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Case Report

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PANCREATOBLASTOMA IN A 9 YEAR OLD FEMALE CHILD

Dr. Kamal Nain Rattan[1], Dr. Omkar Manik Baridabad*[2], Dr. Priyadarashani[2], Dr. Rishabh Batra[2], Dr. Kavitha[2]

[1]Department of Pediatric Surgery, Pt. B. D. Sharma PGIMS, Rohtak, India.
[2]Pediatrics, Pt. B. D. Sharma PGIMS, Rohtak, India.

***Corresponding Author: Dr. Omkar Manik Baridabad**
Pediatrics, Pt. B. D. Sharma PGIMS, Rohtak, India.

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ABSTRACT
Pancreatoblastoma is a rare tumor of childhood and it usually presents with a abdominal mass. We report a case of Pancreatoblastoma in a 9-year-old female child who presented with a huge left sided abdominal mass, the case was successfully managed by surgical excision of mass and pre and post-operative chemotherapy.

KEYWORDS: Pancreatoblastoma, Congenital, Infantile.

INTRODUCTION

Pancreatoblastoma is a uncommon tumor occurring in childhood. Most commonly seen in infants and young children, First described by Frantz in 1959. Later in 1971 it was called as infantile carcinoma of pancreas by Frable et al based-on histology and ultrastructural finding. Then the term Pancreatoblastoma was given by Horie et al based-on histology features in the year 1977. Till 1977 only over 50 cases have been reported in literature.[1] Based on histological appearance, it is considered to be embryonic in origin some studies show endocrine component alfa fetoprotein in acinar cell, suggestive that tumor arises from multipotent stem cells. Tumor is most common in male, median age is 5y. The role of adjuvant chemotherapy, radiotherapy still under discussion as very few cases of pancreatoblastoma have been reported in literature. The prognosis depends upon resectability of the tumor, if it's completely resectable then has good prognosis. The prognosis is poor in unresectable large tumor and in metastatic pancreatoblastoma.

CASE REPORT

A 9-year-old female child presented with history of left sided abdomen mass gradually increasing in size since last 1 year. The mass was noticed accidently by parents. No history weight loss, anorexia, vomiting, hematuria. On examination, the child was well preserved no peripheral lymphadenopathy. On per abdomen examination which revealed huge left sided mass with smooth surface crossing midline, a small nodule was palpable on upper part of tumor. There was no hepatomegaly, no ascites and routine investigation like complete blood count, RFT, LFT with in normal limits but alfa fetoprotein was raised. Ultrasound revealed a huge heterogenous mass of mixed echogenicity occupying left side of the abdomen which was crossing midline. Right kidney was normal but left kidney was displaced inferiorly and laterally by tumor, spleen could not be identified, so it was misdiagnosed as massive splenomegaly and second possibility was of Wilms tumor. CECT large heterogenous enhancing soft tissue attenuation lesion is 16×14×11cm in left hypochondrium with few necrotic areas and foci of peripheral calcification, left kidney is separately visualized. The mass is reaching up to anterior abdominal wall and displacing left kidney posteriorly. [Fig: 1].

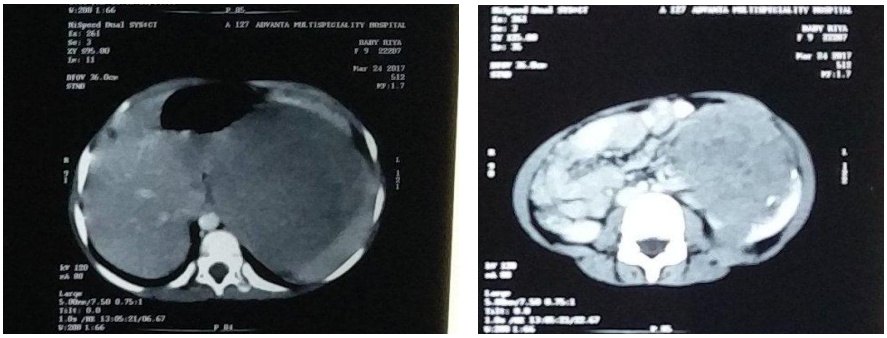

Fig 1: Enhanced axial CT image showing a large well defined encapsulated tumor in body and tail of pancreas.

FNAC specimen consisting of numerous oval-to-cuboidal cells that had a moderate amount of granular cytoplasm. Spindle-shaped, elongated and triangular-shaped epithelial cells were also seen, along with smaller cells that had a higher nuclear/cytoplasmic ratio and a denser cytoplasm. In addition, there were abundant fragments of stroma present, including some surrounded by epithelial cells Cytological features are suggestive of malignant neoplasm favoring pancreatoblastoma.

Due to large size of tumor adjuvant chemotherapy was started with cisplatin and Adriamycin following chemotherapy for one month tumor mass regressed. Patient was taken for surgery. Explorative laparotomy done which reveled a mass of 15x20x25 cm arising from body and tail of pancreas well encapsulated, it was not infilterating the surrounding structure. Tumor mass was excised completely along with body and tail of pancreas during surgery, there was injury to the spleen, hence splenectomy was done, excised tumor mass was size of 12×10×9cm. [Fig: 2].

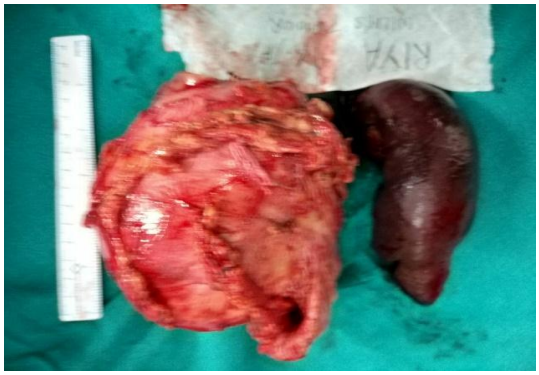

Fig 2: gross specimen showing spleen and pancreas.

