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Roseate and merest-acinar cell carcinoma-pancreas

Abstract

Acinar cell carcinoma pancreas is an epithelial, malignant neoplasm of exocrine pancreas comprised of cells resembling pancreatic acinar cells which are immune reactive to BCL10 and trypsin. Tumefaction arises due to accumulated genetic alterations, chromosomal instability or allelic copy number variation. Cogent clinical symptoms as abdominal pain, dorso-lumbar pain, loss of weight, nausea or vomiting or subcutaneous fat necrosis may ensue. Grossly, a solid, enlarged, well circumscribed, partially encapsulated tumefaction of fleshy consistency is encountered. Neoplastic cells demonstrate granular, eosinophilic cytoplasm pervaded with zymogen granules stainable with periodic acid Schiff's (PAS) stain with diastase resistance, uniform nuclei and a singular, prominent nucleolus. Scanty and fibrous encompassing stroma exhibits foci of perineural and vascular invasion. Acinar cell carcinoma pancreas is immune reactive to keratins as CK7, CK8, CK18, CK19, BCL10, trypsin nuclear beta catenin or CD200. Computerized tomography (CT) and magnetic resonance imaging (MRI) delineates an enlarged tumefaction with well-defined perimeter, an exophytic pattern of tumour evolution and heterogeneous image enhancement. Surgical resection, chemotherapy with gemcitabine or radiofrequency ablation are appropriate modes of therapy.

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Introduction

Acinar cell carcinoma pancreas emerges as a malignant neoplasm of exocrine pancreas. The epithelial tumefaction is comprised of cells demonstrating morphological semblance to pancreatic acinar cells. Tumour cells appear immune reactive to pertinent acinar cell markers as BCL10 and trypsin. Acinar cell carcinoma is additionally designated as acinar cell cysto-adenocarcinoma, mixed acinar neuroendocrine carcinoma or mixed acinar ductal adenocarcinoma. Acinar cell carcinoma pancreas configures ~ 2% of adult pancreatic neoplasms and ~15% of paediatric pancreatic tumours.[1,2]

Acinar cell carcinoma pancreas exhibits a mild male predilection with male to female proportion of 2.1:1. Average age of disease emergence is 60 years. Head of pancreas configures as a frequent site of disease emergence.[1,2] Acinar cell carcinoma pancreas is posited to occur due to cellular accumulation of genetic alterations or occurrence of chromosomal instability a Acinar cell carcinoma nd frequent allelic copy number variation.[3,4] Additionally, acinar cell carcinoma is postulated to arise from factors such as tobacco consumption or cigarette smoking and defective repair of deoxy ribonucleic acid (DNA) (Table 1).

Table 1 Differential Diagnosis of Acinar Cell Carcinoma[3,4]

Differential diagnosis	Gender/age	Imaging/gross features	Histology	Prognosis
Acinar cell carcinoma	Predominant males, mean age 62 years	Solid, bulky, well defined tumour with haemorrhage, necrosis	Predominant solid/acinar architecture, uniform cells, basal nuclei, eosinophilic granular cytoplasm, single, prominent nucleolus, minimal stroma	Aggressive with significant recurrence and metastasis
Pancreatic ductal adenocarcinoma	Mild male predominance, 6th to 8th decade	Solid, poorly defined mass	Large, medium or small malignant ducts, tubular pattern, desmoplastic stroma, mitosis and necrosis	Inferior overall survival
Pancreatic neuro-ectodermal tumour	Equal gender distribution, adult prevalence, mean age 40 years	Solid, well circumscribed, 5% cystic tumours	Variable architecture, uniform cells, oval or spherical nuclei, granular cytoplasm, inconspicuous nucleoli, minimal stroma	Relatively languid, variable outcomes
Solid pseudo-papillary neoplasm	Preponderantly females, average age 28 years	Well defined, encased, cystic degeneration	Pseudo-papillae, cells with hyaline/myxoid stroma, surrounding vessels, large cytoplasmic hyaline globules, nuclear grooves	Low malignant potential. Majority alleviated with surgical resection
Pancreatoblastoma	First decade of life, adults can be affected, mean age 4 years	Partially encapsulated, frequently lobulated, substantial,	Solid & acinar structure, cellular stroma, keratinization of squamoid nests, heterologous mesenchymal elements	Aggressive, superior outcomes in children

Methodology

Molecular alterations as chromosomal mutations within KRAS or SMAD4 genes appear absent. Additionally, recurrent genetic rearrangements within BRAF, RAF1 and RET genes may be discerned.[3,4] A subset of neoplasms are associated with modifications within Wnt signalling pathway in association with APC / CTNNB1 genetic mutations. Majority of neoplasms arise sporadically. However, a minority (< 10%) of tumours are associated with conditions such as Lynch syndrome, familial adenomatous polyposis syndrome or Carney complex.[3,4]

