

## CASE REPORTS

# Unusual involvement in mycosis fungoides: duodenal papilla

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### ABSTRACT

**Background:** Mycosis fungoides (MF) is a type of T-cell lymphoma with cutaneous involvement. It is a rare disease, of low incidence and usually affects middle-aged men. In most cases only the skin is involved although in advanced stages may present with extra cutaneous involvement including the gastrointestinal tract.

**Case report:** We report the first case of MF with compromise of duodenal papilla, emphasizing the diagnostic approach and a brief review of the subject.

**Discussion:** This case report proves the value of the endoscopic studies in patients with lymphoproliferative disorders, because of the impact in the diagnosis and prognosis. Also, this case report is relevant because there is no scientific evidence, as far as we know, of similar cases reported.

**Key words:** Mycosis fungoides. Lymphoma. T-cell. Cutaneous. Papilla of Vater.

### INTRODUCTION

Mycosis fungoides (MF) is a type of T-cell lymphoma with cutaneous involvement (1). It is a rare disease, of low incidence, and usually affects middle-aged men (2). In most cases, only the skin is involved (3), although in advanced stages it may present with extracutaneous involvement, including the gastrointestinal tract (4,5). We report the first case of MF involving the duodenal papilla, emphasizing the diagnostic approach and a brief review of the subject.

### CASE REPORT

This is a 45-years-old man who was diagnosed with cutaneous T lymphoma five years ago and was treated with several lines of chemotherapy, from methotrexate to liposomal doxorubicin. He entered through the emer-

gency room of our hospital complaining of fever, fatigue, weakness for a week and purulent cutaneous lesions in the scalp and eyelids. At physical examination, he presented tachycardia, tachypnea, fever and more than 70% of his body surface was affected by extensive coalescent plaques, nodules and tumors, giving erythrodermic appearance, most with necrotic center and ulcerated. On his scalp there were areas of extensive ulcers draining pus. He received the diagnosis of infected skin lesions by MF and blepharoconjunctivitis. He was treated with broad-spectrum antibiotics but patient's clinical course was torpid, with persistent signs of systemic inflammatory response, thus requiring carbapenem and oxazolidone. Fever persisted after 14 days of antibiotic treatment. A computed tomography (CT) of the chest and abdomen was performed in search of other sites of infection and as staging of the MF, besides bone marrow biopsy. On CT, some pathological cervical and inguinal lymph nodes were observed and at the level of the second portion of the duodenum there was a mass associated to pneumobilia and a dilated common bile duct (Fig. 1). A duodenoscopy showed a large duodenal papilla, its mucosal surface appearing infiltrated, eroded, friable, easily bleeding. Multiple biopsies were taken (Fig. 2). A fistulous suprapapillary orifice draining bile was also observed. An endoscopic ultrasound (EUS) showed duodenal papilla infiltrated by a hypoechoic lesion of 26 mm x 24 mm in diameter, heterogeneous, ill-defined borders, with secondary infiltration of the pancreatic head without vascular invasion or regional lymph nodes. The bile duct and main pancreatic duct were dilated with diameters of 8 and 5 mm respectively (Figs. 3 and 4). Histopathology showed duodenal mucosa infiltrated by atypical lymphoid cells. Immunohistochemistry revealed dense infiltration of T cells positive for CD3 and CD5, CD7 focal loss of dominance and CD4 cell proliferation rate of 15% plus few CD20 positive B cells intermingled, suggestive of infil-

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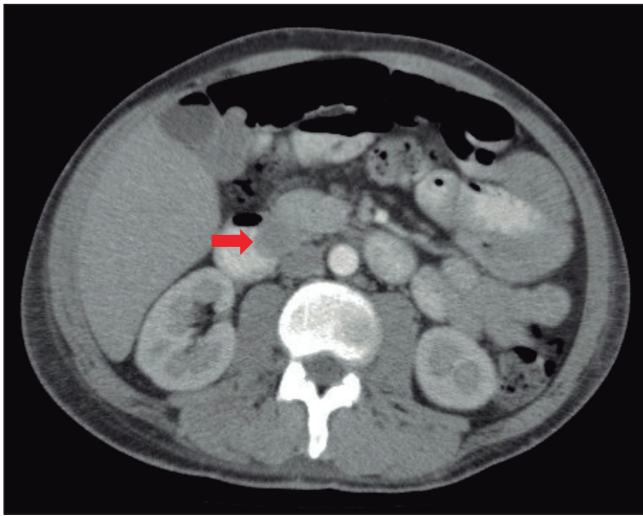


Fig. 1. Abdominal CT scan axial plane. Mass in the second duodenal portion (red arrow).

