

Primary Malignant Duodenal Tumors

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Fourteen patients with primary malignant duodenal tumors are studied. Twelve patients had adenocarcinomas and two patients had malignant lymphomas. Preoperative diagnostic procedures, such as radiographic study of duodenum with hypotonic duodenography, complete duodenoscopy with biopsy and arteriographic studies are discussed. Early diagnosis is the key for curative surgical treatment since these tumors seem to disseminate rather late. In patients with primary duodenal carcinoma the resectability rate was 66.7%—seven Whipple resections and one segmental resection. Only one patient in our series died after a Whipple resection. There was no mortality after other procedures. The five year survival rate was 14.2%. There is a definite correlation between regional lymph node involvement and survival time. The mean survival period after Whipple resection without lymph node invasion is 56.5 months in our series. The survival period after Whipple resection for adenocarcinoma with regional lymph node invasion (6 months) is identical to the survival after palliative internal derivations for duodenal adenocarcinoma (5.8 months). It is concluded that a positive preoperative frozen section of a regional lymph node should exclude resective procedures (except in patients who hemorrhage) because they have a higher mortality rate as derivative procedures. Fifty per cent of the patients treated for malignant duodenal lymphoma is tumor free two years after a radical surgical therapy combined with chemotherapeutical treatment from the early postoperative period.

P RIMARY MALIGNANT duodenal tumors are uncommon. They represent only 0.3% of all gastrointestinal tumors but about 50% of all small intestinal malignancies.^{4,7,12}

About 700 instances of malignant duodenal tumors, mostly case reports, are described in the literature.¹² These tumors are usually diagnosed in the latter stages of development because of unclear and intermittent initial symptomatology.^{4,5,10} Some patients are initially considered neurotic because of their vague intermittent symptomatology.⁴ Symptoms of duodenal obstruction and jaundice are infrequent and depend on the location within the duodenum and also on the characteristics of the tumor itself (growth and size).^{2,3,12} The late diagnosis as well as the localization of these tumors make a curative surgical treatment often impossible. Another

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problem with primary malignant duodenal tumors is the fact that they are frequently grouped together with all other periampullary tumors.⁶ In some reports in the literature 56% of the duodenal carcinomas are in fact pancreatic or biliary duct neoplasms.²

The purpose of this study is to elucidate the diagnostic and therapeutic aspects, as well as the prognosis of primary malignant duodenal tumors. This will make a comparison with the results obtained in patients presenting small intestinal tumors and other periampullary tumors possible.

Clinical Material

This study reviews 14 patients with primary malignant duodenal tumors. There were nine males and five females between 41 and 70 years included in the study (Table 1). The mean age at laparotomy was 53 years.

The diagnosis was based on pre-, per-, and postoperative data and was always confirmed histologically. Ampulloma's, tumors of the pancreatic head and terminal biliary duct neoplasms were excluded from this series.

Signs and Symptoms

The mean duration of symptoms in our patients was 10.4 months. Four of our patients were treated symptomatically or conservatively for more than one year before admission. In two of these patients a postbulbar and a stress ulcer were primarily diagnosed. Jaundice was seen in ten patients, gastrointestinal hemorrhage, weight loss and epigastric or upper abdominal pain in nine of our patients. One patient with malignant lymphoma presented with a malabsorption syndrome (Table 1).

Diagnostic Studies

The preoperative upper gastrointestinal series were mostly diagnostic. Suggestive images were confirmed

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TABLE 1. Primary Malignant Duodenal Tumors

Pat.	Age, Sex	Dur. Sy.	Ict.	Bleed.	Obstr.	W.L.	Pain	Local.	R.L.N.	Dist. M.	Therapy	Morbidity	Survival
Adenocarcinoma													
1 CA	47, f	12 m	-	+	-	+	+	II	-	-	WH	Cholangitis	>96 m
2 GM	60, f	2 m	+	+	+	+	-	II	-	-	WH	No	>78 m
3 HE	47, m	9 m	+	-	-	+	-	II	-	-	WH Ant. res.	No Sigm. ca.	30 m
4 BS	51, f	10 m	+	-	-	+	-	II	-	-	WH	No	>22 m
5 MP	60, m	1 m	-	+++	-	-	+	III	-	-	Seg. res.	Jej. fistula	20 m
6 DJ	70, m	4 m	+	+	-	-	+	III	+	-	WH	Pan. fistula	7 m
7 SA	60, f	8 m	-	+++	+	+	+	II	+	+	WH	Pan. fistula	5 m
8 CJ	54, m	1 m	+	-	-	-	+	III	+	-	WH	Sept. perit. Pan. fistula Bronchopn.	Postop.
9 RP	41, m	13 m	+	+	+	+	-	II	+	+	Deriv.	No	11½ m
10 MJ	48, m	4 m	+	-	-	-	+	II	+	+	Deriv.	No	6 m
11 DJ	70, m	1 m	+	++	-	-	+	I	+	+	Deriv.	No	3 m
12 SR	43, f	18 m	+	+	-	+	-	II	+	+	Deriv.	No	3 m
Lymphoma													
13 CP	44, m	3 m	+	-	-	+	+	dif	+	+	WH + CH	No	>24 m
14 DP	43, m	5 y	-	+	-	+	+	dif	+	+	DER + CH	No	2 m

W.L.: weight loss.

