Cystic variant of acinar cell carcinoma of the pancreas presenting as pseudo-pancreatic cyst

Sir,

Ductal adenocarcinomas of the pancreas are the most common exocrine pancreatic tumors. Acinar cell carcinoma (ACC) that arise from the pancreatic acinar epithelium are unusual, account for 1% of all exocrine pancreatic neoplasms.[1] A rare variant of ACC is acinar cell cystadenocarcinoma where the neoplasm shows varying degree of cystic changes and consists of innumerable variably sized cysts.[1]

A 65-year-old male presented with complaints of difficulty in voiding urine and burning micturition of two months duration. Systemic examination showed an intra-abdominal mass occupying the epigastric and umbilical region measuring 20x15cms in size. The mass was firm and movable side to side. A provisional impression of a mesenteric mass was made. An abdominal ultrasonography (USG) showed the mass had solid and cystic areas. A CT scan with contrast showed this mass to be retroperitoneal in location. A Fine Needle Aspiration Cytology (FNAC) was performed on the mass. An epithelial neoplasm was suggested on cytology. On laparotomy, a cystic mass was found occupying the whole of the abdomen and apparently arising from the body of the pancreas and located in the lesser sac. Presuming this mass to be a pseudo pancreatic cyst by the surgeon, the cyst was decompressed and the aspirated hemorrhagic fluid from the cyst sent for amylase estimation, which was within normal limits. A part of the mass and cyst which could be excised were sent for histopathological examination. The postoperative period was uneventful.

Grossly, the resected specimen consisted of multiple soft tissue masses. The largest one measured 6x4.5x1.5cms and appeared cystic with multiple cysts ranging in diameter from 1mm to 5mm. The external surface was also hemorrhagic. Cut section through the solid areas at the periphery showed a spongy and hemorrhagic appearance. The other mass, which appeared to be pancreatic tissue, measured 5x3.5x1cms and the cut section showed solid gray white areas [Figure 1].

Microscopically, the sections through the solid and cystic areas of the tumor showed an epithelial neoplasm with a lobulated pattern of growth. Cells within the lobules showed a trabecular and glandular pattern of arrangement consisting of round to oval cells with an eosinophilic to granular cytoplasm and uniform vesicular nuclei with occasional nucleoli [Figure 2]. The cystic spaces were lined by similar cells [Figure 3]. Nuclear pleomorphism and mitotic activity was minimal. A periodic acid-Schiff stain did not show evidence of mucin. An impression of acinar cystadenocarcinoma of pancreas was made at morphology.

On immunohistochemistry, the tumor cells were negative for cytokeratin, EMA, synaptophysin, chromogranin, CEA and AFP. An endocrine and ductal differentiation was ruled out confirming the diagnosis of acinar cystadenocarcinoma. The patient was referred to an oncology center.

As per review of available literature, only five cases of acinar cystadenocarcinomas of pancreas have been reported since its first description in 1981.[1-3] The present case adds to the

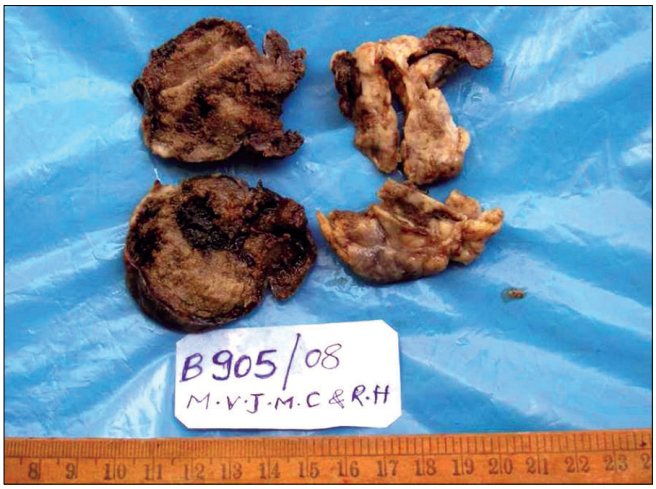

Figure 1: Gross photograph of the cut section of tumor, through solid areas, showing a spongy and hemorrhagic appearance

