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ALPHA-FETOPROTEIN SECRETING PANCREATOBLASTOMA IN CHILDHOOD

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SUMMARY: A case of pancreatoblastoma, a rare epithelial pancreatic tumor of childhood is presented. A six year old boy, admitted to the hospital with fatigue, anorexia, headache and an abdominal mass was diagnosed to the pancreatoblastoma. Moreover, serum alpha-feto protein (AFP) levels which were very high and decreased after the excision of the tumor, were evidence of the secretory nature of the tumor. AFP was also shown on immunoperoxidase staining. Postoperatively, the patient has been well for 24 months.

Pancreatoblastoma in childhood is reevaluated on the scope of recent literature.

Key Words: Pancreatoblastoma, Alpha-feto protein (AFP).

Primary epithelial tumors of the pancreas are quite rare in childhood (2,4,5,6). Pancreatoblastoma, which is the exocrine tumor of pancreas presents itself with a big and fast-growing mass in the abdomen (2). Although other tumors of the pancreas have been recognised for their hormonal secretions, it was usually believed that pancreatoblastomas had no endocrine activity (4,6). However, in the recent years there are some reports that the tumor secretes alpha-fetoprotein, adrenocorticotrophic hormone, gastrin and glucagon (5,6). A case of pancreatoblastoma with elevated levels of APF is presented.

CASE REPORT

A six year old white boy was admitted to the Pediatric Hematology-Oncology department of the İstanbul Medical School on Augst, 27, 1987 for the evaluation of an abdominal mass together with complaints of fatigue, anorexia and headache for the last 15 days. He had used various analgesics and vitamins in the meantime and was referred to our hospital when his abdominal mass began to enlarge rapidly. His family history and previous medical report was non-contributory. Physical examination revealed a slender, pale boy with a 15x12 cm. firm, mobile abdominal mass which extended from the left upper abdomen, across the midline an epigastrium to the lower quadrants. The area of Traube was dull on percussion. The liver was of normal size. No lymphadenopathy was palpable. Arterial blood pressure was between the normal limits and the examination of the remaining systems did not reveal any significant pathology.

Laboratory findings included a serum APF level of 200 mIU/ml (N=10 mIU/ml), serum amylase level of 16 IU/L (N=8-32 IU/L), an alkaline phosphatase level of 119 IU/L (N=38-138 IU/L). Serum proteins, aspartate aminotransferase and alanine aminotransferase and carcinoembryogenic antigen (CEA) were all within normal limits. Roentgenograms of the lung were normal. The direct roentgenogram of the abdomen revealed speckled calcification at the left-side at L.2-L.3 vertebra level. At the ultrasonography of the abdomen, a solid, heterogenous mass between the hila of the liver and spleen and urinary bladder was diagnosed. Intravenous pyelography showed normal kidneys and ureters. On August 31, 1987 the tumor was completely removed with the rescetion of the tail of the pancreas and splenectomy. Postoperatively, serum AFP level returned to normal limits.

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The patient was started on a regimen of 5-fluorouracil, doxorubicin and mitomycin-C and was continued 18 months. postoperative course was uncomplicated and 24 months of follow up disclosed no evidence of recurrent tumor.

PATHOLOGICAL FINDINGS

Gross pathology (fig.1): At operation, an 14x13x8 cm, xantochromic colored coarsely lobulated, smooth surfaced tumor which has infiltrated the meso of the colon, the tail of the pancreas and the artery and vein of the spleen was totally removed with excision of the tail of the pancreas and splenectomy. The cut surface was pink-yellow colored, partially multicystic and necrotic. Light microscopy (fig.2,3): The tumor cells which had hyperchromatic nuclei and narrow cytoplasms were lined in a rosette-like fashion around the PAS (+) material, among these groups, fusiform cells with oval nuclei formed organoid structures and fibrotic connective tissue was discernible in between the cells. The pancreatic tissue was edamatous with fibrotic and inflamatory changes and atrophic asini were seen among the increased connective tissue. Immunoperoxidase staining (fig.4): The tumor tissue displayed fine granuler deposition of AFP whereas other immunoperoxidase staining including CEA was negative. The histological examination of the spleen and the lymph nodes were normal.

DISCUSSION

Epithelial tumors of the pancreas are rare in children (2,4,5,6). These tumors show a diphasic distribution; some of them are seen in early children as are the other blastomas, mainly in boys and are malignant, whereas the others are seen in late childhood, mainly in girls and are more benign. Pancreatoblastoma has a favorable outcome and therefore requires differentiation from more agresive neoplasms (3,6).

