Composite Diffuse Large B-Cell Lymphoma and Peripheral T-Cell Lymphoma Presenting as an Esophageal Perforation: A Case Report

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BACKGROUND
Primary lymphoma of the upper aerodigestive tract is rare. Patients present with nonspecific complaints of dysphagia, odynophagia, dysphonia, and weight loss. Tracheoesophageal fistula (TEF) can be among the initial findings. Histologic proof of lymphoma can be difficult to obtain, resulting in delayed diagnosis and treatment. We present a case that presented with a TEF, and who was ultimately diagnosed with composite B and T cell lymphoma. It illustrates the diagnostic challenges and management issues encountered in this patient’s treatment.

CASE REPORT
A previously healthy 42-year old male presented with a 3-month history of dysphagia, shortness of breath and 40-pound weight loss. Past medical history was remarkable for Hodgkin disease treated 22 years prior with radiation. He was transferred to our institution with left vocal cord paralysis and a Computed Tomography (CT) scan concerning for a ruptured esophageal diverticulum (Fig. 1-2).

Work-up and Diagnosis
The patient was taken to the operating room (OR) for direct laryngoscopy (DL) and esophagoscopy with biopsies. DL and bronchoscopy were normal. Esophagoscopy revealed a large pouch with foul-smelling contents. The esophageal introitus could not be identified. Biopsies returned as “fibropurulent exudate and granulation tissue, no evidence of malignancy.” The patient was made NPO and antibiotics were initiated. The patient continued to be febrile with leukocytosis. The left neck was opened, a drain was placed, and local wound care was initiated. The patient developed progressive respiratory issues. He returned to the OR for tracheotomy and gastrostomy tube placement. Retrograde esophagoscopy revealed the drain in the lumen of a dilated esophagus. Repeat biopsies showed only inflammatory tissue. Several days later, the patient was noted to be refluxing gastric contents into his wound. The gastrostomy was converted to a jejunostomy and discharged with home health for wound care and antibiotics.

Repeat CT 1 month later showed persistent air and fluid surrounding the larynx with interval development of a defect in the subglottic trachea (Fig. 3). Transnasal esophagoscopy revealed marked laryngeal edema, ulceration and near complete laryngeal stenosis. The esophageal inlet was obliterated. At this point, the patient had non-functional larynx and swallow and multiple non-diagnostic biopsies. A PET scan showed multiple areas of increased metabolic activity, suggestive of lymphoma. After consultation with the Oncology service, the decision was made to proceed with laryngectomy to obtain tissue diagnosis.

Operative findings included marked inflammatory tissue of the soft tissue of the neck, an obliterated proximal esophagus, and multiple defects of the tracheal wall. Intra-operative pathology consultation showed only marked inflammation and necrosis. Reconstructive options after would normally include pectoralis flap or tubed free flap. The concern was that, in the presence of the extensive and persistent soft tissue involvement by this process, these options would ultimately fail. The decision was made to temporize the pharyngoesophageal defect with a salivary bypass stent and thick AlloDerm (Fig. 4), with plans for intensive post-operative monitoring and delayed definitive reconstruction.

DISCUSSION
Primary lymphoma of the esophagus and trachea is rare, with approximately 90 cases reported in the literature. Extranelar involvement by lymphoma typically involves skin, bone, soft tissue, lungs, and liver. Patients with primary aerodigestive lymphoma typically present with nonspecific symptoms, including dysphagia, odynophagia, dysphonia, and weight loss. Tissue diagnosis can be elusive. In one review of primary esophageal lymphoma, all but one biopsy on initial exam was negative for malignancy, showing only chronic inflammation. This is attributed in part to the fact that esophageal lymphomas arise in the submucosal lymphoid patches and grow toward the outer wall rather than the inner lumen. This makes accurate biopsy difficult and may explain increased the risk of fistula formation after the initiation of therapy. TEF may be the first indication of the underlying disease process.

Management of malignant TEF is challenging. Silastic esophageal stents can alleviate obstruction and bypass fistulas. AlloDerm has been investigated as a tissue scaffold for patch esophagoplasty. For more substantial lesions, options include gastric pull-up and pectoralis major flaps. Recently, free tissue transfer with a tubed fasciocutaneous or jejunal flap has become the mainstay of cervical esophageal reconstruction. The radial forearm free flap is frequently used for partial or circumferential repair of defects <5cm. This patient’s tumor created a 4 cm circumferential defect. While free flap would typically be considered for a defect of this size, there was no suitable tissue to which a reconstructive flap could be sewn. The defect was temporized using tubed AlloDerm surrounding a salivary stent, with plans for definitive reconstruction after treatment.

Several wound issues evolved as the patient progressed through his therapy and awaited reconstruction. Dissolution of the tumor resulted in a large cervical defect involving the proximal trachea and cervical esophagus. The silastic tube placed at the time of surgery effectively served to span the absent esophagus, diverting saliva to the distal segment. In anticipation of respiratory difficulties, plans were made with the critical care team for fiberoptic intubation though the wound. This was necessary to avoid unintentional esophageal intubation through the defect in the party wall and successful placement of the tube into the distal carina or right mainstem bronchus. This plan was successfully enacted twice when the patient had bleeding events. Close coordination between the Otolaryngology, Critical Care and Oncology Services was key to managing these issues.

CONCLUSION
While primary lymphoma of the esophagus is uncommon, it should be kept in the differential diagnosis, especially in the setting of an otherwise unexplained TEF. A definitive diagnosis may be difficult to obtain and extensive biopsies may be required. This report highlights one of the major pitfalls of treating aerodigestive lymphoma. While the tumor may respond to chemotherapy, as tumor bulk is depleted, the integrity of vital structures can be compromised, resulting in complex wound issues.

REFERENCES