being performed. The standard surgical practice is to approximate the muscle layer following a myotomy and enucleation, although some investigators believe that even large myotomies or extra-mucosal defects can be left open without the development of a subsequent complication. Esophageal resection as a treatment is reserved for those with very large tumors. Asymptomatic tumors which are less than 1 cm are managed by regular follow-up strategies.

Suggested Readings

Chen (2017) Minimally invasive surgery for giant esophageal leiomyoma: a case report & review of the literatures. J Thorac Dis 9(1):E26–E31

Chen (2019) A novel hybrid approach for enucleation of esophageal leiomyoma. J Thorac Dis 11(6):2576–2580

Donatelli (2017) Submucosal tunneling endoscopic resection (STER) with full-thickness muscle excision for a recurrent para-aortic esophageal leiomyoma after surgery. Endoscopy 49(S 01):E86–E87

Zhu (2019) Successful en bloc endoscopic full-thickness resection of a giant cervical esophageal leiomyoma originating from muscularis propria. J Cardiothorac Surg 14(1):16