European Journal of Surgical Oncology xxx (2018) 1–6

Contents lists available at ScienceDirect

European Journal of Surgical Oncology

journal homepage: www.ejso.com

Pancreaticoduodenectomy in the surgical management of primary retroperitoneal sarcoma

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ARTICLE INFO

Article history:
Accepted 15 January 2018
Available online xxx

Keywords:
Pancreaticoduodenectomy
Retroperitoneal sarcoma
Liposarcoma
Leiomyosarcoma

ABSTRACT

Background: In retroperitoneal sarcoma (RPS), the optimal extent of resection must balance adequate disease control with potential for morbidity. We sought to study the frequency and outcomes after a Whipple procedure or pancreaticoduodenectomy (PD) in patients undergoing resection for primary RPS.
Methods: Participating referral centers within the Trans-Atlantic Retroperitoneal Sarcoma Working Group provided retrospective data from January 2007 to December 2016 for patients with primary RPS who underwent PD along with the total number of consecutive resections done during the same time period. Data from participating centers were combined for analysis.
Results: In total, 29 patients underwent PD among 2068 resections performed for primary RPS (1.4%). The predominant histologic subtypes were liposarcoma and leiomyosarcoma. All PD patients underwent concomitant resection of additional organs (median: 2, range: 1–5), including 13 patients (45%) who also received vena cava resection. Definitive evidence of microscopic invasion of the duodenum or pancreas was seen in 84% of patients. Postoperatively, 10 patients (34%) had major complications including 8 (28%) that developed a clinically-significant pancreatic leak. One postoperative death (3.4%) occurred. With a median follow-up of 4.8 years, 19 patients (66%) developed disease recurrence. The patterns of recurrence were dependent on histologic subtype.
Conclusion: Although infrequent, when PD is done for primary RPS, resection of additional organs is often required and major complication rates are moderate. The recurrence rate is overall high and the pattern of recurrence is dictated by histologic subtype.

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https://doi.org/10.1016/j.ejso.2018.01.086
0748-7983/

Introduction

Soft tissue sarcomas are rare malignancies that together represent only 1% of all solid cancers in adults. Although soft tissue sarcomas can develop at any anatomic location, 15–20% occur in the retroperitoneum [1,2]. Surgery is the mainstay of treatment and the only chance for cure; however resection of retroperitoneal sarcoma (RPS) is often challenging due to the large size of the tumors and the potential involvement of major organs and vessels. Complete resection of RPS is critical. The optimal extent of resection in RPS must balance adequate oncologic disease control with morbidity, including the potential for complications. Although there are some retrospective data to support the utility of radiation therapy for local control [3,4], results of an ongoing prospective trial (STRASS) are still pending. The role of systemic therapy in RPS is currently limited [5,6].

The Whipple procedure or pancreaticoduodenectomy (PD) is in itself a complex operation most frequently performed for adenocarcinoma or neuroendocrine tumors affecting the head of the pancreas or duodenum. In RPS, this operation may be indicated for right-sided tumors that abut these structures. In some cases the need for PD may be obvious preoperatively based on symptoms (e.g. bleeding, obstruction) or clear evidence of invasion by cross sectional imaging or endoscopy (Fig. 1). For the majority of RPS patients however, obvious organ involvement is not seen and the decision to perform PD is made intraoperatively. When performed, the combination of PD with RPS resection significantly increases the complexity and magnitude of the operation. To date there are only case reports and small case series [7–11] of this unique cohort of patients.

In 2013, the Trans-Atlantic Retroperitoneal Working Group (TARPSWG) was formed as a multi-institutional, international collaboration dedicated to improve our understanding of RPS and to optimize the treatment of this rare disease [12]. Several original studies and consensus guidelines have been published by the group to help guide the surgical management of RPS [13–17]. In the current study, through the combined efforts of TARPSWG referral centers, we were able to collectively identify a substantial number of primary RPS cases in which PD was performed during resection. We sought to define the frequency of these cases and to study the outcomes after surgery.

Methods

Participating TARPSWG centers – each a sarcoma referral center - were asked to identify and provide study cases of patients with primary RPS who underwent PD as a component of their RPS resection, from January 2007 to December 2016. Each center also provided data for total number of primary RPS resections done over the same time period (= denominator) and the total number by histologic subtype. With this design, patients who were unresectable were not included. Appropriate ethical and institutional approvals for this study was obtained by each participating center.

