was noted to be attached to the esophagus by a broad stalk. There appeared to still be a layer of muscle fibers between the stalk and the mucosa, so division of the stalk with an endo-GIA stapler seemed like a feasible option. The concern in doing this was that if the mucosal tracts did indeed communicate with the mass, then the stapled mucosa that remained would present a risk of leak. Our plan if that were to occur was to ensure satisfactory mucosal closure and to cover the site of resection with an intercostal muscle flap, because a two-layered closure would be compromised by our need to resect a significant portion of the adjacent muscular layer with the specimen. We performed the resection over a lighted 56-Fr bougie to prevent unintentionally entraining mucosa into our staple line and to detect perforation. After resection, intraoperative endoscopy revealed smooth mucosa, with no evidence of disruption or remaining fistuluous tracts, making it likely that these findings were actually traction dimples. Pathologic examination demonstrated pancreatic heterotopia with a negative margin of resection at the staple line.

The patient had an uneventful recovery. A swallow study performed on postoperative day 3 showed no evidence of leak. He was advanced to a soft diet and discharged home. The patient returned for a 2-month follow-up visit and was tolerating a regular diet with no recurrence of epigastric pain.

Comment

Heterotopic pancreas is found in 0.6% to 13.6% of autopsies [1]. It is more common in male individuals and has the highest incidence in the fourth to the sixth decades of life. It is uncommon to find heterotopic pancreas in the esophagus. Only 13 cases of esophageal heterotopic pancreas have been reported. Although the condition is mostly asymptomatic, symptoms associated with esophageal heterotopic pancreas include epigastric pain, dysphagia, and upper gastrointestinal bleeding [2]. The most common site is the distal esophagus. It can be associated with other anomalies. Two cases have been associated with malignancy [3, 4].

Management has varied from observation to more radical surgical procedures such as Ivor-Lewis esophagectomy. Resection by VATS has not previously been described, to our knowledge. Because of the presence of malignancy in previously reported cases, it is prudent to remove the abnormal tissue surgically instead of relying on observation. Diagnosis is often difficult and is usually determined once the specimen is removed in its entirety [5].

In this case, multiple biopsy specimens were nondiagnostic and the imaging studies inconclusive, making preoperative diagnosis impossible. The differential diagnosis included both benign and malignant causes, although the patient’s age argued for a benign cause. Although the lesion looked benign under direct visualization from the left hemithorax, its complete surgical resection was indicated, given the patient’s symptoms, the failure to obtain a tissue diagnosis by less invasive means, and the malignant potential of the lesion.

References


Leiomyoma Presenting as a Massive Calcified Circumferential Esophageal Mass

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Esophageal leiomyoma is the most common benign esophageal neoplasm and often presents as an incidental finding or with nonspecific symptoms such as dysphagia.
or chest pain. Surgical enucleation is the mainstay of treatment and may be accomplished using both open and thoracoscopic approaches. We present a case of a 57-year-old man who presented with a massive circumferential calcified leiomyoma.


Esophageal leiomyoma is the most common benign esophageal tumor but represents only 1% of all esophageal masses [1]. The disease entity was first described by Morgagni in 1761, and close to 2 centuries later Sauerbruch performed the first successful resection of a leiomyoma in 1932 [2]. Generally, these lesions are discovered incidentally or are brought to attention after reaching sufficient size to cause symptoms such as dysphagia. Treatment involves complete surgical resection, which is curative. Recurrence rates are low.

A 57-year-old El Salvadoran man presented for evaluation of a painful flank hernia. On review of systems, the patient additionally reported long-standing dysphagia and a history of endoscopic dilation of an esophageal stricture in his home country. Computed tomography (CT) of his abdomen and pelvis was performed to evaluate the hernia, with an incidental finding of an esophageal mass. He subsequently underwent CT of his chest, which demonstrated a large, coarsely calcified posterior mediastinal mass (10.6 × 6.9 × 5.7 cm) displacing the esophagus to the right and resulting in significant luminal narrowing (Fig 1A). The mass extended from the thoracic inlet nearly to the gastroesophageal junction.

The patient subsequently underwent esophagogastroduodenoscopy, which showed significant external compression of the esophageal lumen over a 12-cm length, with intact overlying esophageal mucosa and a normal squamocolumnar junction. Endoscopic ultrasonography demonstrated a large heterogeneous mass with hyperechoic features (Fig 1B). Endoscopically guided biopsy was nondiagnostic. Because of the large size of the mass and the patient’s symptoms, he was offered surgical resection.

The patient was taken to the operating room and underwent general anesthesia, including intubation with a double-lumen endotracheal tube. He was placed in the left lateral decubitus position, and a right posterolateral thoracotomy was performed. On inspection of the pleural cavity, a large periesophageal mass was identified deep to the muscular layer. A myotomy was performed and carried superiorly and inferiorly, revealing a firm light yellow mass circumferentially encapsulating the esophagus, requiring tedious dissection. After resection, the overlying muscular layer was reapproximated with interrupted 4-0 polypropylene suture and buttressed with a pericardial fat pad flap.

Postoperatively, the patient recovered well. He underwent a fluoroscopic swallow study on postoperative day 7 and was discharged the following day. A year later, he continues to do well with no signs of recurrence and good resolution of symptoms.

Gross pathologic review of the surgical specimen demonstrated a solid tan mass measuring 4.2 × 7.7 × 3.1 cm (Fig 2). Cut surfaces revealed firm white whorled surfaces with multiple areas of focal calcifications. Hematoxylin and eosin staining demonstrated no atypia and only 1 mitotic figure per 10 high-power fields (Fig 3A). Immunohistochemical staining was positive for smooth muscle actin and negative for c-Kit (Fig 3B, 3C), consistent with a leiomyoma.

