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Case report

Multimodal approach and long-term survival in a patient with recurrent metastatic acinar cell carcinoma of the pancreas: A case report

Sarah F. Jauch, Van K. Morris, Corey T. Jensen, Ahmed O. Kaseb*

The University of Texas MD Anderson Cancer Center, 1515 Holcombe Boulevard, Unit Number: 426, Houston, TX 77030, USA

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ABSTRACT

Pancreatic acinar cell carcinoma is an uncommon neoplasm of the exocrine pancreas associated with a poor prognosis, especially when found to be metastatic. Since there are a lack of large studies and prospective, randomized data, no consensus treatment guidelines are available. Here, we report a case of a patient with recurrent metastatic acinar cell carcinoma involving the liver who had presented initially with pancreatic panniculitis. She received chemotherapy with capecitabine and oxaliplatin prior to resection of her primary tumor and liver metastases, after which she experienced a 30 months recurrence-free survival. Upon relapse, she was treated with a combination of capecitabine and oxaliplatin followed by maintenance capecitabine. Now, more than seven years after initial diagnosis, the patient remains stable without evidence of active disease. This case highlights the possibility of therapeutic success even for a patient initially deemed unresectable due to a poor performance status who responded to fluoropyrimidine-based therapy.

Introduction

Pancreatic cancer is the fourth leading cause of cancer mortality in the United States [1–3], with approximately 40,000 deaths estimated in 2015 [1]. The vast majority of pancreatic neoplasms arise from the exocrine pancreas, and over 85% of all pancreatic malignancies are pancreatic ductal adenocarcinomas (PDAC) [4]. However, acinar cell carcinoma (ACC) is a rare subgroup of 1–2% of pancreatic exocrine tumors in adults [4,5] with a prevalence of less than one per million in the United States [6]. The diagnosis of ACC is challenging due to various morphologic characteristics [7] and non-specific clinical symptoms [5,6,8–11]. A distinctive but rare syndrome called Schmid's Triad is characterized by subcutaneous fat necrosis, eosinophilia and polyarthralgia, secondary to lipase hypersecretion [11]. Associated subcutaneous nodules can be misdiagnosed as erythema nodosum [10]. The overall five-year survival of pancreatic ACC from prior reports has ranged from 6 to 50% [5,6,11–13]. These series have suggested that survival outcomes in ACC may be more favorable than for patients with adenocarcinoma [6,12]. In the absence of prospective data for this rare disease, no standard therapeutic approach exists [14]. Surgery is the treatment of choice for patients with localized disease [10]. In addition, chemotherapeutic agents established in PDAC and colorectal carcinoma are often used [14].

We report a multimodal approach in a patient diagnosed with stage IV pancreatic ACC who remains without evidence of active disease on maintenance capecitabine.

Case report

In June 2007, a 61-year-old Caucasian woman with a past medical history notable for vasculitis presented with partially ulcerated, erythematous/violaceous subcutaneous nodules and swelling throughout her lower extremities. With a presumed diagnosis of erythema nodosum (given her aforementioned history of autoimmune disease), she was started empirically on prednisone yet developed rapid progression of her skin lesions. Biopsy of the skin demonstrated septal panniculitis with areas of necrosis. Infection was excluded as an etiology with negative blood and wound cultures. Further diagnostic evaluation included a computed tomography (CT) scan (Fig. 1A + B), which detected a

* Corresponding author. Tel.: +1 713 792 2828; fax: +1 713 563 0541.
E-mail address: akaseb@mdanderson.org (A.O. Kaseb).

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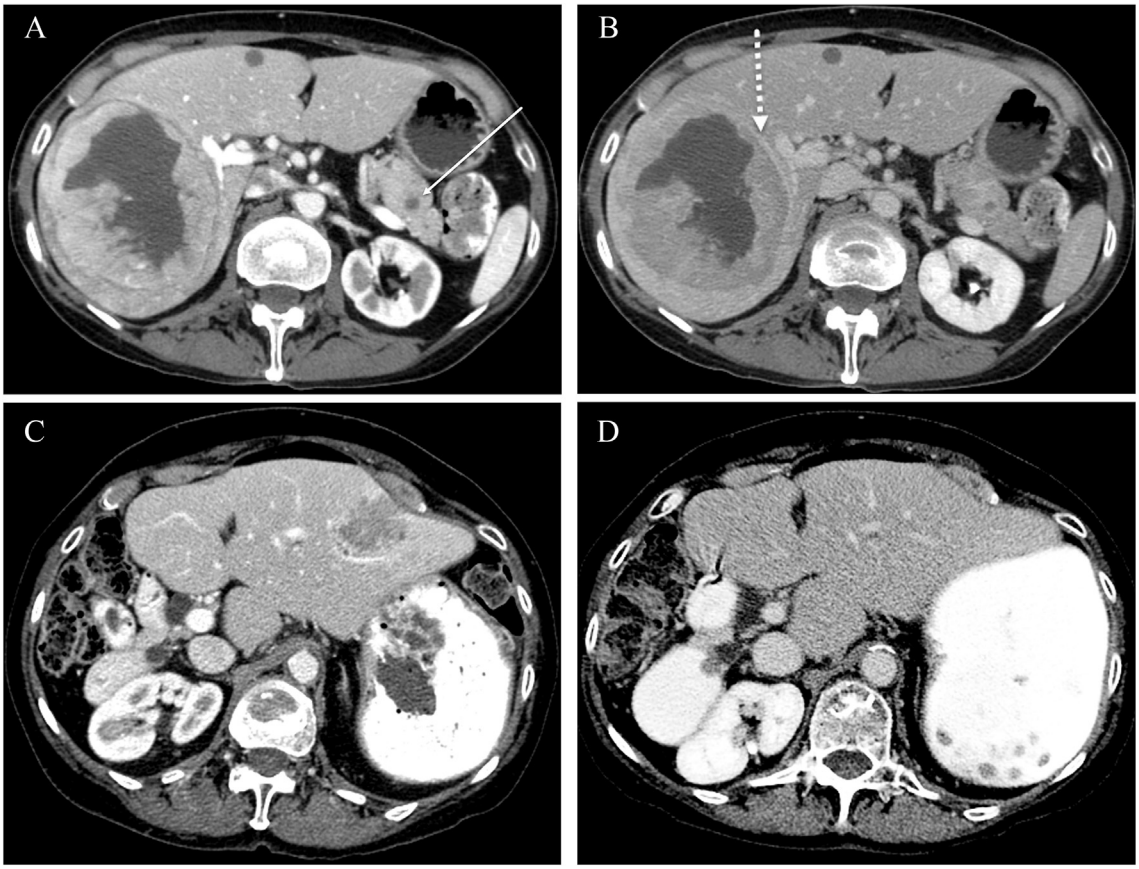