In each study case (= PD during primary RPS resection), clinicopathologic data were collected including patient age, sex,

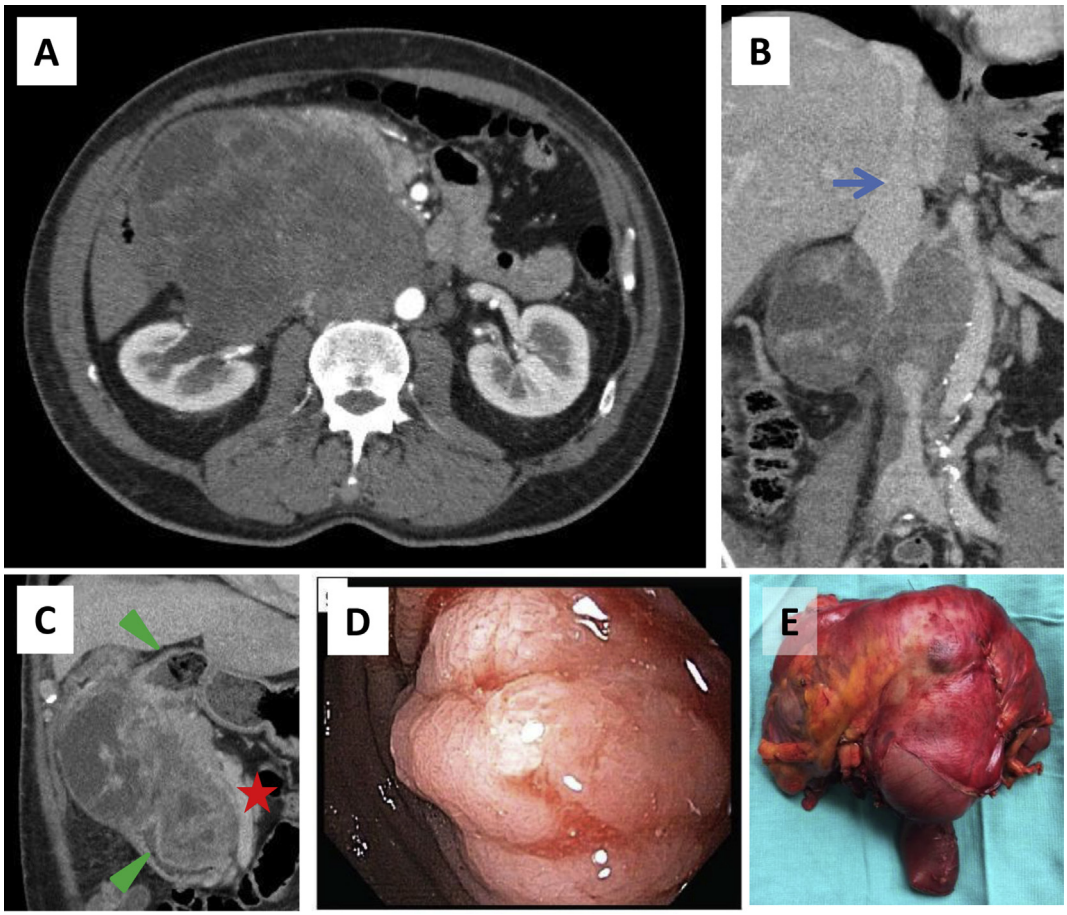

Fig. 1. Case example of a patient with retroperitoneal leiomyosarcoma who required pancreaticoduodenectomy for complete resection. A-C. Tumor involvement of adjacent organs was evident by preoperative cross sectional imaging. The tumor involved the inferior vena cava (arrow), duodenum (arrowheads) and head of pancreas, coming close to the superior mesenteric vessels (star). D. Endoscopic view of tumor invasion of the duodenum. E. Gross tumor after en bloc complete resection.

Please cite this article in press as: Tseng WW, et al., Pancreaticoduodenectomy in the surgical management of primary retroperitoneal sarcoma, European Journal of Surgical Oncology (2018), https://doi.org/10.1016/j.ejso.2018.01.086