Comment

Esophageal leiomyoma is the most common benign esophageal tumor but represents only 1% of all esophageal tumors [2]. These tumors most frequently occur in men in the fifth decade of life but have been described in much younger and older patients [2–4]. Half are
asymptomatic, whereas the remainder present with a variety of symptoms, including dysphagia, chest or abdominal pain, dyspepsia, and weight loss [1–4]. Leiomyomas are generally slow-growing tumors and, even in symptomatic patients, diagnosis is often delayed because of nonspecificity or the mild nature of symptoms. Given the distribution of smooth muscle in the esophagus, leiomyomas most commonly occur in the distal and middle thirds of the esophagus [1]. The lesions are usually isolated, with less than 10% of patients having multiple masses [2].

A variety of diagnostic modalities may be used in the initial workup. Barium swallow classically demonstrates a smooth convex mass encroaching into the esophageal lumen. Esophagogastroduodenoscopy should be performed to rule out carcinoma. In contrast to carcinomas, leiomyomas are associated with intact mucosa with a freely mobile submucosal mass. Endoscopic ultrasonography generally reveals a homogeneous, intramural, hypoechoic, and well-demarcated mass. If leiomyoma is suspected, biopsy is unnecessary, especially given the procedural risk of infection, bleeding, or perforation and the fact that biopsy may result in scarring, which increases the difficulty of subsequent extramural resection. In this case, biopsy was performed because of the size, atypical features, and heterogeneity of the patient’s mass. Cross-sectional imaging, either by computed tomography or magnetic resonance imaging, most commonly demonstrates a homogeneous round or oval mass, but calcifications may be observed on computed tomography, as in this patient.

Gross histologic examination of an esophageal leiomyoma demonstrates a firm, rubbery white or tan lesion. These tumors are smooth, round or ovoid, and nearly half are lobulated. Leiomyomas may be distinguished from gastrointestinal stromal tumors (GISTs) on the basis of histologic characteristics. Leiomyomas stain positively for desmin and smooth muscle actin, whereas GISTs do not; additionally, leiomyomas are CD34- and c-Kit (CD117)-negative, whereas GISTs stain positive. Leiomyomas are
composed of smooth muscle fibers bundled with fibrous and neural tissue. They have little cytologic atypia and low mitotic activity. Eosinophilic interlacing spindle cells with elongated cigar-shaped nuclei are observed.

Surgical resection is the treatment of choice and is generally recommended for large or symptomatic tumors. Because of the exceedingly rare incidence of malignancy [2], small asymptomatic leiomyomas may be observed. Resection is most commonly performed by thoracotomy or a video-assisted thoracoscopic approach, although robotic techniques have also been reported, even for complex tumors [5]. Video-assisted thoracoscopic resection offers the advantage of decreased postoperative pain and length of hospital stay [6]. If thoracotomy is indicated, a right-sided approach is used for lesions in the upper two thirds of the esophagus, whereas a left-sided approach is used for more distal lesions. Right thoracotomy was chosen for the patient in this case because of the location, large size, and circumferential nature of the tumor. Enucleation is preferred and is generally feasible; however, mucosal injury is fairly common (12.7%) and is associated with preoperative endoscopic biopsy within 1 month of resection but not with tumor size, location, or operative approach [7]. When injury occurs, the mucosa should be repaired in 2 layers with absorbable suture. Formal esophageal resection is rarely indicated but is more common if the tumor is large (> 8 cm), annular, or densely adherent to surrounding structures [2].

Large series have demonstrated excellent outcomes after resection, with long-term symptomatic relief, low recurrence rates, and low mortality rates [4, 7]. Malignant transformation is exceedingly rare but has been described in large case series [2].

Although the majority of esophageal leiomyomas are smaller homogeneous submucosal tumors, some tumors may become quite large and present as heterogeneous calcified circumferential masses, as in this patient. Careful operative planning and meticulous dissection are critical for optimal patient outcomes.

References


Expansile Kaposiform Hemangioendothelioma Deformed Thoracic Cage in an Adult

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Kaposiform hemangioendothelioma is a vascular tumor that commonly presents as a cutaneous mass, is observed in children, and is associated with Kasabach-Merritt phenomenon. Herein we report a case of kaposiform hemangioendothelioma with chest wall deformity in an adult who did not show the Kasabach-Merritt phenomenon or cutaneous lesions. To our knowledge, this is the first case of asymptomatic kaposiform hemangioendothelioma arising from the pleura and deforming the chest wall. The patient was treated with tumor excision and chest wall reconstruction.


Kaposiform hemangioendothelioma (KHE), first described by Zukerberg and colleagues [1] in 1993, is a rare vascular tumor. Like most vascular tumors, KHE is usually benign or of intermediate malignancy [2]. KHE is a locally aggressive vascular neoplasm that usually develops as an ill-defined, red-to-purple indurated plaque in the skin or deep soft tissue of infants and adolescents [3]. KHE is classically associated with Kasabach-Merritt phenomenon (KMP), which presents with severe thrombocytopenia, consumptive coagulopathy, and microangiopathic anemia [4].

A 21-year-old man presented with a chest wall tumor in our clinic. Right-sided chest wall protrusion and scoliosis had developed when he was 15 years old, and he started to feel mild pain in his right chest at that time. He had not experienced any inciting trauma but described that the pain had worsened with time. On physical examination, protrusion of the ribs with localized tenderness to palpation was noted over the