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Case Report

Primary lymphoma of the entire esophagus diagnosed by endoscopic ultrasound-guided fine needle aspiration (EUS-FNA) ☆

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ABSTRACT

Primary esophageal lymphoma is extremely rare, with fewer than 30 cases reported in the literature. Presentation is nonspecific with multiple radiological and endoscopic appearances, posing its diagnosis a challenge. We report a case of a primary esophageal lymphoma diagnosed by endoscopic ultrasound-fine needle aspiration in a 68-year-old woman referred to our hospital for evaluation of a submucosal tumor spreading all over the esophageal wall. We describe its clinical and imaging features and stand out the importance of having a specific preoperative diagnosis in order to avoid a major surgery.

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Introduction

Esophageal involvement in lymphomas usually results from metastasis from cervical or mediastinal lymph nodes, or extension from gastric lymphoma. Primary esophageal lymphoma is an extremely rare condition, accounting for <1% of all gastrointestinal (GI) lymphomas. [1] Radiological and endoscopic findings show several morphologic characteristics and are nonspecific, posing diagnostic challenges [2,3], and many

of these cases are finally diagnosed surgically. We report a case of an esophageal non-Hodgkin lymphoma involving the whole length of the esophagus.

Case report

A 68-year-old woman was referred to our institution for diagnosis and treatment of an extensive esophageal submucosal

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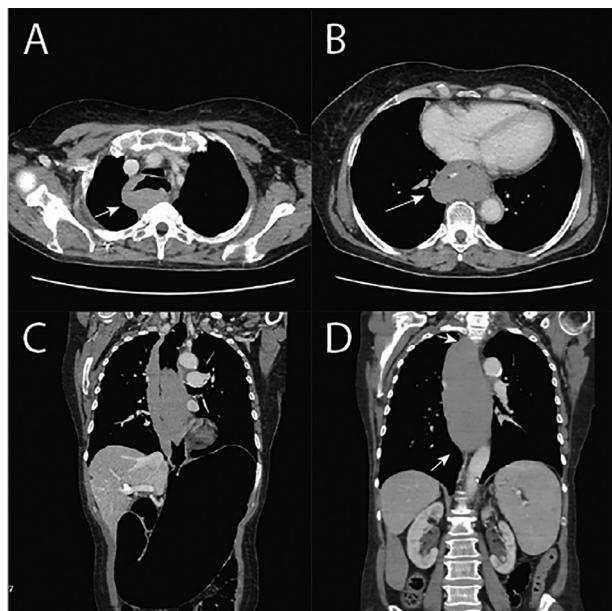


Fig. 1 – PneumoCT showing a lesion in the esophageal wall affecting from the proximal to the distal third, with parietal maximum diameter of 70 mm, producing severe luminal narrowing. No mediastinal lymphadenopathy were observed. (A, B) Axial portal venous phase showing the thickening of the esophageal wall (white arrow). (C, D) Coronal portal venous phase demonstrating the full extension of the lesion (between two arrows D)

lesion. She referred progressive dysphagia and weight loss in the last three months. She had no evidence of any immunosuppressive disease, laboratory findings and physical examination were unremarkable, except for a slightly elevation of lactate dehydrogenase at 250 UI/L.

Computerized pneumotography (pneumoCT) was performed. PneumoCT is a technique used for the diagnosis of lesions of the esophagus, esophagogastric junction, and stomach. Consists of the introduction of a transnasal or transoral foley catheter below the cricopharyngeal muscle with the objective of achieving good distention of the esophageal and gastric lumen. This is done with continuous and sustained insufflation with a CO₂ pump. The distention of the lumen allows best visualization of areas of parietal thickening of the esophageal and gastric wall. PneumoCT showed a lesion in the esophageal wall affecting from proximal to distal third, with parietal maximum diameter of 70 mm, producing severe luminal narrowing. No mediastinal lymphadenopathy were observed (Fig. 1).

Upper gastrointestinal endoscopy revealed a large soft, elastic mass protruding into the lumen with an intact mucosa, extending from 18cm from incisor teeth to 41cm without compromise of gastroesophageal junction (Fig. 2A). Bite-on-bite biopsies (repeated forceps bites in the same spot for unroofing the submucosal lesion) showed normal squamous epithelium. Gastric biopsies showed normal oxyntic mucosa (helicobacter pylori-negative). Endoscopic ultrasound (EUS) performed with a curvilinear scope (Pentax 3870UTK), showed a thickening of

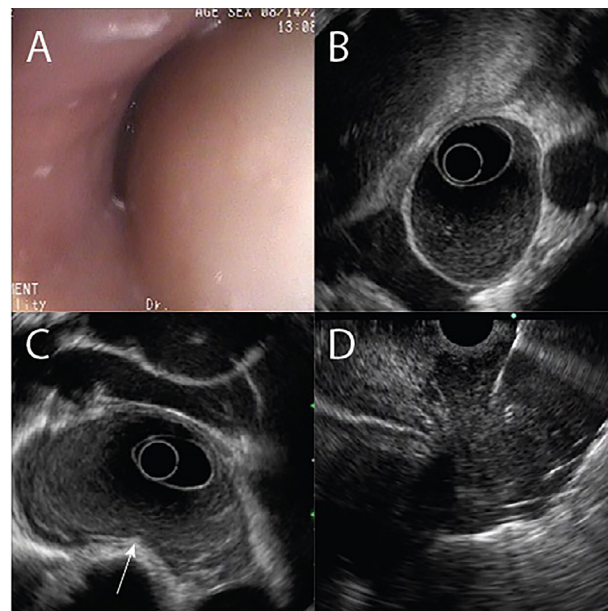


Fig. 2 – Esophagogastroduodenoscopy and Endoscopic ultrasound with Fine needle aspiration (EUS-FNA). (A) Upper gastrointestinal endoscopy showing a large mass protruding into the lumen with an intact mucosa, extending from 18 cm from incisor teeth to 41 cm just above the gastroesophageal junction. (B,C) EUS demonstrating thickening of the whole esophageal wall, with net limits and loss of normal wall layer pattern. This thickening was accentuated in the middle third (arrow) were some microcalcifications were seen. (D) Fine needle aspiration (FNA) with 19G-needle was performed.

