

Isolated Primary non-Hodgkin's Lymphoma of the Esophagus

Abstract

Isolated primary esophageal lymphoma, defined as a lymphoma developing in the esophageal wall, is a distinctly rare presentation and accounts for <1% of all gastrointestinal lymphomas and <0.1% of all malignant lymphomas. The clinical and biological characteristics of the disease are not well known due to its rarity, compounded with a low index of suspicion during diagnosis. We report a rare case of an immunocompetent patient diagnosed with an isolated primary esophageal non-Hodgkin's lymphoma of diffuse large B-cell type. Clinicoradiological and histopathological features, treatment, and outcome, along with a brief review of the literature, are discussed herewith.

Keywords: *Esophageal neoplasm, lymphoma, non-Hodgkin's*

Introduction

Non-Hodgkin's lymphoma (NHL) is most commonly a disease primarily involving lymph nodes; however, it has a well-known tendency to involve extranodal organs as its primary site. The gastrointestinal (GI) tract is the most common extranodal site of NHL, accounting for 5%–20% of all cases,^[1] with the stomach affected in approximately 50%, the small bowel or ileocecal valve in approximately 33%, and the large bowel in approximately 10%. Esophageal lymphoma accounts for <1% of all GI lymphomas and is usually secondary to local extension from adjacent structures, such as mediastinal lymph nodes or from a gastric lymphoma. Primary esophageal lymphoma in an immune competent patient is very rare.^[2] In a study of 79 cases of isolated lymphomas of the GI tract in HIV-seronegative patients, sites of involvement were the stomach (55%), small intestine (31%), large intestine (11%), and esophagus (1%). Lymphomas account for <1% of all malignant tumors of the esophagus.^[3] We report a case of histopathologically confirmed primary esophageal NHL of diffuse large B-cell type (DLBCL), describe its clinicoradiological features, and review the literature.

Case Report

A 60-year-old man presented to us with recent onset progressive dysphagia

for solids over the past 6 months. There was no history suggestive of gastroesophageal reflux or esophagitis before initiation of his present symptoms. His physical examination was normal without enlarged lymph nodes or palpable hepatosplenomegaly. Routine hematological and biochemical investigations were normal. Esophagogastroduodenoscopy showed a circumferential ulceroproliferative friable tumor from 30 to 35 cm with luminal narrowing. The remaining part of the esophagus, stomach, and duodenum were normal. The biopsy showed malignant round cells arranged in a diffuse pattern, suggestive of a NHL [Figure 1a and b]. This was further confirmed by immunohistochemistry which showed diffuse positivity of tumor cells for leukocyte common antigen [Figure 1c] and CD20 [Figure 1d] and negativity for CD3 antigen. Mib-1/Ki-67 (proliferation marker) labeling index highlighted 98% tumor cell nuclei. Cytokeratin, epithelial membrane antigen, and CD138 were negative. Positron emission tomography (PET) scan [Figure 2a and b] revealed 18-F fluorodeoxyglucose-avid circumferential wall thickening of the mid and lower third of the esophagus for a segment of 10 cm with hypermetabolic pretracheal, precarinal, and bilateral hilar lymph nodes. A bone marrow biopsy was done which did not show involvement by NHL. Based on a final diagnosis of isolated esophageal NHL of DLBCL type, and the multidisciplinary

**Pavan Thulasi Sugoor,
Sabita Jiwnani¹,
Bharat Rekhi²,
George Karimundackal¹,
Nilendu Purandare³,
C S Pramesh¹**

Departments of Surgical Oncology, ¹Thoracic Oncology, ²Pathology and ³Nuclear Medicine, Tata Memorial Centre, Mumbai, Maharashtra, India

Address for correspondence:

*Dr. C S Pramesh,
Department of Surgical Oncology, Division of Thoracic Surgery, Tata Memorial Centre, Parel, Mumbai - 400 012, Maharashtra, India.
E-mail: prameshcs@tmc.gov.in*

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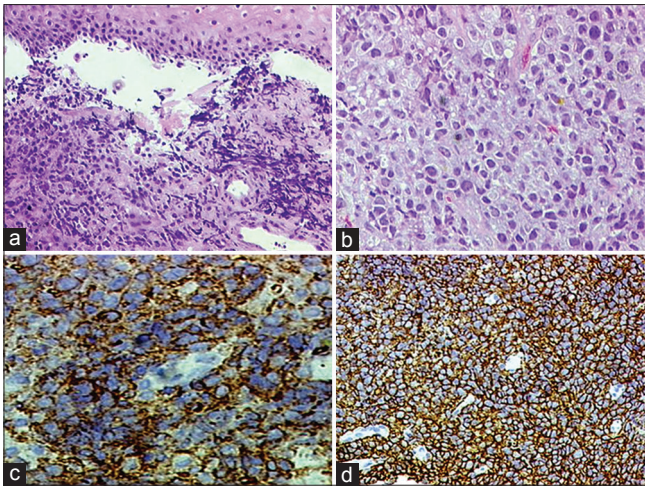


Figure 1: (a) Tumor underlying esophageal squamous mucosa. (b) Malignant tumor composed of round cells arranged in a diffuse pattern. (c) Tumor cells diffusely expressing leukocyte common antigen. (d) Diffuse CD20 positivity within tumor cells

tumor board decision, we treated the patient with chemoradiation with six cycles of chemotherapy, including cyclophosphamide, etoposide, vincristine, and prednisolone, along with involved field radiotherapy) in view of the bulky primary disease. Posttreatment response assessment PET [Figure 2c and d] revealed complete metabolic and morphological response of the primary and aortopulmonary lymph nodes. Endoscopic mapping after completion of treatment showed only posttreatment changes in the form of mucosal tags with scarring and pseudodiverticula. The patient is symptom and disease free on a 3-year follow-up.

