Obstructive Jaundice Caused by Primary Duodenal Lymphoma

Case Report and Review of the Literature

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Dear Sir,

Primary duodenal lymphoma is a rare neoplasm, and initial presentation with obstructive jaundice is even rarer. Primary B-cell lymphoma of the duodenum was shown to be the cause of biliary obstruction in the present case.

A 70-year-old woman was referred to our hospital in July 2005 because of pruritic jaundice and dyspepsia for a few days. There was no weight loss, fever or nocturnal sweats. The patient had a history of bronchoalveolar adenocarcinoma (pT1pN0) for which she had had a resection of the left lower pulmonary lobe 4 years ago. Physical examination on admission was normal apart from jaundice. Abnormal laboratory findings included bilirubin 154 $\mu\text{mol/l}$ (normal value <20), alkaline phosphatase 547 IU/l (<117), and ALT 284 IU/l (<31); LDH was normal. CT scan showed dilated biliary and pancreatic ducts and a slight thickening of the duodenal wall without pancreatic or peripancreatic mass. Endoscopic retrograde cholangiography revealed an ulcerative tumor in the descending part of the duodenum including the papilla of Vater. There was massive dilatation of the biliary and pancreatic ducts with a stenotic section of about 3 cm in length in the distal common bile duct. A sphincterotomy was performed, and two 7-Fr plastic double pigtail stents were placed and later replaced by a single 10-mm self-expanding metal stent. Unexpectedly, his-

tology of the tumor showed diffuse large B-cell lymphoma. The patient was treated with six courses of the CHOP scheme of chemotherapy, accompanied by immunotherapy with rituximab and followed by consolidating radiotherapy. In the CT scan, a good remission of the duodenal wall thickening could be documented, and the patient has been well for 18 months since the first course of chemotherapy.

Discussion

Gastrointestinal lymphomas account for approximately 40% of all extranodal lymphomas and for 4–12% of all non-Hodgkin lymphomas [1–3]. The incidence has been reported to be 0.8–1.2 per 100,000 persons per year [4]. By far the most common site of primary gastrointestinal lymphoma is the stomach, accounting for 75–80% of all primary gastrointestinal lymphomas, followed by the ileum (12–15%) and colorectum (approx. 5%). The duodenum only accounts for 0.8–2% of all gastrointestinal lymphomas [1, 5].

Symptoms most often associated with primary lymphoma of the small intestine include abdominal pain, anorexia, weight loss and manifestations of bowel obstruction. Less often observed are nausea, vomiting or diarrhea and only rarely gastrointestinal bleeding, and obstructive jaundice

is extremely rare [5, 6]. Up until now 14 cases have been reported in the literature (table 1). Only 4 of them had symptoms suggesting lymphoma including fever and nocturnal sweats, while most had a history of weight loss probably due to local effects of the tumor.

Treatment in our case consisted of re-establishing biliary drainage by endoscopic retrograde cholangiography in order to relieve symptoms and to enable chemotherapy, followed by radiotherapy. In most previous cases, biliary drainage was established before starting chemotherapy. To the contrary, Dudgeon and Brower [7] reported 5 patients with obstructive jaundice due to non-Hodgkin's lymphoma who had chemotherapy using attenuated doses of doxorubicin and vincristine without prior biliary drainage, and jaundice disappeared in all. Although based on a small group of cases, the prognosis appears to be relatively favorable as shown in table 1. Of 14 cases found in the literature, 7 were reported to be in complete remission during a mean follow-up of 19 months.

In conclusion, obstructive jaundice caused by primary duodenal lymphoma is a very rare event. In view of the minor risks of biopsy and the important consequences in case of finding a treatable disease like lymphoma, histological clarification should be attempted whenever feasible.

Table 1. Primary duodenal lymphoma with obstructive jaundice

Year	Country	Localization	Age	Sex	Symptoms	Histology	Nonsurgical biliary drainage	Surgery	CT/RT	Follow-up	Ref.
1986	Kuwait	duodenum	24	f	pain, nausea, weight loss	poorly differentiated			RT	exitus 3 months	8
1986	Kuwait (Egyptian)	stomach/duodenum	30	m	pain, weight loss	poorly differentiated lymphocytic	PTCD		RT	lost	8
1986	USA	duodenum	72	m	anorexia, weight loss	diffuse histiocytic	PTCD		CT	exitus 2 months	9
1987	South Africa	duodenum	56	m	pain, anorexia, weight loss	large cell (Western variant)		Roux-en-Y	CT	lost	10
1993	USA	duodenum	35	m	bleeding	poorly differentiated			CT	CR 36 months	7
1994	Australia	papilla	57	f	nausea	MALT		Whipple		CR 26 months	11
1995	USA	duodenum	42	m	pain, weight loss, pancreatitis	large cell	ERCP		CT	CR 21 months	12
1997	USA	papilla	56	f		follicle center	ERCP	Whipple	RT	CR 5 months	13
2001	USA	papilla	67	f	weight loss	T-cell	ERCP	Whipple		exitus 13 months	14
2003	Morocco	duodenum	10	m	nausea	Burkitt			CT	CR 1 month	15
2004	Switzerland	papilla	54	m	anorexia, weight loss	T-cell	PTCD		CT	exitus 2 months	16
2005	Turkey	duodenum	33	m	pain	intermediate grade B-cell		gastroentero- and choledo-choenterostomy	CT	CR 12 months	2
2005	Turkey	duodenum	38	m	itching	diffuse B-cell			CT	exitus	2
2006	Turkey	duodenum	16	f	pain	Burkitt			CT	CR 36 months	17

CT = Chemotherapy; RT = radiotherapy; CR = complete remission.

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