the whole esophageal wall, with net limits and loss of normal wall layer pattern. This thickening was accentuated in the middle third were some hiperecoic strands compatible with microcalcifications were seen (Fig. 2 B, C). Fine needle aspiration (FNA) was performed with a 19-gauge(G) needle (Fig D). Once targeted the lesion, the needle-catheter assembly was passed through the working channel of the echoendoscope until the system handle locked in at the end of the biopsy channel. The needle was then advanced under real-time EUS guidance into the middle part of the target lesion. 19G needle can be used to acquire tissue samples for histological examination as described by Yasuda et al. [4] Histopathological examination showed a dense infiltration by a low-grade lymphoma composed of small mature B-cells, admixed with scattered large B cells and mature accompanying T-cells. Neoplastic cells showed positivity for CD20 and Bcl2, whereas CD5, CD23, CD10, cyclin D1, and Bcl6 were negative. Ki67 proliferation index showed a positivity rate of 10% (Fig. 3). Bcl2 rearrangement was negative using FISH break apart approach. With this techniques mantle cell lymphoma, follicular lymphoma and small lymphocytic lymphoma were discarded.

Final diagnosis was a low-grade B-cell lymphoma, favor MALT lymphoma. She started chemotherapy with 6 sessions of cyclophosphamide-vincristine-mabthera and prednisone

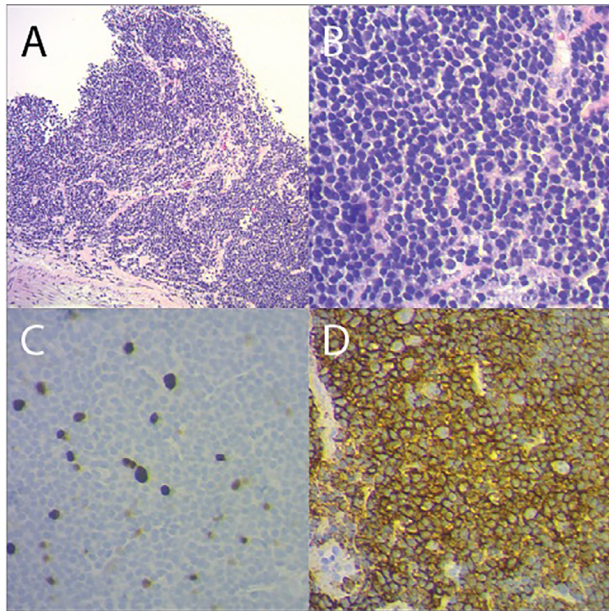


Fig. 3 – Microscopic examination. (A, B) Hematoxylin and eosin (H&E). The papilla is filled by dense infiltrate of small, monomorphic lymphocytes. (C) Ki67. Very scattered positive cells. (D) Intense positivity with B cell markers.

(R-CVP). Her symptoms of dysphagia improved significantly after initiation of chemotherapy.

Discussion

The GI tract is the most commonly involved extranodal site in non-Hodgkin's lymphoma. [1] Primary GI lymphoma, however, is rare, and the esophagus is an extremely rarely involved site, with less than 30 cases in the literature [2,3,5,6]. They are predominantly B-cell type, with few cases of mucosa-associated lymphoid tissue (MALT) type described, and only one diagnosed by EUS-FNA [7,8]. Imaging findings in esophageal lymphoma are unspecific, and vary greatly, including polypoid masses, strictures, ulcers, thickening of the mucosal folds, submucosal nodules, among others [2,3]. Given such a diverse spectrum of presentations, histological assessment is mandatory to confirm diagnosis. In our patient we firstly performed endoscopic biopsy of the lesion, but accordingly to the literature, endoscopic and bite-on-bite biopsy in submucosal infiltrating tumors have a very low diagnostic yield. With the clinical application of EUS, great progress has been made in the diagnostic accuracy of structural abnormalities and depth of invasion in various GI diseases, including lymphoma [9]. EUS provides more accurate information on the involvement of the wall layers. EUS-guided FNA is a safe and effective method in the diagnosis of esophageal and mediastinal pathologies [10]. Moreover, in the last years, with advances in immunology and cytogenetic techniques, FNA biopsies have been excellent samples providing a high diagnostic yield in GI lymphomas

[11]. In conclusion, although a rare disease, it is important for the gastroenterologist to be mindful of esophageal lymphoma and recognize the different patterns of GI involvement in order to make a precise preoperative diagnosis avoiding unnecessary surgical approaches.

Author Contributions

Ines C. Oría: writing original draft. Juan E. Pizzala: writing original draft. Augusto M. Villaverde: Investigation. Dalila C. Urgiles: Visualization. Fernando G. Wright: writing original draft. Federico Jauk: Investigation. Dana Kohan: Supervision. Mariano M. Marcolongo: Supervision.

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