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Histiocytic sarcoma of the esophagus

Carlos Comesaña Castellar[1], Maria Antònia Payeras Capo[1], Joaquín Fernández García[1], Cristina Calvo Martínez[2], Maria Dolors Ramis Estelrich[1], Raúl José Díaz Molina[1], Pere Vaquer Grimalt[1], Willy Denis Sánchez Fernández[1]

[1]Gastroenterology and [2]Anatomic Pathology Departments. Hospital Universitari Son Espases. Palma de Mallorca, Spain

Correspondence: Carlos Comesaña Castellar
e-mail: carloscome@gmail.com

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Dear Editor,

A 20-year-old male with no medical history of interest presented to the Emergency Room because of retrosternal pain, odynophagia, dysphagia and fever. On physical examination the axillary temperature was 37.7 °C, he had a poor general condition and central chest pain on palpation. The blood tests showed: 16,200 x 10^6/l white blood cells, 12,800 x 10^6/l neutrophils and 11.66 mg/dl C reactive protein. The rest of the complete blood count, coagulation and biochemistry were within normal values.

There were no pathological findings by computed tomography of the chest. Thus, an upper gastrointestinal endoscopy was performed, finding an extensive superficial 7 cm long ulcer with geographical edges in the middle third of the esophagus, involving almost the entire circumference (Fig. 1). Biopsies were taken and serologies were requested to rule out an infectious etiology, all of which were negative. The patient received high dose proton pump inhibitors, and was discharged from hospital after a good clinical evolution.

The pathological analysis revealed an esophageal mucosa with intense ulceration and acute inflammation, finding vessels with perivascular infiltration of blast-like cells, compatible with an atypical perivascular myelomonocytic/histiocytic neoplasm. After discussing the case with the Hematology Department, a positron emission tomography/computed tomography (PET/CT) was performed, as well as a bone marrow aspiration and biopsy. No malignant lesions were seen with the PET/CT scan, the bone marrow aspiration was normal and no neoplastic infiltration was observed in the biopsy. The lesion was finally diagnosed as histiocytic sarcoma of the esophagus and radiotherapy was started in the affected area.

Discussion

Histiocytic sarcoma (< 1 % of all hematological neoplasms) results from the malignant proliferation of mature tissue histiocytes. The mean age at diagnosis is 52 years and it normally affects extranodal sites, mainly the skin and gastrointestinal tract. Pathological analysis and immunohistochemistry are key to establish the diagnosis. Staging is performed from a bone marrow study (aspiration and biopsy) and PET/CT, with most patients having disseminated disease at diagnosis. Treatment depends on extension, consisting of surgery and radiotherapy for localized tumors; and alemtuzumab (monoclonal antibody against CD52), thalidomide and/or bone marrow transplant for disseminated cases. The mean survival of these patients is less than two years. So far, only one histiocytic sarcoma with primary esophageal involvement has been reported.

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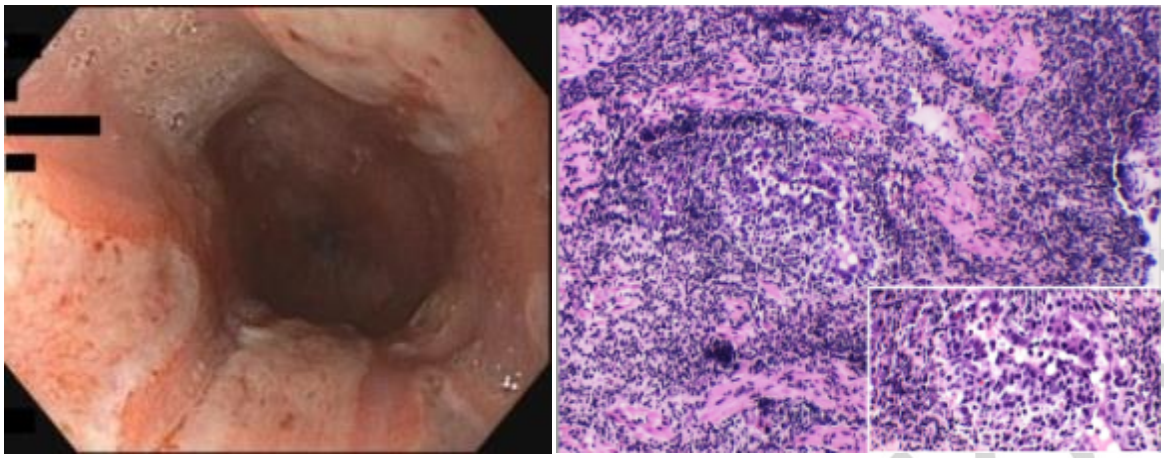

Fig. 1. Histiocytic sarcoma of the esophagus. A. Extensive superficial ulcer in the middle third of the esophagus seen by endoscopy. B. Ulcer biopsy with hematoxylin-eosin staining with high inflammatory infiltrate. C. Same image as B, magnified with high inflammatory infiltrate and the presence of small blood vessels.