Discussion

Epithelial tumors, namely squamous cell carcinoma and adenocarcinoma, are the most common types of esophageal cancer. Nonepithelial malignancies comprise only about 0.5% of all primary esophageal neoplasms. Primary lymphoma of the GI tract accounts for about 5%–20% of all cases of lymphoma. Isolated NHL of the esophagus is an extremely uncommon occurrence, accounting for <1% of patients with lymphoma and occurs more often in the distal esophagus.^[4] Furthermore, isolated primary lymphoma of the esophagus without an extraesophageal location is extremely rare and only 20 such cases have been reported in literature to date.^[5,6]

The etiology of the disease is unknown, with the role of Epstein–Barr virus being controversial. It has been noticed that it is most common in immune compromised patients, with HIV infection as a probable risk factor. Dysphagia is the most common symptom at presentation with less common symptoms being odynophagia, fever, and weight loss. The age at presentation of the disease is highly variable. Endoscopic and radiological findings of esophageal lymphoma are nonspecific and nondiagnostic. The diverse spectrum of endoscopic/radiological patterns

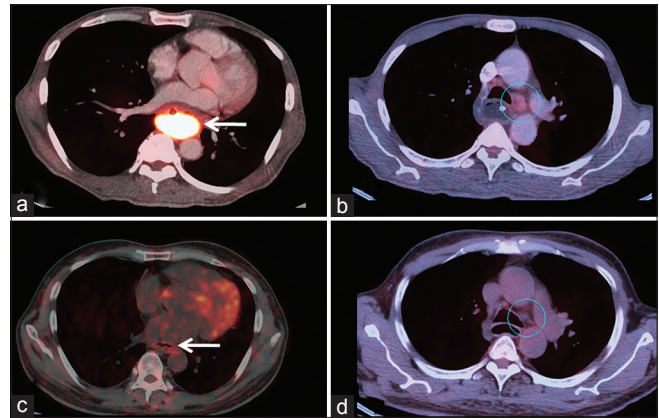


Figure 2: Pre- and post treatment 18-F fluorodeoxyglucose positron emission tomography-contrast-enhanced computed tomography study in a patient diagnosed of esophageal Non-Hodgkin's Lymphoma. Axial-fused positron emission tomography-computed tomography images show the bulky 18-F fluorodeoxyglucose avid esophageal mass (arrow in 2a) and aortopulmonary lymph node (circle in 2b). Post treatment positron emission tomography-contrast-enhanced computed tomography study showing complete metabolic and morphological response of the circumferential thickening in the middle and lower third esophagus (arrow in 2c) and also the aortopulmonary nodes (circle in 2d)

that have been described for esophageal lymphoma include stricture, ulcerated mass, multiple submucosal nodules, varicoid pattern, achalasia-like pattern, progressive aneurysmal dilatation, and tracheoesophageal fistula formation. Computed tomography (CT) scan is valuable for the evaluation of the extraluminal component of an esophageal mass, its mediastinal extension, fistula formation, and status of the lymph nodes; therefore, it has a role in disease staging, assisting in stratification of various available treatments, evaluating treatment responses, monitoring patient progress, as well as detection of any relapses. PET-CT scanning has emerged as an indispensable tool in the staging and follow-up of patients with extranodal involvement in lymphoma. PET-CT has also significantly increased the detection of indolent lesions that were undetected by conventional cross-sectional imaging.^[7]

Dawson's criteria^[8] to identify primary GI lymphoma include nonpalpable superficial lymphadenopathy, no splenic involvement, and no enlargement of mediastinal or hilar lymph nodes. Our patient fit Dawson's criteria fully, except that he also had enlarged mediastinal nodes. The application of Dawson's criteria for the diagnosis of primary GI lymphoma may not hold true for the esophagus. Modifications as exclusion of mediastinal lymphadenopathy in the criteria for primary esophageal lymphoma have been suggested, because the esophagus is a mediastinal structure with an extensive lymphatic network which results in wide lymph node basins. Therefore, based on the predominant lesion being in the esophagus with no involvement of liver, spleen, and peripheral lymph nodes, we did not consider the absence of mediastinal lymphadenopathy as strict criteria for a diagnosis of primary esophageal lymphoma;

the positive endoluminal biopsy in the absence of nodal disease infiltrating the esophageal wall confirmed the primary origin in the esophagus.

Conclusion

Primary NHL of the esophagus is an extremely rare esophageal neoplasm. Imaging modalities are nonspecific, thus posing a diagnostic dilemma. Endoscopic biopsy is the gold standard to confirm the diagnosis. Accurate histopathological diagnosis supplemented with necessary immunohistochemical stains helps in accurate diagnosis; complete staging work-up using PET-CT scanning aids in treatment decision and prognostication. Appropriate chemotherapy with radiotherapy may offer a significant chance of long-term survival.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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