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Algorithmic Approach

Periampullary adenocarcinoma (PAC) is a clinicopathologic entity comprised of four distinct subtypes in one high-density anatomic region. Formally defined as tumors arising ≤ 2 cm from the ampulla of Vater, PAC can originate from the pancreatic head, duodenum, distal bile duct, or ampulla of Vater [1]. Pancreatic ductal adenocarcinoma (PDAC) is the most common PAC with a recent longitudinal study demonstrating its occurrence at 66% compared to 16%, 12%, and 6% for ampullary, biliary, and duodenal adenocarcinomas, respectively [2]. Though not a common tumor type, PAC has low overall survival rates that differ significantly by location of origination [3]. To illustrate, one series reported primary site-specific 5-year PAC survival rates of only 17% for pancreas, 23% for bile duct, 37% for ampulla, and 51% for duodenum [4]. The high and variable mortality of PAC is likely attributable to stage at presentation and inherent biologic differences in pancreatobiliary versus intestinal histology [1, 5].

Diagnosis and Preoperative Evaluation

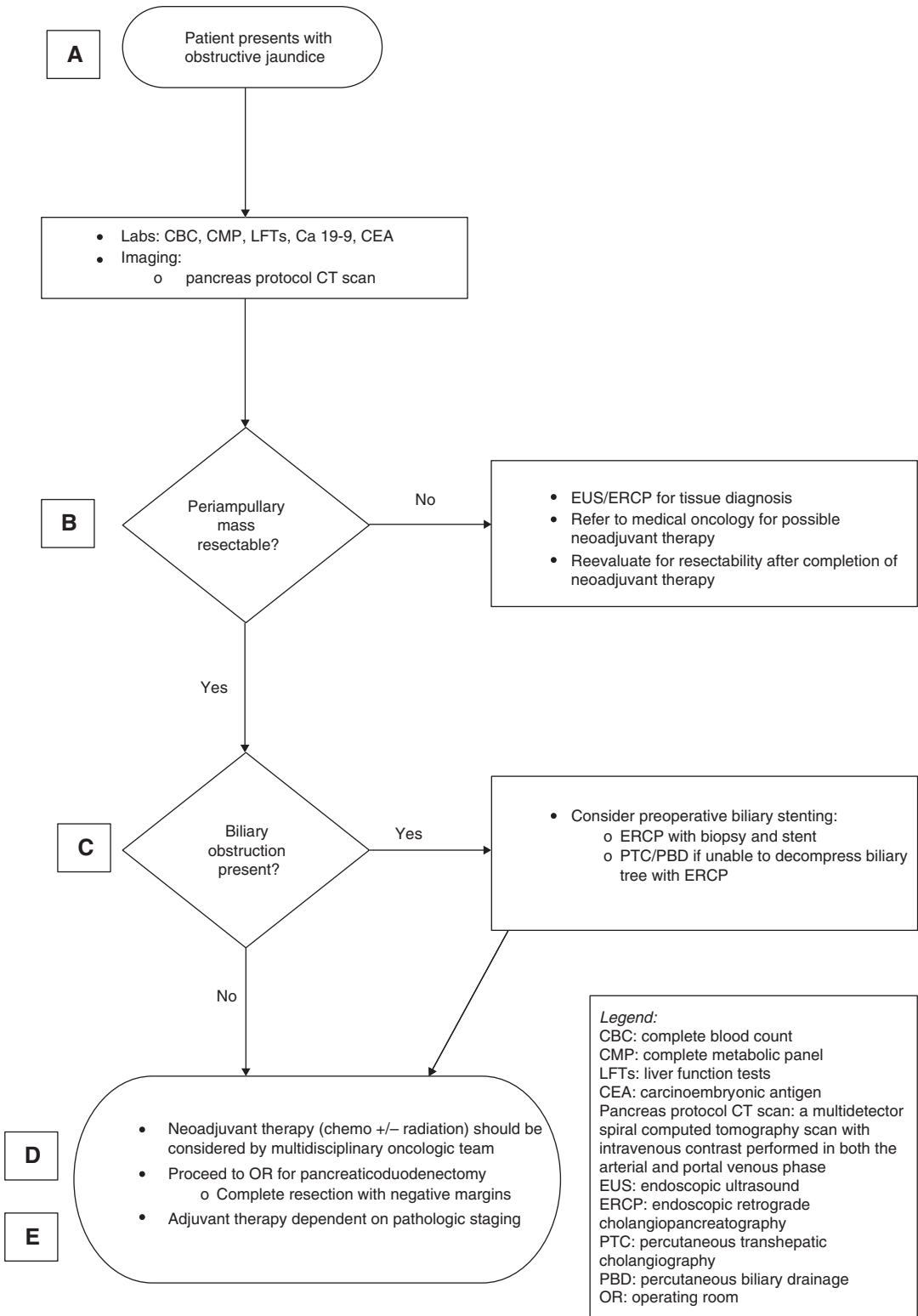
- A. Patients most frequently present with obstructive jaundice possibly accompanied by vague abdominal pain, nausea, and weight loss [6]. PAC should always be considered when evaluating a patient with obstructive jaundice, and basic workup of laboratory studies including liver function tests (LFTs), cancer antigen 19-9 (CA 19-9), carcinoembryonic antigen (CEA), and cross-sectional imaging should be obtained [6–8]. A multidetector spiral computed tomography scan with intravenous contrast performed in both the arterial and portal venous phase (pancreas protocol CT) is ideal to evaluate for a periampullary mass [7]. Findings on pancreas protocol CT are critically important to determining the next steps in the patient's management.
- B. Recent advances in radiographic technology now facilitate assessment of many preoperative staging factors necessary to determine resectability of a periampullary mass, including involvement of mesenteric vessels [9].

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Resection and Clinicopathologic Staging

As surgical resection remains the best chance of cure for PACs, the time from identification of the mass to operation should be optimized [2]. Proceeding to surgery based on pancreas protocol CT findings alone is acceptable, but the clinical scenario may require further workup [7].

- C. Biliary drainage procedures are typically recommended only in cases of cholangitis, substantially elevated bilirubin, or prolonged period of elevated bilirubin [10, 11]. Preoperative tissue diagnosis is not necessary if there is a high index of suspicion for cancer on cross-sectional imaging and immediate operative intervention is planned. However, tissue diagnosis is necessary if neoadjuvant therapy is considered, as is frequently the case for PDAC [12, 13].
- D. The standard procedure for PAC is pancreaticoduodenectomy (PD) with the goal of complete tumor resection with negative oncologic margins [2]. Full descriptions of the operative technique and complications of PD are out of the scope of this chapter. Clinicopathologic staging is based on the American Joint Committee on Cancer (AJCC) Cancer Staging Manual for each location of PC origination [14]. Key factors are resection margins, nodal involvement, microvascular invasion, and perineural invasion [14].
- E. As chemotherapy and immunotherapy continue to improve for PAC, further important features are histomolecular markers such as KRAS and DPAC that are more prevalent in PDAC and offer opportunity for targeted therapy [15]. Multidisciplinary oncologic care is essential along the continuum of treatment [16].



Algorithm 98.1

Conclusion

PAC consists of pancreatic, biliary, ampullary, and duodenal carcinoma which all present with similar symptoms but have variable survival

rates depending on site and specific histology. Pancreas protocol CT scan is the most useful imaging modality, and surgical resection with pancreaticoduodenectomy offers the best chance for cure.

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