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Primary non-Hodgkin lymphoma of the esophagus: case report

Linfoma não-Hodgkin Primário de Esôfago: Relato de Caso

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Resumo

O linfoma não-Hodgking primário do esôfago é muito raro, representando menos de 1% de todas as neoplasias do trato gastrointestinal. Relatamos o caso de um paciente de 60 anos com história de disfagia progressiva de dois anos, sem história de imunodeficiência e sem anormalidades laboratoriais ou radiológicas. A endoscopia mostrou uma lesão esofágica vegetante de dois centímetros invadindo a cárdia com limites imprecisos. Três esofagoscopias com biópsias foram realizadas revelando, contudo, resultados inconclusivos. Sem um diagnóstico histopatológico definitivo, o paciente foi submetido a uma esofagectomia trans-hiatal com linfadenectomia e reconstrução, utilizando-se o estômago. A análise histológica do tumor revelou citoplasma eosinofílico e marcante hipercromatismo, e pleiomorfismo nucleares com atividade mitótica aumentada. As células do tumor estavam invadindo a camada muscular. O diagnóstico de linfoma não-Hodgkin foi feito e confirmado pela imunoistoquímica usando dois marcadores: o Ki67 e o CD20. Após a cirurgia, o paciente recebeu três ciclos de quimioterapia. Até o momento, após 49 meses de acompanhamento, ele mantém-se assintomático. Concluímos que, embora o linfoma de esôfago seja muito raro, ele deve ser considerado como um diagnóstico diferencial em situações como esta. Além disso, a análise imunoistoquímica constitui um poderoso instrumento para confirmação diagnóstica.

Palavras-chave: Neoplasias gastrointestinais, Neoplasias esofágicas, Linfoma não-Hodgkin.

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Abstract

Primary non-Hodgkin lymphoma of the esophagus is quite rare, representing less than 1% of all gastrointestinal neoplasms. We report the case of a 60-year-old male with 2-year progressive dysphagia and weight loss. The patient had no previous history of immunodeficiency, and laboratory work-up and chest radiography revealed no abnormalities. Endoscopy showed an ulcerated and vegetative lesion in the esophagus, 2 cm long, deeply invading the gastric cardia, with imprecise borders. Three esophagoscopies with biopsies were performed, all of which showed inconclusive results. Without a definitive histopathological diagnosis, the patient underwent one-stage distal transhiatal esophagectomy with lymphadenectomy and reconstruction using the stomach. Histological examination of tumor cell sheets revealed eosinophilic cytoplasm, and the nucleus showed marked hyperchromatism, pleomorphism, and increased mitotic activity. The tumor cells were infiltrating the muscularis propria. A diagnosis of non-Hodgkin's lymphoma was made and confirmed by immunohistochemistry using two markers: Ki67 and CD20. Surgery was followed by three cycles of chemotherapy. The patient now remains asymptomatic after 49 months of follow-up. In conclusion, although esophageal lymphoma is very rare, it should be considered as a differential diagnosis in situations like this. Furthermore, immunohistochemical analysis is a powerful tool for confirming the diagnosis.

Key words: Gastrointestinal neoplasms; Esophageal neoplasms; Lymphoma non-Hodgkin.

INTRODUCTION

Squamous cell carcinoma and adenocarcinoma are the most frequent malignant esophageal neoplasms, occurring in 95% of cases. Primary non-Hodgkin lymphoma of the esophagus is very rare, representing less than 1% of all gastrointestinal neoplasms1. We report an immunocompetent patient with primary non-Hodgkin lymphoma involving the lower third of the esophagus.

CASE REPORT

A 60-year-old male was admitted to our hospital with 2-year progressive dysphagia, allowing him to swallow only liquids at the time of presentation. He also complained of weight loss, but did not present anorexia. He had no previous history of immunodeficiency, and on physical examination showed no peripheral lymphadenopathy.

Laboratory work-up revealed normal red blood cell levels and total and differential leukocytes. Chest radiography showed no abnormalities. A barium meal in the esophagus evidenced a slower rate of contrast progression and normal esophagus situation. A narrowed segment 2 cm long was detected involving the lower third of the esophagus, with an exuberant irregularity and a thick pleated mucous membrane (Fig.1). Stomach capacity and contour were normal.

Endoscopy showed an ulcerated and vegetative lesion in the esophagus, 2 cm long and located 38 cm from the incisor teeth. The lesion was deeply invading the gastric cardia, with imprecise borders. Three esophagoscopies with biopsies were performed, but all three showed only an ulcerated lesion with a fibrinoleukocytic crust and necrosis, with no malignant cells.