*: malabsorption.

by hypotonic duodenography. In two patients (14%) the x-ray examination was not conclusive; duodenoscopy with biopsies confirmed the malignancy in these cases. The literature states that upper gastrointestinal studies may be false positive or negative in about 20–30%.^{1,7,11,12} Table 2 shows the pathology which, in our opinion, may suggest a duodenal tumor image.

Peroperative Diagnosis and Histology

Adenocarcinoma was found in 12 patients and a malignant lymphoma was found in two patients. In eight patients the adenocarcinoma was localized in

the second part of the duodenum (Fig. 1), in three patients in the third part and in one patient in the first part of the duodenum. In both patients with lymphoma, there was a diffuse spread of disease in the duodenum. Fifty per cent of all patients presented distant metastases whereas nine of the 14 patients had local lymph node invasion (65%).

TABLE 2. Differential Diagnosis in Malignant Duodenal Tumors

Prolapsing gastric mucosa
Gallbladder dilatation or gallbladder impression by local peritonitis
Postbulbar ulceration
Chronic pancreatitis
Incancerated cholelithiasis with oedematous prolapsing Papilla of Vater
Spec./aspec. duodenal inflammation
Plica longitudinalis
Diverticulum
Periduodenal lymphnode
Retroperitoneal or intramural haematoma following (old) duodenal trauma
Annular pancreas
Brunner gland hyperplasia

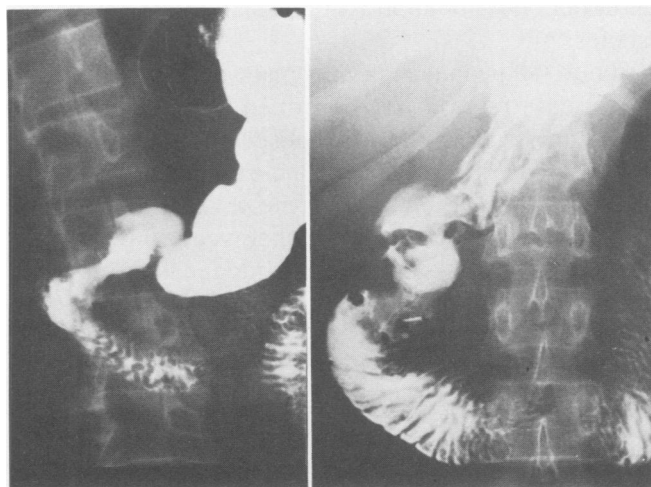


FIG. 1a. X-ray series showing a multinodular infiltrating duodenal process (Adenocarcinoma; Patient 1 in Table 1).

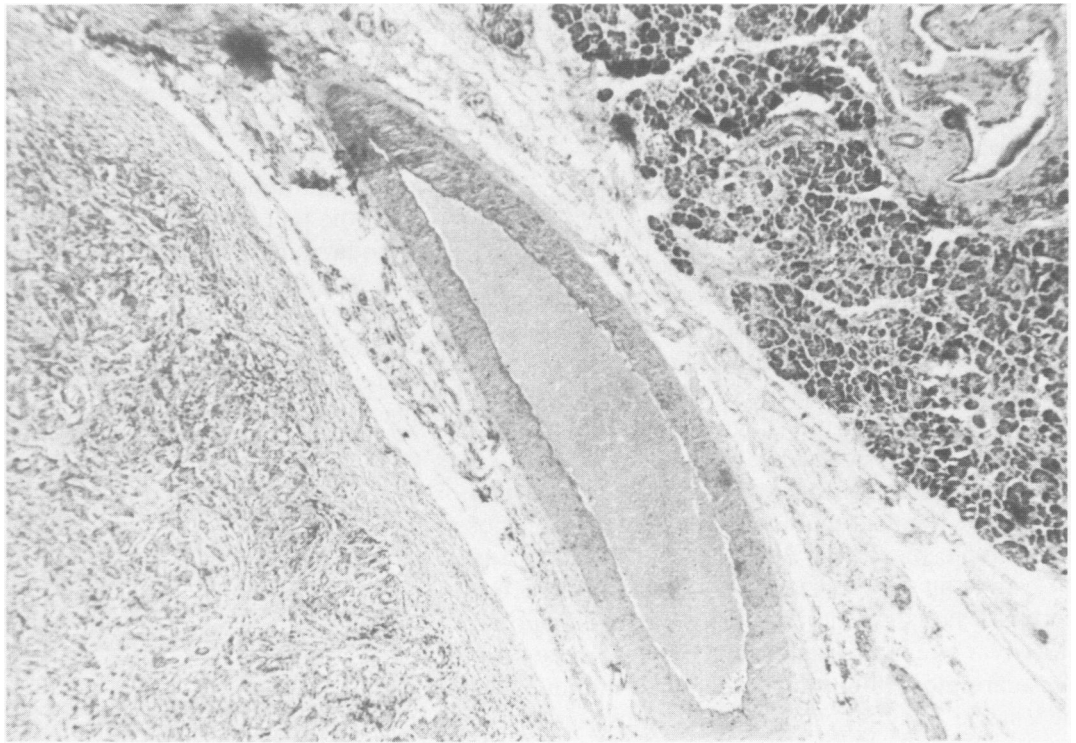


FIG. 1b. Microscopic examination in the same patient. Adventitial tissue separates normal pancreatic tissue on the right from an invasive duodenal adenocarcinoma on the left.

