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Preoperative Treatment With FOLFIRINOX and Successful Resection for a Patient With Mixed Acinar-Endocrine Carcinoma of the Pancreas

Motokazu Sugimoto, M.D.,

Department of Surgery, UCLA Medical Center, Los Angeles, California

O. Joe Hines, M.D.,

Department of Surgery, UCLA Medical Center, Los Angeles, California

David W. Dawson, M.D.,

Department of Pathology and Laboratory Medicine, UCLA Medical Center, Los Angeles, California

V Raman Muthusamy, M.D.,

Division of Digestive Diseases, UCLA Medical Center, Los Angeles, California

Howard A. Reber, M.D., and

Department of Surgery, UCLA Medical Center, Los Angeles, California

Timothy R. Donahue, M.D.

Department of Surgery, UCLA Medical Center, Los Angeles, California

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To the editor

Mixed acinar-endocrine carcinoma (MAEC) of the pancreas is an extremely rare entity. Therefore, the biological behavior of the MAEC of the pancreas has not been clearly determined and is generally considered as clinically aggressive. We report a case of a 48-year-old man with MAEC who underwent pancreaticoduodenectomy after neoadjuvant chemotherapy.

A 48-year-old man was referred to our institution for a large mass within the head of the pancreas. He was in his usual state of health until he developed severe abdominal pain a few weeks prior to presentation. A pancreas-protocol, contrast-enhanced CT showed a 7.7×5.9 cm enhancing solid mass with cystic portions in the pancreatic head and uncinate process (Fig. 1A). On the CT, the mass abutted the superior mesenteric vein (SMV) without narrowing it and also appeared to abut the posterior aspect of the superior mesenteric artery (SMA) (Fig. 1B). The patient underwent endoscopic ultrasonography (EUS) with fine needle aspiration (FNA) of the mass. This revealed a malignant epithelioid neoplasm with both acinar and endocrine features. Immunohistochemical staining of the tumor cells showed

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Corresponding to: Timothy R. Donahue, 10833 Le Conte Ave, CHS 72-215, Box 956904, Los Angeles, CA 90095, USA, Tel: 310-206-7440, Fax: 310-206-2472, tdonahue@mednet.ucla.edu.

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strong positivity for trypsin and synaptophysin. The Ki-67 labeling index was 10–20%. No distant metastasis or lymphadenopathy was suggested on either the CT or EUS. Serum carbohydrate antigen 19–9 (CA 19–9) level was 374 U/ml.

Given the size and potential vascular involvement of the tumor, the patient received 6 cycles of FOLFIRINOX without any adverse effects. On the follow-up CT, the tumor showed a significant decrease in size to 1.9×1.4 cm (Fig. 1C). The tumor appeared to have retracted away from the SMV (Fig. 1D). There was still some soft tissue density on the CT scan involving the posterior aspect of the SMA, however, no evidence of metastatic disease or lymphadenopathy was seen. His serum CA 19–9 level had decreased to 29 U/ml. Since downstaging of the tumor was confirmed at this point, he underwent exploratory laparotomy and a pylorus-preserving pancreaticoduodenectomy. No vascular resection was required. The patient recovered uneventfully and was discharged on postoperative day 6.

Pathological examination with immunohistochemistry of the surgical specimen was consistent with MAEC of the pancreas: it was diffusely positive for trypsin and synaptophysin. The gross tumor size was 2.0 cm. The tumor cells invaded the retroperitoneal soft tissue. Margins were negative. No metastasis was detected within 34 harvested lymph nodes. Neither perineural invasion nor lymphovascular invasion were observed. Mild treatment response was seen. At last follow-up, the patient was alive 21 months later without any evidence of recurrence.

DISCUSSION

MAEC of the pancreas is an extremely rare entity, with only 30 resected cases reported in the English literature.¹ The criteria for diagnosis of MAEC is that more than 25% of each component of acinar and endocrine cells are present in the specimen. Immunohistochemistry demonstrates the presence of each component: trypsin, chymotrypsin, lipase, or periodic acid-Schiff as an acinar marker and chromogranin A, synaptophysin, CD56, or neuron-specific enolase as an endocrine marker. Although MAECs show both exocrine and endocrine differentiation, they usually predominantly consist of acinar cell cancer (ACC) and considered clinically as a variant of ACC.² Therefore, optimal treatment strategy is surgical resection if resectable for the localized disease, conforming to the treatment for pancreatic ductal adenocarcinoma or ACC.¹ Response to chemotherapy has reportedly been poor.³ The median overall survival was calculated to be 12 months after surgical resection for the 21 cases with the information available from published papers.¹

In our case, the patient was diagnosed as MAEC by EUS with FNA. Based on the borderline resectability on CT, 6 cycles of FOLFIRINOX as neoadjuvant therapy were administered and resulted in significant reduction of the tumor size and retraction of the tumor from the surrounding vessels (SMV and SMA) on CT. Potentially curative surgical resection to negative margin was performed. FOLFIRINOX has been used as a standard regimen of neoadjuvant therapy for pancreatic ductal adenocarcinoma, but it has also been reported to be effective for unresectable ACC in a few papers.⁴ Use of FOLFIRINOX for the patients with MAEC or ACC may be appropriately considered as a choice of downstaging therapy for patients with borderline resectable or locally advanced disease.

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Fig. 1.

A) An initial CT showed a 7.7×5.9 cm tumor located in the pancreatic head and uncinate process. B) On the initial CT, the mass abutted the superior mesenteric vein (SMV) without narrowing it and also appeared to abut the posterior aspect of the superior mesenteric artery (SMA). SV; splenic vein. C) On the followup CT after 6 cycles of FOLFIRINOX, the tumor showed a significant decrease in size to 1.9×1.4 cm. D) On the followup CT, the tumor appeared to have retracted away from SMV. There was still some soft tissue density involving the posterior aspect of the SMA.