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Case report

Primary mucosa-associated lymphoid tissue lymphoma of the esophagus masquerading as a benign tumor

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ABSTRACT

We report a case of primary esophageal low-grade B-cell lymphoma of MALT type in a 50-year-old Saudi male patient who presented to our hospital with a history of dysphagia and heartburn for more than 2 years. Endoscopy showed a large esophageal mass with an intact mucosa located in the distal esophagus, 28 cm–35 cm from the incisor teeth. Endoscopic ultrasonography (EUS) showed a large well demarcated sub-epithelial lesion 4 cm in width and 10 cm in length arising from the muscularis mucosa with mixed echogenicity consistent with benign leiomyoma. Subsequently, the patient underwent surgical resection of the tumor; the histopathology confirmed the diagnosis of esophageal lymphoma. The tumor was considered to be completely resected and therefore additional treatment was not administered. The patient was doing well on follow up after treatment. Clinically and radiologically he did not reveal any signs of recurrence. Surgical resection is beneficial as a primary treatment option in incipient primary low grade MALT esophageal lymphomas.

1. Introduction

Primary esophageal lymphoma is extremely rare, with only a few cases of primary mucosa-associated lymphoid tissue (MALT) lymphomas of the esophagus reported in the literature and none of them from Saudi Arabia. Although lymphomas are malignant neoplasms which are usually confined to the lymph nodes, one-fifth of lymphomas present with extra-nodal localization [1]. The esophagus is an uncommon localization accounting for less than 1% of patients with lymphoma and is usually seen secondary to mediastinal nodes or gastric lymphoma [2,3]. Primary MALT lymphoma of the esophagus is exceptionally rare, and when seen, is usually the non-Hodgkin's type [4] with few reports of primary MALT lymphoma of the esophagus in the English-language literature and a few cases in Japan and no reports from Saudi Arabia. The findings in esophageal lymphomas may appear similar to tumors of benign origin like leiomyoma [5,6]. We report a case of primary esophageal MALT lymphoma presenting as a large sub-mucosal tumor (SMT).

2. Case report

A 50-year-old Saudi male, with no significant medical history, presented to our hospital with history of dysphagia and heartburn for more than 2 years. He denied hemoptysis, hematemesis, and weight loss and reported an active smoking history for 20 years and no alcohol consumption. Physical examination was unremarkable, with no palpable lymphadenopathy, ascites, or organomegaly. Laboratory data were normal including: white blood cell count 4.32 K/uL, hemoglobin value 14.4 g/dL, and platelet count 247 K/uL. Fasting blood sugar level was 6 mmol/L. HBs-Ag negative, HBs-Ab 852.2 mU/mL, HCV-Ab negative, HIV 1&2 negative, rapid urease test for *Helicobacter pylori* was negative. Initial upper gastrointestinal endoscopy (UGIE) showed a large mass protruding to the lumen with an intact mucosa extending from 28 cm to 35 cm from incisor teeth above the cardia (Fig. 1). EUS demonstrated a large well demarcated sub-epithelial (SE) lesion about 4 cm in width and 10 cm in length arising from the muscularis mucosa (MM) with mixed echogenicity and echogenic strands. The mass is located posteriorly in front of the aorta respects completely the sub-mucosal layer deeper without any suspicious regional lymph nodes. The margins were very well demarcated as well consistent with benign leiomyoma features. Chest CT was notable for an 11 cm intramural lesion in the lower third of the esophagus, causing

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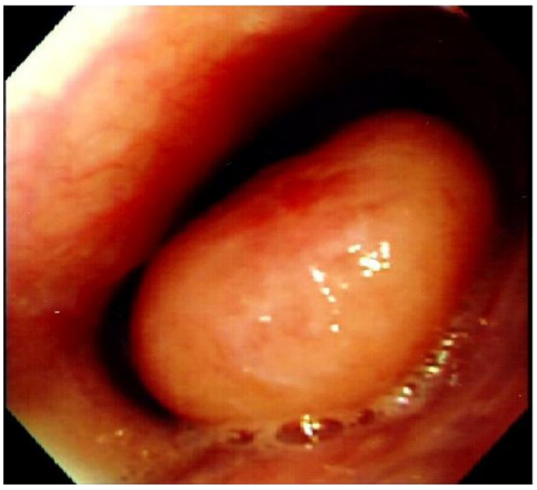