Acinar cell carcinoma pancreas manifests cogent clinical symptoms as abdominal pain, dorso-lumbar pain, loss of weight,

nausea or vomiting. Hyperbilirubinemia is exceptional, in contrast to ductal adenocarcinoma. Extensive metastatic disease accompanying acinar cell carcinoma pancreas may demonstrate symptoms contingent to lipase hypersecretion as subcutaneous fat necrosis.[5,6]

Upon gross examination, a solid, enlarged, well circumscribed, partially encapsulated tumefaction with fleshy consistency is encountered. Upon frozen section, acinar cell carcinoma pancreas appears as a hyper-cellular neoplasm composed of neoplastic cells reminiscent of pancreatic acinar cells. Cytological examination demonstrates hyper-cellular or moderately cellular smears exemplifying dissemination of monomorphic nuclei with prominent nucleoli.[5,6]

Acinar cell carcinoma pancreas is a significantly cellular neoplasm. Neoplastic cells appear incorporated with moderate quantities of granular, eosinophilic cytoplasm and are pervaded with zymogen granules stainable with periodic acid Schiff's (PAS) stain with diastase resistance. Tumour cell nuclei are uniform and characteristically enunciate a singular, prominent nucleolus. Circumscribing stroma is scanty and fibrous. Foci of perineural invasion and vascular invasion are frequently observed.[5,6] Tumefaction is endowed with variable architecture and exhibits configuration of cystic, acinar, glandular or intra-ductal growth patterns.

Acinar cell carcinoma pancreas may represent a non-neuroendocrine component composed of an admixture of neuroendocrine and non-neuroendocrine neoplasms (MiNEN), a tumefaction which may be appropriately discerned with morphological assessment and precise immunohistochemistry.[5,6]

Pancreatic acinar cell carcinoma appears immune reactive to keratins as CK7, CK8,CK18, CK19, BCL10 or trypsin. Besides, immune reactivity to nuclear beta catenin or CD200 may ensue. Pancreatic acinar cell carcinoma appears immune non reactive to chromogranin or synaptophysin.[7,8] Pancreatic acinar cell carcinoma requires segregation from tumours such as Acinar cell carcinoma exhibiting glandular configurations neuroendocrine neoplasms, pancreatoblastoma or intra-ductal tubulopapillary neoplasm (Figure 1).[7,8]

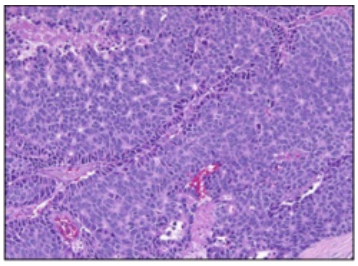

Figure 1 Acinar cell carcinoma exhibiting glandular configurations lined by acinar cells pervaded with moderate, granular cytoplasm, bland nuclei with singular, prominent nucleolus and circumscribing minimal, fibrotic stroma.[7,8]

Computerized tomography (CT) and magnetic resonance imaging (MRI) emerge as a preferred imaging modality for ascertaining pancreatic acinar cell carcinoma. Upon computerized tomography (CT) and magnetic resonance imaging (MRI), pancreatic acinar cell carcinoma delineates an enlarged tumefaction with a well-defined tumour margin, an exophytic pattern of tumour evolution and heterogeneous image enhancement.

Conclusion

Exceptionally, elevated levels of serum alpha feto protein (AFP) can be encountered, especially with incrimination of young subjects. Metastatic disease is accompanied by elevated levels of serum lipase. The neoplasm can be appropriately discerned with cogent tissue sampling or surgical resection.[9,10] Acinar cell carcinoma pancreas can possibly be subjected to surgical resection. Chemotherapy with gemcitabine or radiofrequency ablation appear as optimal alternative modes of therapy. Besides, molecular target therapy may be adopted as an option.[9,10]

Prognostic outcomes are inferior. Cogent Tumour, Node, Metastasis (TNM) staging of the neoplasm emerges as a singular significant prognostic indicator. Acinar cell carcinoma pancreas exhibits an average survival of ~19 months. Tumefaction with an intra-ductal component is accompanied with superior prognosis, in contrast to conventional acinar cell carcinoma pancreas (Figure 2).[9,10]

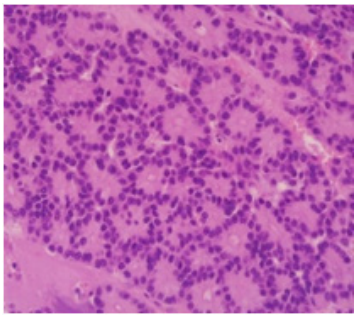

Figure 2 Acinar cell carcinoma delineating glandular articulations layered with acinar epithelial cells imbued with moderate, granular cytoplasm, uniform nuclei with prominent nucleolus and surrounding scanty, fibrotic stroma.[11,12]

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Conflicts of interest

The author declares that there is no conflict of interest.

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11. Image 1 Courtesy: Wikimedia commons.

12. Image 2 Courtesy: Science direct.