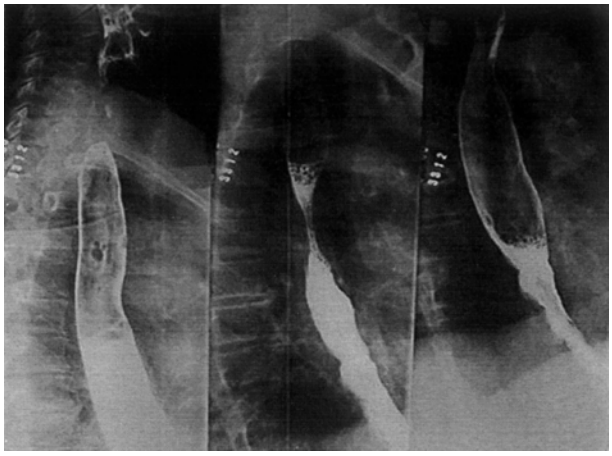

Figure1. A barium meal of the esophagus shows lower progression of contrast and normal esophagus situation. A narrowed segment - 2 cm long - is shown involving the lower one third of the esophagus, with an exuberant irregularity and a thick pleated mucous membrane in this region of the esophagus

Ultrasound examination demonstrated no evidences of abdominal lymphadenopathy, liver involvement, or ascites. Without a definitive histopathological diagnosis, the patient was submitted to one-stage distal transhiatal esophagectomy with lymphadenectomy and reconstruction using the stomach. At laparotomy, there was no splenomegaly, no abdominal lymphadenophaty, no involvement of the gut beyond the esophagus, and no liver injury.

Gross examination of the resected specimen showed a 4 X 4-cm tumor involving the lower third of the esophagus. There was no evidence of metastasis in the

periesophageal lymph nodes analyzed.

Histological examination of tumor cell sheets revealed eosinophilic cytoplasm and a large vesicular round nucleus with two to three prominent nucleoli. The nucleus showed marked hyperchromatism, pleomorphism, and increased mitotic activity (Fig.2).

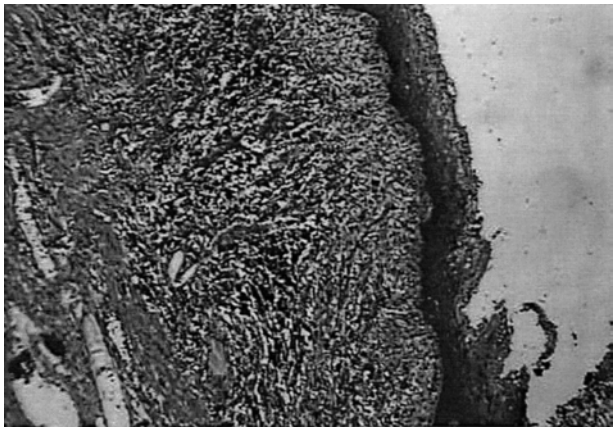

Figure 2. Tumor cells with eosinophilic cytoplasm and large vesicular round nucleus with two to three prominent nucleoli. The nucleus showed marked hypercromatism, pleomorphism, and increased mitotic activity (HE 40X)

The tumor cells were infiltrating the muscularis propria. A diagnosis of diffuse large-cell non-Hodgkin's lymphoma was made (Fig. 3) and confirmed by immunohistochemistry using two markers: Ki67 and CD20. Ki67 showed 75-100% expression in the nucleus and C20 showed high membrane positivity. This pattern classified the tumor as a B-cell non-Hodgkin lymphoma with high proliferative activity.

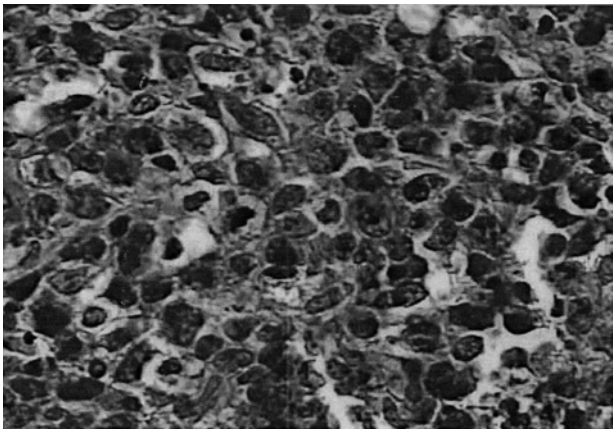

Figure 3. The tumor cells showed infiltration in the muscular propria layer (HE 400X)

Surgery was followed by three cycles of chemotherapy (cyclophosphamide, vincristine, and predinisolone) at 2-month intervals. Fifteen days later the patient underwent another surgery due to a small intestinal intussuception. Patient is now asymptomatic after 49 months of follow-up.

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