

Acinar cell carcinoma – a rare tumour of the pancreas

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Case report

A 32-year-old man presented with a progressively enlarging, firm, non-tender abdominal mass, more prominent on the left. He was fit and had no significant past medical history. Routine blood tests were all normal (including liver functions, amylase and α -fetoprotein). A computed tomography (CT) scan showed a 10×13×11.5 cm well-defined, solid and cystic mass in the lesser sac, with no evidence of distant spread. At laparotomy, a distal pancreatectomy and splenectomy were performed with a satisfactory plane around the tumour allowing complete macroscopic excision. Histological results were consistent with a pure acinar cell carcinoma. After discussion at a multidisciplinary oncology clinic, he was placed on a trial of gemcitabine-based chemotherapy.

Discussion

Pathology

The acinar cells constitute most of the mass of the pancreas. Acinar cell carcinoma is a rare malignant tumour of the pancreatic exocrine cells; it comprises approximately 1% of malignant pancreatic tumours.¹ The majority are pure acinar cell cancers; however, a minor endocrine component may occur. If the endocrine component constitutes more than 25% of the tumour, it is termed a mixed acinar-endocrine tumour; the clinical behaviour mimics that of the pure form. Rarely, a cystic pattern may predominate, and these are termed acinar cell cystadenocarcinomas.¹ All tumours are characterised by secretion of pancreatic exocrine enzymes (trypsin, chymotrypsin, and lipase).^{1,2} A small subgroup (10%) may exhibit a lipase hyper-secretion syndrome characterised by subcutaneous fat necrosis, polyarthritis and eosinophilia.¹ The characteristic histological features are those of a stroma-poor cellular tumour composed of sheets and small glands (acini) comprising polygonal atypical cells with prominent nucleoli. Trypsin and chymotrypsin immunohistochemistry are almost always positive.^{1,2}

Clinical behaviour

The clinical behaviour of these tumours is believed to fall between that of pancreatic neuroendocrine tumours and pancreatic adenocarcinoma. Among the first series reported, median survival was 18 months in 28 patients from multiple institutions.² In a more recent and larger series of 39 patients from the Memorial Sloane Kettering Institute, the median survival was 19 months. For those with resected lesions, survival was 36 months. Five-year survival was less than 10%.³ Johns Hopkins had 14 patients of whom all had successful resections with a median survival of 33 months. Eight of the 14, however, developed recurrence.⁴

Therapy

Complete excision remains the best treatment. However, the majority (72%) of patients will present with recurrent disease.^{3,4} This raises the question of adjuvant treatment. Again, owing to the small numbers, exact protocols do not exist. Chemotherapeutic regimens are modelled on those with a favourable response in the setting of metastatic disease. The results are disappointing. The MSK series had 18 patients, none had a complete response, and only 2 had a partial response to combined chemotherapy.³ Radiotherapy is recommended for regional irresectable disease. A multidisciplinary approach is necessary to adequately cover the range of treatment required for these rare tumours.

REFERENCES

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