Pancreatoblastoma

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Pancreatoblastoma is a rare pancreatic neoplasm involving predominantly the younger age group. It has a distinct histological appearance with elements of acinar and ductal differentiation. This case is presented because of its rare incidence and good prognosis on early diagnosis and surgical management. To the best of our knowledge no similar case has been reported in the Indian literature.

Case Report

A 5-year-old boy was admitted to Kalawati Saran Children's Hospital with the complaints of vague abdominal pain and a gradually increasing lump in the left hypochondrium for one and half year prior to admission. There was no history of fever, weight loss or other constitutional symptoms.

On examination, an intra abdominal mass was felt in the left hypochondrial region measuring 7 x 6 cm. It was moving with respiration, was soft to firm in consistency and nontender.

Sonography revealed a 6.5 x 5.4 cm soft tissue mass predominantly cystic with some solid areas adjacent to the tail of pancreas, separate from spleen anterior to left kidney *(Fig. 1)*. The head and body of pancreas were normal.

Plain X-rays of abdomen were normal. Barium study revealed displacement of lesser curvature of stomach, fourth part of duodenum and duodenojejunal junction to the right.

Pre-operative fine needle aspiration cytology showed clusters of epithelial cells, cyst macrophages, neutrophils and

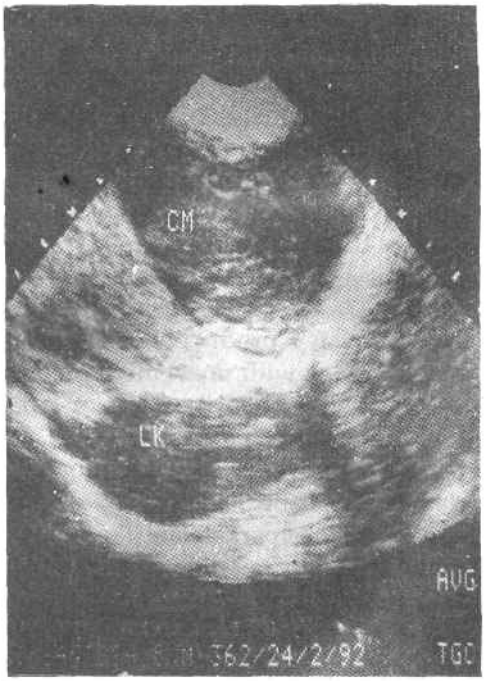

Fig. 1. Sonogram of left hypochondrium showing cystic mass (CM) with solid areas anterior to left kidney (LK).

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lymphocytes in a hemorrhagic background. No definite opinion was possible.

Peroperatively a 7 x 8 cm lobulated tumor mass was seen in relation to tail of pancreas and extending into mesocolon. A successful extirpation of the tumor mass was performed.

Grossly, the tumor was well encapsulated, 7 x 8 cm with nodular external surface and friable granular soft grey white cut surface. Microscopically encapsulated tumour tissue showed two types of cells, small round cells with ill defined cell outlines present in sheets *(Fig. 2a)* or acinar fashion *(Fig. 2b)*. These cells had PAS positive diastase sensitive zymogen granules. Large polygonal cells with moderate amount of pink cytoplasm *(Fig, 2c)* were also seen. Squamad differentiation was also observed *(Fig. 2d)*. A diagnosis of pancreatoblastoma was given.

Post operative period was uneventful. One month postoperatively sonography revealed no evidence of residual mass or metastasis. The patient is doing well and there is no evidence of recurrence or metastasis after one year.

Discussion

Pancreatic carcinomas are classified on basis of cell of origin into: *(a)* ductal cell, *(b)* islet cell, *(c)* acinar cell, and *(d)* uncertain histogenesis. The most common tumor in adults and children is ductal cell carcinoma. Except for islet cell tumor, the three year survival of pancreatic tumor is 2%. Notable exception to this prognostic statistics is a tumor of uncertain histogenesis which was variously reported as pancreatoblastoma or infantile type pancreatic-carcinoma(l). Embryologically, pan-

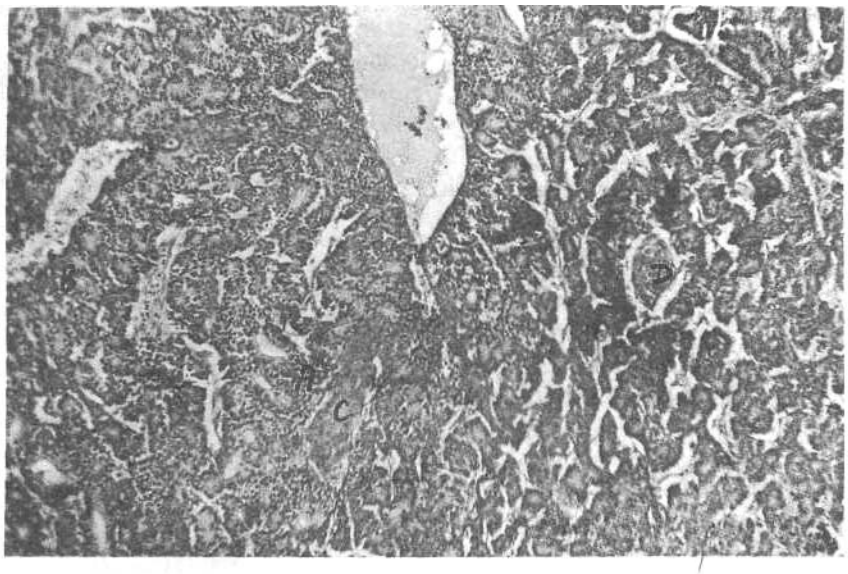

Fig. 2. Microsection showing small round cells (a); acinar arrangement of cells (b); larger cells with vesicular nuclei (c); squamoid differentiation (d). (H&E ×100).

creatoblastoma arise from the ventral pancreas isolated by lack of communication with duct of Wirsung(2).

In 1964, 16 cases of childhood pancreatic carcinoma were reviewed(3). In 1973, 12 cases were reviewed from the Japanese literature(4). The first case of infantile pancreatic carcinoma was reported in 1971(5). To the best of authors knowledge, no case has been reported in the Indian literature.

Pancreatoblastoma is a rare tumor comprising of 0.5% of epithelial neoplasm of pancreas(6). This tumor has peripheral lobulation and central necrosis, arises from the body or tail of pancreas. CT scan shows the mass as hypodense with hypodense areas in liver and porta suggestive of metastasis. Magnetic resonance imaging may show low signal mass on Tl image and bright signal on T2 image(7). The mass is relatively avasulcar and has good prognosis if metastasis is not present at the time of presentation. The treatment is surgical. Chemotherapy and radiotherapy are required in case of metastasis.

Our case, a 5 year old presented with an asymptomatic encapsulated cystic mass with solid areas arising form the tail of pancreas. Early diagnosis and surgical management led to complete cure with no recurrence or metastasis seen after one year follow up.

The prognosis is good since only two out of eight cases of pancreatoblastoma died, one of which was due to postoperative complications and other due to pulmonary metastasis in follow up varying from 15 months to 13 years(l). The terminology is objectional to some because histolcrgically this tumor does not have a typical blastemal appearance and also because of good prognosis.

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