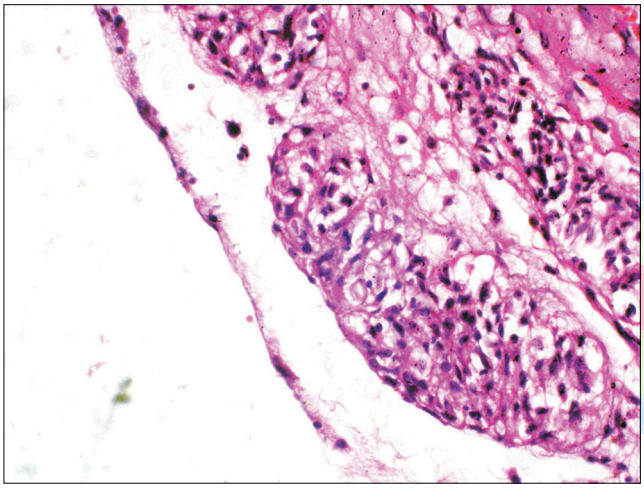

Figure 3: Photomicrograph showing cells with eosinophilic granular cytoplasm lining a cystic space (H and E, x400)

list and is the first one reported at our institution. In this case, the tumor presented as an abdominal mass and arose from the body of the pancreas as in the three reported cases of acinar cell cystadenocarcinomas of the pancreas.[1-3] Grossly, the ACCs are usually large, solid, well circumscribed with necrosis and hemorrhage.[4] On the other hand, the rare cystic variant termed as acinar cell cystadenocarcinoma consists of innumerable variably sized cysts with a spongy appearance on the cut surface.[1,3,4] The present case showed a partially cystic mass with a spongy appearance and hemorrhagic areas.

The most common histologic patterns seen in ACC are acinar and solid. The acinar pattern consists of small lumina surrounded by cells with eosinophilic granular cytoplasm and basal nuclei and in solid areas the tumor cells are in sheets and nests with central nuclei and little cytoplasm. The nuclei of the tumor cells are vesicular, show moderate pleomorphism with centrally placed round nucleoli. Mitotic activity is variable. Vascular and

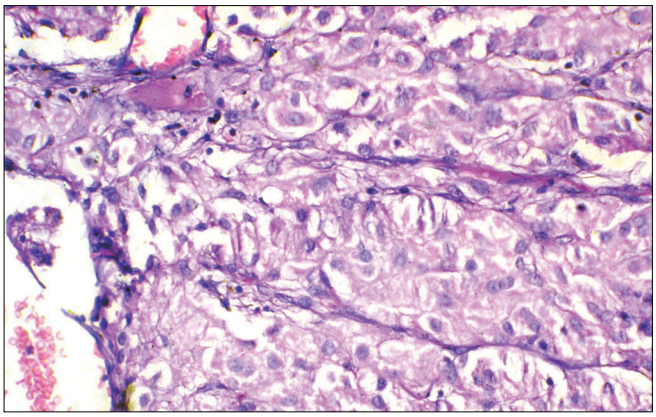

Figure 2: Photomicrograph showing an acinar pattern of cells with granular eosinophilic cytoplasm and vesicular nuclei with nucleoli (H and E, x400)

perineural invasion is common.[4] Microscopically, in the cystic variant, the cysts are lined by a single layer of cuboidal cells which have characteristics of acinar cells. Immunohistochemically, the cells express α_1-antitrypsin, trypsin and lipase in their cytoplasm, thus confirming the acinar origin of the tumor.[1] The present case showed varied architectural pattern as well as cystic change. The cells in the tumor had characteristics of acinar cells. The tumor showed no evidence of an endocrine differentiation (chromogranin –ve) and showed negativity for EMA and CK as opposed to a ductal carcinoma. Trypsin and chymotrypsin were not performed, however, cystic change was seen grossly with a spongy appearance. It was supported by the histologic characteristics of acinar cells with a granular cytoplasm and the typical growth pattern favored a diagnosis.

Other cystic lesions of pancreas such as serous microcystic adenoma and benign acinar cell cystadenoma of the pancreas were ruled out. Serous microcystic adenomas show a central scar grossly and microscopically composed of cystic spaces lined by clear cells with bland nuclear morphology. Benign acinar cell cystadenoma of the pancreas is cytologically bland with rare mitotic figures, where as acinar cell cystadenocarcinomas consists of moderately atypical cells with prominent nucleoli, frequent mitosis and show infiltration.

Acinar cyst adenocarcinoma of pancreas is not prognostically different from the classic type.[1] Follow-up data, available for four out of five reported cases of acinar cyst adenocacinomas, showed that all the affected had metastasis at presentation or a few months later and two died of disease at 13 and 37 months after diagnosis.[1,2]

This case is reported for its rarity and to highlight its need to be diagnosed when presenting as a predominantly cystic mass where a clinical impression of a pseudo pancreatic cyst is possible.

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