Pancreatic tumors can be divided into four groups as exocrine, ductal, mesenchymal and endocrine. Pancreatbolastoma is the exocrine tumor of the pancreas and can be located at any region of the organ (2.6). Herie et al. suggested that the tumor could be located at the body of the pancreas and can have a relationship with the islet cells (3). In our case the tumor was located at the body of the pancreas and had grown towards the hilus of the spleen. Hence tail of the pancreas and the spleen was removed at the operation. pancreatoblastomas are usually benign but the prognosis for those located at the distal part is slightly

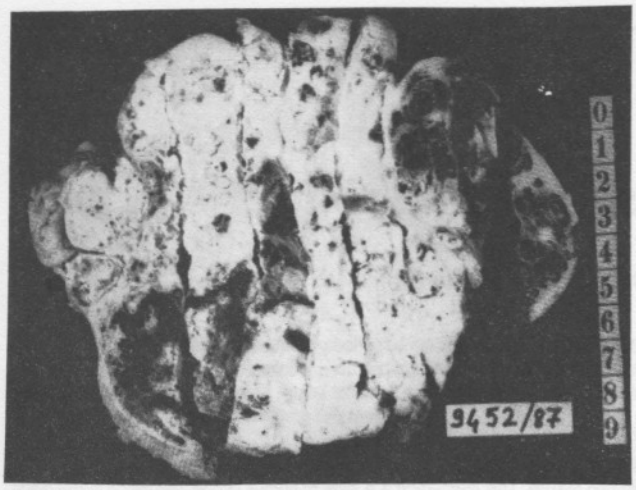

Fig.1: Gross pathology of pancreatoblastoma.

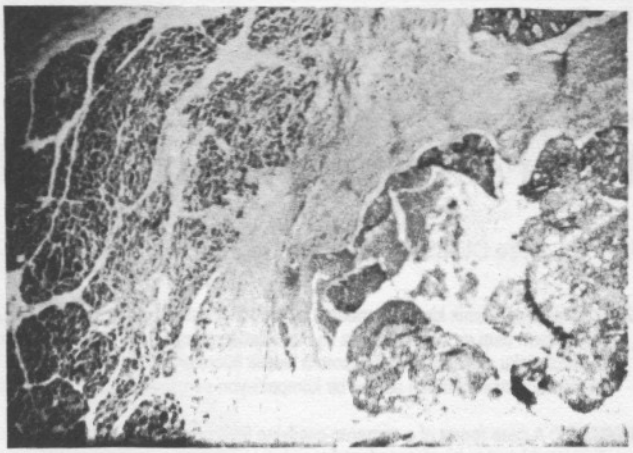

Fig.3: Light-microscopy view of the tumor.

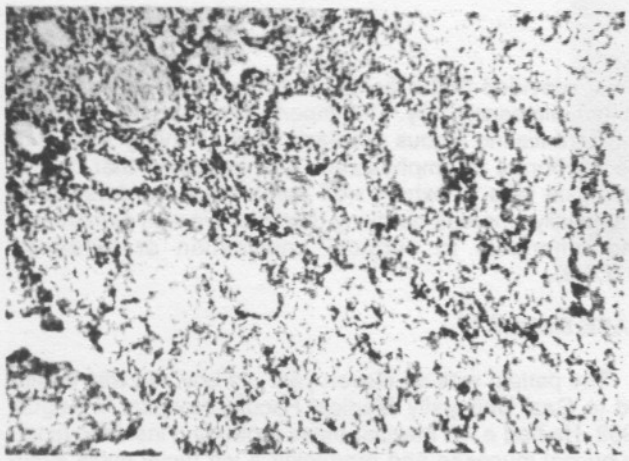

Fig.2: Light-microscopy view of the tumor.

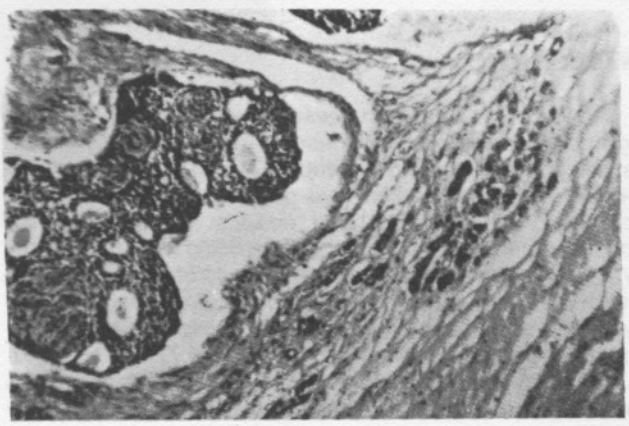

Fig.4: Granular deposition of AFP in the tumor tissue

less favorable (2,5,6).

The islet cell tumors of the pancreas secrete many hormones but the exocrine tumors are known to be nonfunctional. In recent years, insulin, gastrin, somatostatin and ACTH have been shown by immunohistochemical assays in pancreatoblastoma cases. Elevated serum AFP levels are extremely rare and only very few cases have been reported (1,5). The findings of elavated serum AFP levels before surgery, normalisation after surgery and the demonstration of AFP in tumor cells by immunoperoxidase technique clearly substatiates the production of this oncofetal protein by the tumor. The presence of AFP in the upper gastrointestinal system and pancreas is no surprise because it is the usual production site of this protein in the fetus. The presene of AFP within the tumor cells further substantiates the embryonel nature of this tumor and in this regard shows analogy to the histologically different, but embryologically related hepatoblastoma.

The relationship of AFP and prognosis is not well-established. According to Buchino et al., AFP secreting pancreatoblastomas have good prognosis (1), but prognosis may be totally dependent upon histology. In fact, the prognosis for pancreatic tumors of childhood is not well known and non-functional tumors have been reported to have a better prognosis Our cases has been followed up for 24 months and has not shown any signs or symptoms of recurrent tumor.

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