Fig. 1. Endoscopic image of the esophagus showing a large mass with an intact mucosa.

marked narrowing of the esophageal lumen distally and dilatation proximally; no mediastinal or hilar lymphadenopathy were found (Fig. 2a, b). The patient underwent surgical resection of the mass through left postero-lateral thoracotomy approach; tumor was excised completely with part of the mucosa attached to it. Microscopic examination revealed a diffuse proliferation of small lymphoid cells that are covered by stretched and unremarkable stratified squamous epithelium. The lymphoid cells were small to medium sized, slightly irregular nuclei with variably conspicuous nucleoli and abundant pale cytoplasm, scattered large blast like cells are identified with infrequent mitosis and apoptosis (Fig. 3a–d). Immunohistochemistry revealed that the small lymphoid cells are positive for LCA, CD20, CD79a, CD43, bcl2a (Fig. 4) and negative for the rest of markers including CD3, CD5, CD23 (Fig. 5a), Cyclin D1 and bcl6 (Fig. 5b). The tumor was classified as a primary low grade B-cell MALT esophageal lymphoma. On subsequent follow up for more than 12 months duration, the

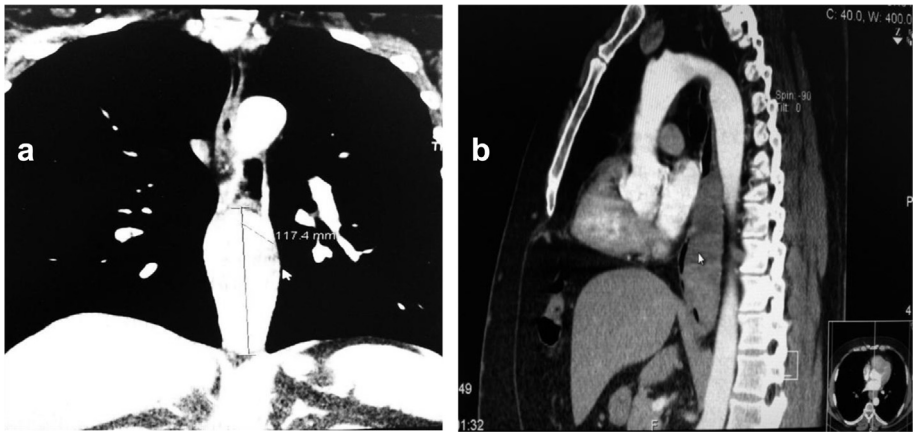

Fig. 2. Chest CT revealed an 11 cm intramural lesion in the lower third of the esophagus, causing marked narrowing of the esophageal lumen distally and dilatation proximally. No mediastinal or hilar lymphadenopathy.

