Transoral Endoscopic Resection of Esophageal Liposarcoma

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A 63-year-old man presented with a 1-month history of dysphagia. Endoscopy showed the patient had a mass in the cervical esophagus. Multiple biopsy specimens were taken, which were inconclusive for a diagnosis. A positron-emission tomography scan showed a hypermetabolic lesion. A transoral endoscopic approach was used to resect the tumor. Final histologic examination revealed a completely resected low-grade liposarcoma.

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The first esophageal liposarcoma was described by Mansour and colleagues in 1983 [1]. Liposarcoma is one of the most common soft tissue sarcomas in adults, affecting soft tissue of the extremities, the trunk, or the retroperitoneum. Liposarcomas are rarely seen in the gastrointestinal tract, especially in the esophagus. Very few cases have been reported, and most of these originated in the cervical esophagus [1–3]. The incidence of gastrointestinal liposarcomas is 0.1% to 5.8% at autopsy. Of those, the esophagus is the least common location affected (1.2% to 1.5% of all gastrointestinal lipomas) [4].

Curative intent management requires complete surgical removal, traditionally achieved through an esophageal resection. In this case report we describe a transoral approach to these rare tumors that may be applied under favorable conditions.

A 63-year-old man with a profound history of severe coronary artery disease, including coronary artery bypass grafting and multiple coronary artery stents, presented with a 1-month duration of dysphagia to solids, without weight loss. Physical examination did not reveal an oral, neck, or abdominal mass.

An esophagoscopy found a subepithelial mass about 20 cm from the incisors. The mass measured 1 × 1.5 cm, was pedunculated with normal-appearing overlying mucosa, and occupied approximately 40% of the esophageal lumen. Biopsy specimens were taken, but analysis was inconclusive and noncontributory. This prompted several additional endoscopies as well as an endoscopic ultrasound, which showed a subepithelial homogenous hypoechoic lesion, 2 to 3 cm in length and measuring 1 × 1.5 cm. The mass appeared solid and seemed to rise from the muscularis mucosa, without invading the muscularis propria. Analysis of additional biopsy specimens failed to reveal a definitive diagnosis.

Additional imaging was performed, including a computed tomography and positron-emission tomography–computed tomography scan, which revealed a hypermetabolic focus involving a short segment of the proximal esophagus of 2.5 cm at a standardized uptake value of 4. There was no evidence of regional or distant metastasis.

The patient was presented to the multidisciplinary upper gastrointestinal cancer tumor board with a suspected, but not confirmed, cervical esophageal malignancy. The recommendation was up front resection.

Before a planned transcervical esophagotomy and resection of the mass, a transoral approach was entertained. Under general endotracheal anesthesia, an anterior commissure direct laryngoscope was used to visualize the hypopharynx and proximal esophagus. A flexible gastroscope was advanced into the esophagus, and the pedunculated mass was withdrawn into oropharynx, still tethered on a narrow stalk to the cervical esophageal mucosa. A stabilizing retraction suture was placed with an endoscopic suturing device, and the stalk was divided with ultrasonic shears, releasing the tumor (Fig 1).

The patient's postoperative course was uneventful, and he was discharged on the first postoperative day on a regular diet. At the 6-month follow-up, the patient remained disease-free.

Pathologic Findings

Grossly, the mass consisted of a submucosal pedunculated lesion measuring 4 × 2 × 2 cm, covered by normal squamous mucosa. The cut section showed a fatty tumor away from the surgical margins (Fig 2A). The polypoid mass was covered by an unremarkable esophageal squamous mucosa and was situated in submucosa. It was multilobulated and showed readily recognizable lipogenic component.

The main histologic evaluation of the tumor showed proliferation of immature fat cells in the form of lipoblasts of various sizes, embedded in a collagenous stroma. Multinucleated hyperchromatic stromal cells (Floret cell) are pathognomonic to the diagnosis of well-malignant liposarcoma.

Fig 1. Photographs show the (A) esophageal mass resection process and (B) the mass after resection.
differentiated liposarcoma (Fig 2B). The characteristic molecular amplification of \textit{MDM2} and \textit{CDK4} genes on chromosome 12q13-15 in well-differentiated liposarcoma can now be detected with commercially available immunostains. This slide demonstrates strong nuclear expression to MDM2 immunostain, a further confirmation to the diagnosis.

Comment
The most common malignant tumor of the proximal esophagus is squamous cell carcinoma, and mesenchymal tumors account for less than 0.5% of primary malignant esophageal tumors [5]. Liposarcoma is one of the most common soft tissue sarcomas in adults, affecting soft tissue of the extremities, trunk, or retroperitoneum, but is rarely seen in the gastrointestinal tract, especially in the esophagus. Only a few cases have been reported, and most of them originated in the esophagus [1–3]. The incidence of gastrointestinal liposarcomas is 0.1% to 5.8% at autopsy, and the esophagus is the least common location (1.2% to 1.5% of all gastrointestinal lipomas) [4].

The esophageal liposarcomas reported to date have been pedunculated, large, and intraluminal tumors, some tumors have been reported to be transmural masses instead of a polyp [2]. The lesion usually grows until it reaches a considerable size and causes symptoms. Three main variants of liposarcoma have been described, with the well-differentiated variant being the most common, comprising 68% of all lesions reported [6], and to a lesser degree, the myxoid and the pleomorphic type [2].

Depending on the size of the mass, symptoms range from dysphagia to solids or to solids and liquids, foreign body sensation, and in some cases, the mass was protruding through the mouth after an episode of vomiting [7]. Imaging studies show an esophageal mass, but the attachment of the stalk is not identified. Although usually inconclusive, computed tomography and magnetic resonance imaging may add some information about soft tissue densities. Liposarcoma is usually diagnosed from the histologic examination. When applicable, newly available specific antibodies can further confirm the diagnosis, as in this case of the well-differentiated variant.

As with most sarcomas, curative intent treatment requires complete surgical resection [2]. This usually requires resection of the esophagus, although transcervical approaches have been described [7]. In this case, we have presented the utility of a transoral endoscopic approach that can be used for selected esophageal liposarcoma tumors with certain favorable features, such as proximal location, pedunculated morphology, and small size.

References