Unusual esophageal infiltration by lymphoma in a woman with Sjögren's syndrome

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RESUMEN

Presentamos un caso de infiltración esofágica por un linfoma difuso de células grandes B en una mujer de 63 años de edad con síndrome de Sjögren (SS). Identificamos en el tercio superior del esófago una lesión ulcerada de base granular y márgenes elevados, que midió 18 mm de eje mayor, la cual correspondió a infiltración linfomatosa. La infiltración esofágica por linfoma como lesión ulcerada única en un paciente con SS es muy rara.

Palabras clave. Linfoma MALT. Esófago.

ABSTRACT

We present a case of esophageal infiltration by a diffuse large B-cell lymphoma in a 63 year-old woman with Sjögren's syndrome (SS). We identified in the upper third of the esophagus, an ulcerated lesion with a granular base and elevated margins that measured 18 mm in greatest dimension corresponding to a lymphomatous infiltration. An esophageal infiltration by lymphoma as only ulcerated lesion in a patient with SS is very rare.

Key words. MALT-lymphoma. Esophagus.

One month before admission at the hospital a 63 year-old woman with Sjögren's syndrome (SS) had severe abdominal pain and fever. She had been treated with steroids and cyclophosphamide during the last 8 years. At the admission, the physical examination revealed a left positive Giordano's sign. A clinical diagnosis of renal and urinary infection was considered, and she received antimicrobial treatment without symptomatic remission. A computed tomography showed a 43 mm tumor in the upper pole of the right kidney with direct invasion into the right hepatic lobule as well as multiple pulmonary nodules and another 15 mm left hepatic nodule, which were considered as metastases of a probable renal cell carcinoma. The patient had an unfavorable evolution and died by gram-negative bacteria septicemia. An autopsy study was performed. Microscopically, all these nodules, including hepatic and renal tumors, correspond to an infiltration by a CD20-positive diffuse large B-cell lymphoma (DLBCL). Interesting, in the upper third of the esophagus, we identified an ulcerated lesion with a granular base and elevated margins, which measured 18 mm in greatest dimension (Figure 1A). This lesion was also lymphomatous infiltration (Figures 1B-1D). The other portions of the esophagus and the digestive tract were unremarkable.

The digestive tract is the extranodal site most frequently involved by non-Hodgkin lymphoma. The stomach is the most commonly affected organ, followed by the small intestines, colon, and rarely, the esophagus (1% or less).[1] When a lymphoma affects the esophagic wall, only 10% will be primary, and they occur mainly in males over 50 years old. In these patients, HIV-AIDS infection and other immunosuppressed states increase the risk for lymphoma in this anatomic location.[1,2] In case report series, the most common subtypes of lymphoma involving the esophagus

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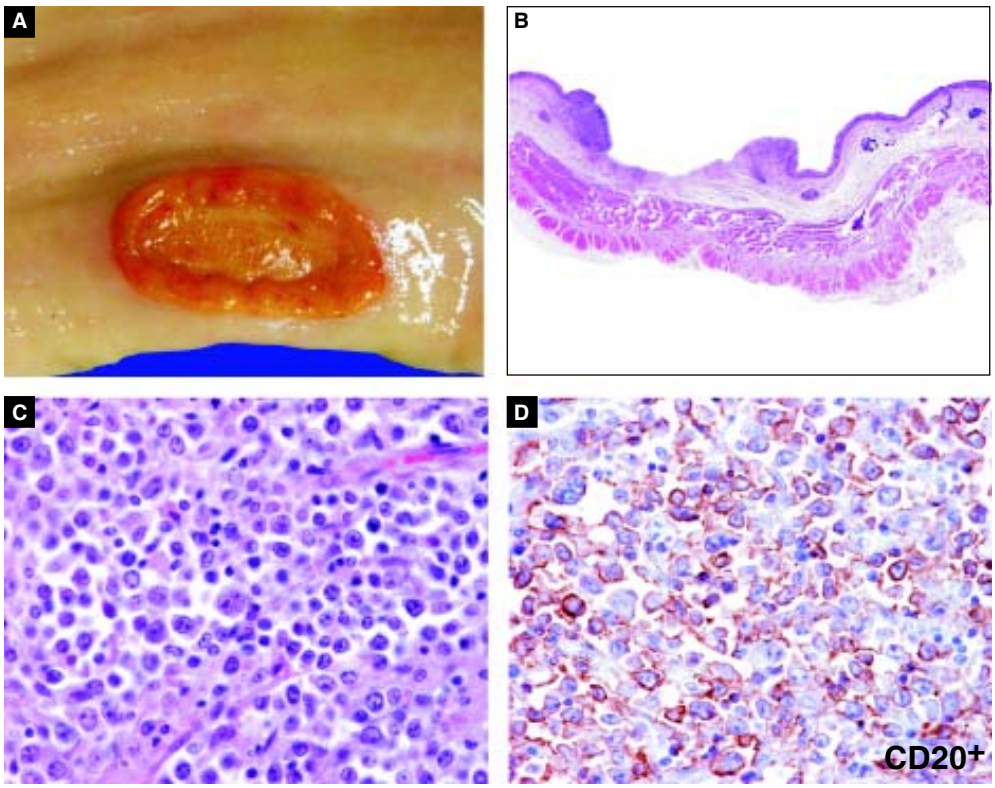

Figura 1. A. The esophagus showed an ulcerated lesion, which measured 18 mm in greatest dimension. **B.** A whole mount section of the tumor showed an infiltrating tumor localized in the esophageal mucosa and submucosa. **C.** The tumor was composed of large malignant cells with marked nuclear pleomorphism and prominent nucleoli, as well as, scant eosinophilic cytoplasm. **D.** The malignant cells were strongly positive for CD20.

were DLBCL and extranodal marginal zone B-cell lymphomas of mucosa-associated lymphoid tissue (MALT-lymphoma); but mantle cell lymphoma and rarely T-cell lymphomas have been also reported.[1,3-5] The endoscopic and gross features of these cases include diffuse, infiltrative, polypoid, nodular, and less commonly ulcerated patterns.[2] In some patients an initial clinical diagnosis of poorly differentiated carcinoma was considered. By the way, in a cohort study of patients with SS, MALT-lymphomas constitute the majority of cases (59%) of lymphomas, followed by nodal marginal zone lymphomas (15%) and diffuse large B-cell lymphomas (15%), but the esophagic infiltration was not mentioned.[2] In conclusion, an esophageal infiltration by lymphoma as only ulcerated lesion in a patient with SS is very rare.

CONFLICT OF INTEREST

The authors disclose no conflicts.

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