Surgical Treatment

Operative Procedure

We performed a surgical resection in nine of the fourteen patients (64.3%). Eight patients underwent a hemipancreaticoduodenectomy, *i.e.* Whipple resection (six curative and two palliative). A segmental resection of the third part of the duodenum was performed in one patient; in seven of our 14 patients the resection was curative (six Whipple resections and one segmental resection). In the five other patients a gastrointestinal and/or biliodigestive anastomosis was performed.

Additional Treatment

Both cases of malignant lymphoma were additionally treated with chemotherapy which was started during the early postoperative period.

Postoperative Morbidity

The overall postoperative morbidity was 28.5% (4:14 patients). Three pancreatic fistulas were observed after Whipple resection and one jejunal fistula after segmental duodenal resection. One patient (Patient 1) presented recurrent cholangitis, intrahepatic cholestasis and calculus formation caused by stenosis of the

hepaticojejunostomy. This patient underwent a hepaticojejunoplasty six years after the Whipple resection.

Postoperative Mortality

One patient (Patient 8) died of a pancreatic fistula, bilateral bronchopneumonia and septic peritonitis 30 days after pancreatoduodenectomy despite reinterventions. The operative mortality of the global series is 7.1% (1:14 patients) and 12.5% in the Whipple series (1:8 patients).

Survival After Surgical Procedures

The mean survival period of the entire group is 23 months (four patients are still alive). Two patients are alive more than five years after surgical treatment (14.2% five year survival rate). The mean survival after Whipple resection for adenocarcinoma is now 39.6 months (three patients alive) and more than 24 months in one patient with duodenal lymphoma (the patient is still alive). The survival period after Whipple resection for adenocarcinoma without regional lymph node involvement is now 56.5 months (three of four patients alive; one patient died six months after resection for sigmoid adenocarcinoma), in contrast with six months (all patients died) after a palliative Whipple resection for duodenal adenocarcinoma.

One patient who underwent a segmental resection of the third part of the duodenum for adenocarcinoma survived 20 months.

Discussion

This study confirms that primary malignant duodenal tumors are often diagnosed late (on average 10.5 months after initial symptoms).^{4,5,10} Diagnosis is primarily based on upper gastrointestinal contrast medium radiographic studies. When a duodenal tumor is suspected, a duodenoscopy with biopsy should be performed. This should extend throughout the whole length of the duodenum.¹⁰ Definitive diagnosis should always be confirmed by histological examination. Two years ago we introduced systematically coeliac and superior mesenteric angiographies in all patients proposed for Whipple resection. Resectability, vascular anomalies and the different types of inflammatory or tumorous lesions can be judged by this examination.¹⁻³ Moreover late radiographic images of the venous filling phase are most important to demonstrate an eventual mesenteric and/or portal vein compression or tumor invasion. In all of our cases the preoperative angiographic findings were confirmed peroperatively. Cavography, although not performed in our series, may also give useful information about tumor invasion. Sonography and CAT-scanning complete the preoperative investigation of these patients.

The resectability of primary malignant duodenal tumors is about 60–70%.^{8,9,12} Our 64.3% resectability rate for primary malignant duodenal tumors is much higher than the resectability rate in our series of malignant cephalic pancreatic tumors (20.6%), slightly lower than in our series of primary malignant small bowel tumors (76%) and significantly lower than the resectability rate in our series of ampullomas (88.2%).

The five year survival rate of primary malignant duodenal tumors remains low (10–20%).^{8,9,12} The mean survival period after Whipple procedure for primary malignant duodenal tumors is about two years.⁸ Some authors report a five year survival of about 40%;⁵ however this result has not been confirmed in the literature. In our series we have a mean survival rate of 39.6 months after Whipple procedure. There is a good correlation between the survival period and the presence of regional lymph node involvement. In cases with lymph node invasion the survival period was six months

in contrast with 56.5 months in patients without lymph node invasion. Palliative biliary and/or gastrointestinal derivations for primary duodenal adenocarcinoma have a mean survival time (5.8 months) which is comparable with that after Whipple resection in the presence of regional lymph node invasion. These findings as well as the fact that the five year survivals after segmental and Whipple resection are comparable¹² indicate that a Whipple resection should not be performed in the presence of regional lymph node invasion. Consequently, the first step of surgical intervention for duodenal adenocarcinoma should be a frozen section examination of the lymph nodes at the superior mesenteric artery, hepatic artery and truncus coeliacus.

The use of radio- and chemotherapy is confined to malignant lymphomas.^{3,9} In one of our patients with a malignant lymphoma there was no further tumor activity two years after a Whipple resection and administration of chemotherapy, despite initial tumoral invasion of the renal capsule.

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