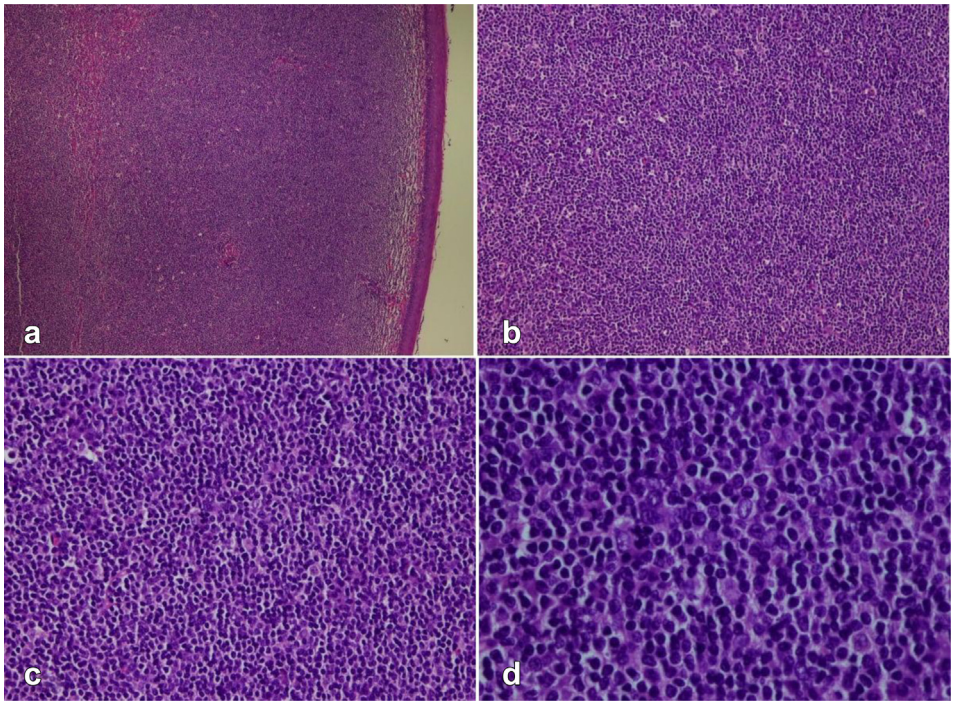

Fig. 3. H&E slides showing the lymphoid cells with small to intermediate sized irregular nuclei and scattered blast like cells.

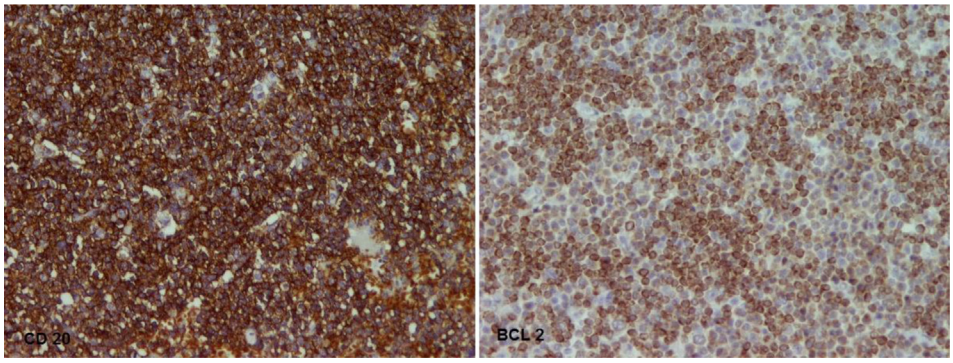

Fig. 4. Slide showing the positivity of tumor cells for CD20 and BCL2.

patient is asymptomatic with no clinical or radiological evidence of recurrence.

3. Discussion

Previously described cases have reported a variety of radiographic appearances for primary esophageal lymphoma. The most common include polypoid masses with or without erosions, stricture with ulceration mimicking esophageal cancer, thickening of the mucosal folds producing a varicoid appearance, narrowing of the distal segments (achalasia-like appearance), or submucosal nodules [5,6,8]. Given such a diverse spectrum of radiographic appearance, EUS, CT and biopsy of the esophageal wall is needed in order to confirm the diagnosis. In our case, CT revealed an 11 cm smooth intramural lesion in the lower third of the esophagus, causing marked narrowing of the esophageal lumen distally and dilatation proximally; no mediastinal or hilar lymphadenopathy were found. With the clinical application of EUS, great progress has been made in the diagnostic accuracy of structural abnormalities and depth of invasion in various gastrointestinal diseases, including lymphoma [7]. EUS provides more accurate information on the involvement of the wall layers. However in our case, it is revealed a large well demarcated SE lesion about 4 cm in width and 10 cm in length arise from the MM with mixed echogenicity and echogenic strands reaching the GE junction, mimicking the leiomyoma features which usually presents as a homogeneous and hypoechoic lesion with clear margins, surrounded by a hyperechoic area, which can easily be differentiated from a lipoma, cyst, or hemangioma in the esophageal wall. Preoperative biopsy of the tumor is a debating issue [9]. Our EUS showed all detailed features which were toward