Histopathological examination of tumor mass measuring 12×10×9cm. cut section is gray white encapsulated and a necrotic cavity measuring 6×5cm is identified. Microsection examined from tumor mass shows histological features compatible with pancreatoblastoma with areas of hemorrhage and necrosis. Microsection examined from spleen are unremarkable, which confirmed the diagnosis of pancreatoblastoma. [Fig: 3].

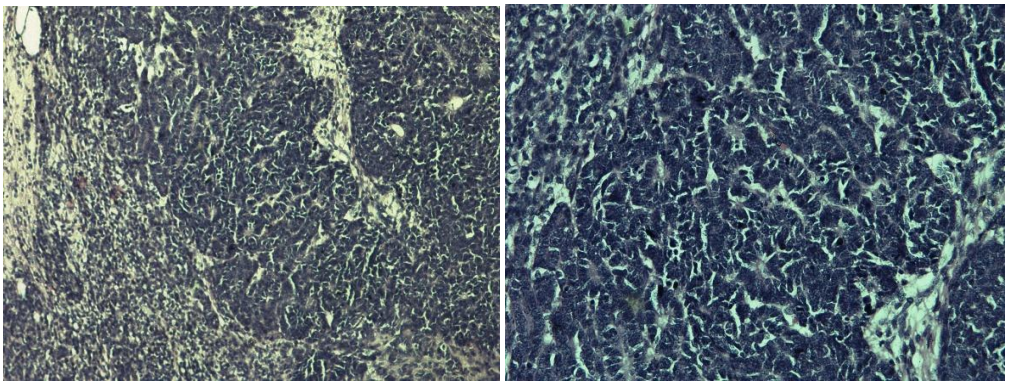

Fig-3a **Fig-3b**

Photomicrograph showing lobules of neoplastic cell separated by fibrous bands reviling. Acinar and ductular differentiation (A:100X , B:200X).

Patient is on post operative chemotherapy Adriamycin and cisplatin. Alfa fetoprotein done in post operative period was decreased. Patient is followed up with USG and afla fetoprotein. In post operative period the child is doing well.

DISCUSSION

Pancreatic tumors are rare in children and pancreatoblastoma comprises only 0.5-1% percent of pancreatic non-endocrine tumors occurring in children.[2] Between 1885 and 1991 only 71 cases of pancreatic tumors in childhood and adolescence have been reported in literature. By immunohistochemistry, the tumors exhibited acinar, endocrine, and ductal differentiation, with positivity for pancreatic enzymes (100%), endocrine markers (82%), and carcinoembryonic antigen (85%).[3] Recently genetic alterations also have been characterized and the commonest change is allelic loss of 11p. Elevated serum alpha-fetoprotein levels have been reported in up 68% of cases.[4] The pathologic features of pancreatoblastoma have been explained well. Microscopic finding shows that it composed of epithelial cells and variable amounts of stroma. Epithelial cells are typically arranged in a glandular or acinar pattern resembling acini and short tubules, which contain zymogen granules. The histogenesis of this tumor is still uncertain. It is hamartomatous or dysembryogenic development of ductal cells of ventral portion of primordial pancreas. Pancreatoblastoma contains pluripotent cells capable of differentiating along the pathway of all three pancreatic cell types. Molecular investigation has disclosed a mosaic paternal 11p15 uniparental disomy in the tumor cells of pancreatoblastoma.[4-7] Pancreatic tumors are classified on the basis of cell of origin, four major categories have been described by Buchino et al: ductal cell, acinar cell, islet cell, and those of uncertain histogenesis.[8] The tumor shows three different types of pattern like acinar, endocrine and ductal differentiation,[9] on ultrastructural examination of the epithelial cells which shows electron dense zymogen granules or small dense neuroendocrine granules.[10] This tumor is more common in Asians than

in the caucasian. Many of the tumors arise from body and tail,[11] however because of the rare site and its unusual presentation with jaundice which could be seen occasionally in the patients with the mass in the region of pancreatic head. The level comes down once the tumor is resected. The tumor has to be differentiated from Wilms, malignant lymphoma, rhabdomyosarcoma, neuroblastoma, hepatoblastoma, Tumors measuring up to 25x20x15 cm and weighing up to 2.5 kg have been reported. The diagnosis is made with help of USG, CT SCAN and MRI. Primary treatment is surgical excision and the tumor is well encapsulated. Encapsulated tumors have a better prognosis. Chemotherapy shows marginal benefit in two patient reported by Kataria et al.[12] The primary therapy is surgery in all cases. Nagaraj et al reviewed the results of 20 resected tumors in children with survival rate of 80%. The most important prognostic factor in pancreatoblastoma is complete removal of tumor. Graffin et al reported a case of locally recurrent pancreatoblastoma successfully treated with radiotherapy.[13] In chun's series of 5 patients, all of the 3 patients who had unresectable tumor died despite of adjuvant chemotherapy.[14]

The tumor has an indolent course and is amenable for various modes of treatment but surgery is the most optimal treatment. Complete resection of the tumor offers the best prognosis. However, in the presence of metastatic disease, it is of limited value. In these situations where there is suspected or documented metastatic lesions, empirical chemotherapy regimens that include cisplatin and Adriamycin have been used. When the tumor is unresectable and the patient is non-responsive to chemotherapy, radiotherapy is given. Significant shrinkage of the tumor has been reported after treatment with.

CONCLUSION

Pancreatoblastoma rare tumor of childhood with non specific presentation with good prognosis when resected completely. Prognosis is poor if metastasis present.

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