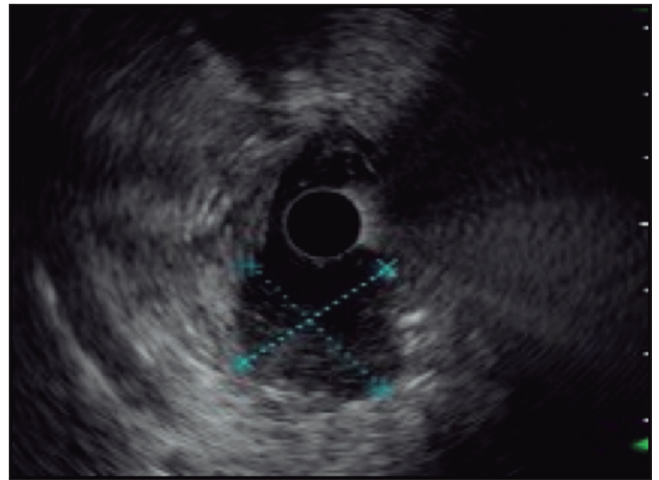


Fig. 3. EUS. Neoplastic appearance of papilla of Vater.



Fig. 2. Gastroscope. Papilla of Vater infiltrated by MF.

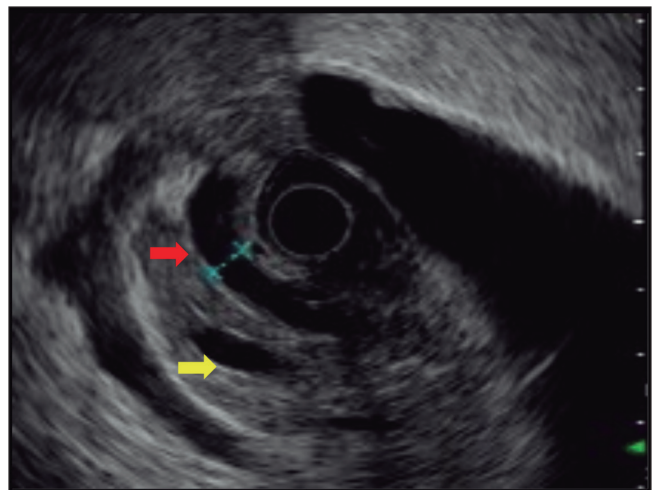


Fig. 4. EUS. Common bile duct (red arrow) and main pancreatic duct (yellow arrow).

tration by MF. Bone marrow biopsy confirmed the same findings.

Considering the extensive commitment to MF, the refractory to several lines of oncological management and the difficulty in controlling the infectious process, no salvage chemotherapy began. Finally, after 20 days of hospitalization the patient died of septic shock and severe sepsis.

**DISCUSSION**

MF is the most common cutaneous T-cell lymphoma (1). The MF incidence is about 3 per million a year, affecting people between 55 to 60 years of age (2). The disease may

have a slow, indolent evolution and causes skin lesions that include nodules, plaques, and tumors that can progress to compromise an extensive area of skin (3). In advanced cases with aggressive disease, it may be seen compromise of bone marrow, lymph nodes, liver, lung and gastrointestinal tract. Some autopsy studies have shown that between 70% and 90% of patients die of visceral MF involvement. Visceral involvement correlates to the severity of skin lesions, and cutaneous infections are common (4,5). It is recommended to confirm visceral involvement with biopsies and immunohistochemically because non-lymphomatous tumors can occur in long term affected patients of less than 30 years of age. A retrospective study of 404 patients found a high risk of primary non-skin neoplasia in MF patients (SIR 3.40; 95% CI, 1.55-6.45), mostly synchronous lymphoma (SIR 12.86; 95% CI, 2.65-37.59) and melanoma (SIR 9.31; 95% CI, 8.75-33.62) (6). Colorec-

tal, liver, biliary tract, lung and urinary tumors have been also reported (7). MF patients' survival depends on disease stage. In early stages it equals general population survival, but in extracutaneous involvement it can be as short as 1 to 2 years. Dismal prognosis factors also include older patients, refractoriness to successive treatment protocols, lymph node involvement and presence of Sézary cells in histology (7,8).

MF gastrointestinal involvement can be primary or spreading from cutaneous disease. Primary forms are quite rare and patients can present diarrhea, perforation, bleeding, dysphagia and intestinal obstruction (9-12). In cases of cutaneous spreading to digestive tract, involvement often occurs in the stomach, small bowel and pancreas (13-15). Arai et al. (13) reported an autopsy study of 107 patients and found pancreas involvement in 20% of cases. Madsen et al. (14) reported a 39 year old patient with refractory MF that presented biliary obstruction due to MF involvement of biliary tract. Gottlieb et al. (15) also reported a patient with biliary MF obstruction due to a pancreatic MF. On EUS they found a hypoechoic infiltrative lesion in the head of the pancreas, without vascular and lymph node involvement. Fine needle biopsy of the lesion shows lymphoma cells, CD45, CD3 positives, CD5, CD20, SYP, cytokeratines and chromogranin negatives, suggesting T-cell MF lymphoma.

We here report an MF patient with extensive and severe cutaneous disease, with papilla of Vater involvement, with secondary biliary tract dilation; however, it never presented jaundice or liver profile alterations, due to the presence of a suprapapillary fistulae. In this patient the diagnosis of MF involvement of papilla of Vater was important to establish a prognosis, supporting the decision of giving only best supportive and compassionate care in a patient refractory to second line treatment protocols. This case report proves the value of the endoscopic studies in patients with lymphoproliferative disorders, because of the impact in the diagnosis and prognosis. Also, this case report is relevant because there is no scientific evidence, as far as we know, of similar cases reported.

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