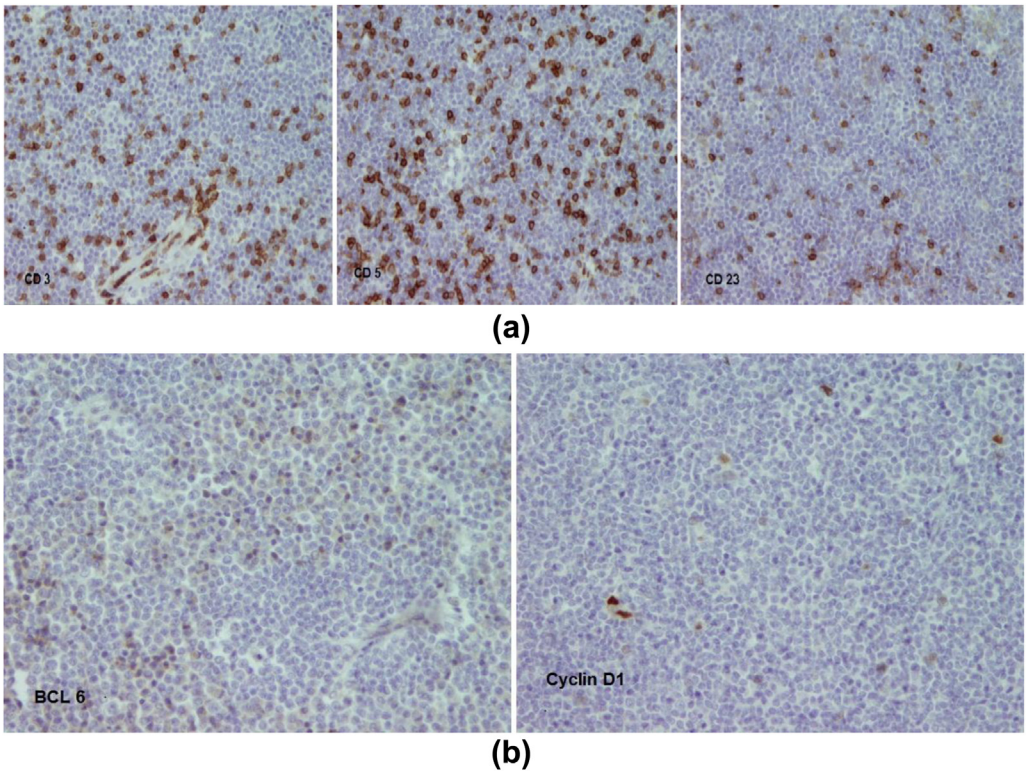

Fig. 5. Two slides showing that the tumor cells were negative for CD3, CD 5, CD 23, Cyclin D1 and BCL 6.

benignity so our policy was not to recommend it. To the best of our knowledge, our case of primary MALT esophageal lymphoma is the first described case in the literature to have a benign appearance on CT, UGIE, and EUS. Given the non-specific clinical, radiological and endoscopic appearance, we recommend the pre-operative endoscopic biopsy of the lesion as a guide to diagnosis even with the benign features.

Intraoperative, we noted that the tumor could not be resected without part of the mucosa, which was a clue that this was not a benign tumor, although this was not evident before the operation. Pathologic evaluation of our lesion showed low grade B-cell MALT esophageal lymphoma. The treatment of esophageal lymphoma either with chemotherapy, radiotherapy or resection endoscopically or through thoracotomy, depends on the histological tumor type and its initial location. Because of the rarity of MALT lymphoma of the esophagus, its diagnostic features have not yet been well recognized and optimal treatment has not been properly discussed. Though radiotherapy is widely preferred for gastric and orbital MALT lymphoma, surgery has been the most frequently reported treatment for esophageal MALT lymphoma. We believe that surgical resection is beneficial as a primary treatment option in incipient primary low grade MALT lymphomas as our patient was asymptomatic post-operatively after more than 12 months of follow up with chest CT and gastrografin swallow which not revealing any residual lesion. Esophageal MALT lymphomas should be considered in the differential diagnosis of immune-competent patients presenting with dysphagia.

Conflict of interest statement

None to declare.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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