Fig. 1. Abdominal CT scans at diagnosis demonstrate the A) pancreatic tail mass with small cystic change (arrow) and the dominant hypervascular hepatic metastasis with B) washout and pseudocapsule (dashed arrow) on delayed imaging. Three years later, C) recurrent tumor was identified in the liver with D) subsequent complete response to therapy.

3.1 × 2.4 cm pancreatic mass between the gastric fundus and distal pancreatic tail. In addition, numerous bilobar liver metastases, the largest in the right lobe measuring 10 × 8 cm, were noted. CT guided biopsy of a suspected liver metastasis was consistent with acinar cell carcinoma. Pathological analysis of the tumor revealed strong immunohistochemical staining for chymotrypsin, lipase, alpha-1 antitrypsin, and pankeratin; and scattered positivity for carbohydrate antigen 19-9 (CA 19-9) and trypsin. Neuron-specific enolase (NSE), synaptophysin, chromogranin and beta-catenin were all negative.

Subsequently, the patient was referred to our institution for further recommendations of metastatic pancreatic ACC. Upon initial evaluation, she was noted to have an Eastern Cooperative Oncology Group (ECOG) performance status of 3 and to be underweight with a body-mass index (BMI) of 17.3 kg/m^2. Her physical exam was otherwise notable for violaceous lesions across her abdomen and lower extremities. Laboratory results showed increased CA 19-9 of 345.6 U/mL, alpha-fetoprotein (AFP) of 343.5 ng/mL, lipase of 7459 U/L, albumin of 3.3 mg/dL, and lactate dehydrogenase (LDH) of 865 IU/L. Carcinoembryonic antigen (CEA), amylase, total bilirubin, alanine aminotransferase (ALT) and aspartate aminotransferase (AST) were all within normal limits. Because of her unfavorable performance status and her poor nutritional status, the patient was not considered to be a candidate for surgical resection. Rather, she received eight cycles of chemotherapy with capecitabine plus oxaliplatin (XELOX). After her first three cycles, laboratory analyses revealed normalization of her CA 19-9, lipase, and LDH laboratory values. After eight cycles, the panniculitis had resolved completely, and both her ECOG performance status and her BMI had improved, findings compatible with clinical improvement. Imaging studies confirmed resolution of the primary tumor and reduction in the size of her right hepatic lesions. Given this profound response to XELOX chemotherapy, surgical evaluation was reassessed, and four months after completion of chemotherapy, the patient underwent distal pancreatectomy, splenectomy, cholecystectomy and right hepatectomy. Pathology revealed a residual moderately differentiated ACC in the pancreas measuring 6 mm in diameter with negative surgical margins, one of eleven regional lymph nodes with tumor, and six tumor nodules in the liver, with the largest containing 30% viable tumor cells (ypT1N1M1, AJCC 6th edition). Due to her slow recovery after surgery and residual sensory neuropathy from oxaliplatin, the patient did not receive adjuvant chemotherapy. She remained disease free for the following 30 months, at which point she was found to have a newly elevated lipase (1111 U/L) and new liver lesions concerning for recurrent metastases (Fig. 1C). Given her initial response to XELOX, this regimen was reintroduced with imaging studies after three cycles showing a response in all measurable lesions. After a total of six cycles of XELOX, the oxaliplatin was dropped, and she has continued on single-agent capecitabine (1000 mg/m^2/day) for the past three and a half years. Most recent imaging studies show complete eradication of all disease with no evidence of macroscopic tumor visualized (Fig. 1D). Her performance status is now 0, and her previously mentioned elevated laboratory values are all within normal limits.

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