Answer

Acinar Cell Carcinoma

Acinar cell carcinoma (ACC) of the pancreas is an uncommon malignancy that accounts for 1% of pancreatic exocrine malignant tumors. This malignancy has a slight male preponderance with a peak age in the seventh decade of life.[1] A specific syndrome of subcutaneous fat necrosis (pancreatic panniculitis) and polyarthralgias is encountered when excessive lipase is secreted from the tumor, as manifested by our patient. Additional endocrine symptoms may be seen because of increased serum levels of insulin, insulinlike growth factors, and glucagon. The tumor size at presentation tends to be larger for ACC than pancreatic adenocarcinoma and typical findings on computed tomography are a solitary, well-defined, heterogenous, hypodense mass with a well-defined enhancing capsule.[1-3]

Acinar cell carcinomas of the pancreas are highly cellular tumors that have 4 well-described growth patterns: acinar, cellular, trabecular, and glandular. Characteristically, the tumor cells will show increased cellularity with minimal stroma and have positive immunostaining for periodic acid–Schiff and trypsin.[1]

The single largest institutional case series included 39 patients diagnosed with ACC over a 21-year period.[4] The lipase hypersecretion syndrome was rare even in this small group. Nine patients were noted to have elevated serum lipase levels and only 3 patients had subcutaneous manifestations in this series. However, the association between underlying pancreatic disease and distant cutaneous fat necrosis has been well characterized. In a Mayo Clinic series of 11 patients with pancreatic panniculitis, 6 of the patients had underlying pancreatic carcinoma and 5 patients had underlying pancreatitis. Arthritic symptoms were present in most of the patients and major morbidity and mortality were high among all patients.[5] Skin changes with underlying pancreatic pathology are also seen in patients with glucagomas. In contrast to the deep and focal areas of adipocyte necrosis seen in pancreatic panniculitis, migratory necrolytic erythema of glucagonoma syndrome has superficial mucocutaneous involvement on areas of trauma or friction that grossly appear as macular erythematous annular plaques and microscopically show hyperkeratosis.[6]

Adjuvant therapy is often used and all patients diagnosed with ACC should undergo serum lipase measurement, which may be a useful marker to assess response to therapy. Surgical resection offers the only chance of cure and prolongs survival.[1-4] Compared with pancreatic adenocarcinoma, patients with ACC tend to have a higher resectability rate with 5-year survival rates of 44% to 72% after surgical resection, according to recent reports from large American[7] and Japanese[8] tumor registries.

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