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CASE REPORT

ADULT PANCREATOBLASTOMA: A RARE ENTITY

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ABSTRACT: Pancreatoblastoma is a rare tumor of neuroendocrine origin affecting paediatric age group. It is an extremely rare neoplasm of adults. Patients with pancreatoblastoma present with complaints of abdominal pain and distension. Clinically diagnosis is usually of a retroperitoneal sarcoma or tumor of colon. This tumor is generally detected accidentally during laparotomy and diagnosis is confirmed by histopathology. The presence of squamoid corpuscles with a morular appearance and acinar differentiation is the most characteristic feature of this tumor. In this article we present one such rare case of pancreatoblastoma in a 50yr old lady.

KEYWORDS: Pancreotoblastoma; Adult; Pancreatic cancer; Histopathology.

INTRODUCTION: The term pancreatoblastoma was introduced in 1977 by Horie et al. to describe a rare pancreatic tumor of childhood, previously known as infantile carcinoma of the pancreas. The histological features are similar to the pancreatic tissue of fetus at approximately at 7th week of gestation.[1-2] A pancreatoblastoma typically manifests in children between 1-8yrs of age.[3] It may also affect neonates and elderly.[4-7]

Nevertheless, it is extremely rare in adults. In 1986, Palosaari et al described the first case in an adult. Men are more commonly affected than women. It is clinically occult and is large at the time of presentation. Clinical features are typically due to mass effect like abdominal pain, weight loss, satiety, vomiting, jaundice and constipation.[8-14] Here we describe a case where the preoperative diagnosis was retroperitoneal sarcoma but was later confirmed to be pancreatoblastoma on histopathological examination.

CASE REPORT:A 50yr old female presented with pain abdomen, mild distention of abdomen and constipation, on and off for the past two months. She also had loss of weight and loss of appetite. On examination she was thin built, afebrile, pale and there was mild distention of abdomen. On examination a palpable abdominal mass was present in the left hypochondrium which was hard in consistency, moving with respiration, had irregular borders, non-tender and ballotable but not bimanually palpable.

Clinically differential diagnosis of retroperitoneal sarcoma or carcinoma of transverse and descending colon was made. Laboratory investigations were within normal limits except for haemoglobin of 7gms/dl. Ultrasound abdomen revealed 8.5x6.5x8.6 cm sized, ovulated heterogeneous mass lesion seen in the left lumbar region, inferior to the left kidney. The lesion appeared to be medial to the descending colon, with no demonstrable fat plane in between. The tail of pancreas was not visualized separately. The lesion was vascularized and spleen appeared encased. Few para aortic and celiac lymph nodes were seen. Diagnosis was confirmed by CT.

On laparotomy a necrotic nodular mass was found to be arising from the tail of the pancreas, encasing the splenic vein and infiltrating the root of mesentry and transverse mesocolon (Fig 1).

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Large Omental and mensenteric deposits were found (Fig. 2). Debulking of the tumor along with distal pancreatectomy, splenectomy and resection of transverse colon was done. Histopathology of the resected specimen confirmed a malignant epithelial tumour.

The microscopic features demonstrated squamoid corpuscles and acinar differentiations which are classical features of pancreatoblastoma (Fig. 3). Immunocytochemistry was used for identifying acinar differentiation.Omentum showed multiple tumor deposits. The patient was referred to regional cancer institute for chemotherapy and was on follow up till she died 2 years after diagnosis.

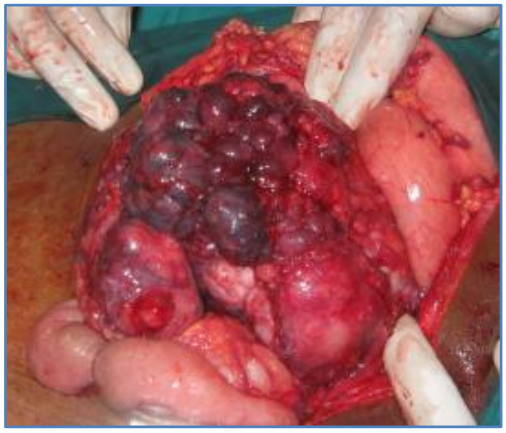

Fig. 1: Pancreatic tumour

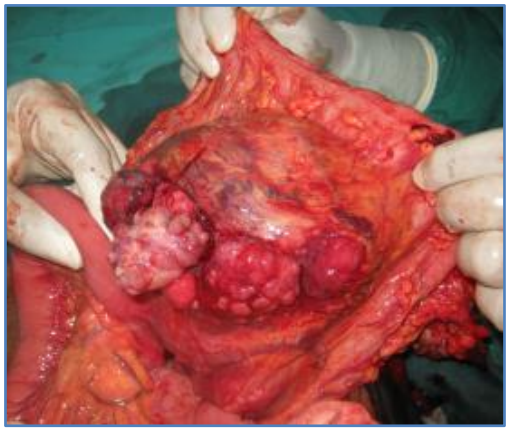

Fig. 2: Mesenteric &Omental deposits

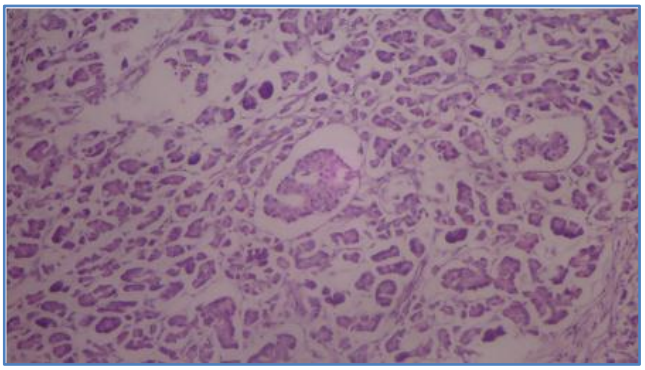

Fig. 3: Histopathology

DISCUSSION: Pancreatoblastoma accounts for 0.5% of exocrine tumors of pancreas, with highest incidence in the first decade of life with a predilection for males and Asians. Differential diagnosis includes nonfunctional pancreatic endocrine tumor, acinar cell carcinoma, solid pseudopapillary tumor and adenocarcinoma[15]. Usually adult pancreatoblastomas are slow growing and large, ranging between 1.5cm and 20cm with majority larger than 8cm.[6] Approximately half of them are located in the pancreatic head.[6-9&12]

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Few cases have been reported to arise from tail.[6, 10&13] Du et al have reported a case located at ampula of Vater and Zhu et al have reported a case in the body of pancreas. Distant metastasis commonly involves liver (26%) and then lymph nodes (14%). CT scan shows a well circumscribed, heterogenous mass with features suggestive of peripheral vascularity and viability with areas of central necrosis.[15]

Histologically the presence of squamoid corpuscles is the most characteristic feature besides acinar differentiation. Treatment includes surgical resection, followed by adjuvant chemotherapy or radiotherapy because of high metastatic potential of the tumor. But prognosis is poor and more than half of them die within 3yrs of initial diagnosis.[6]

An atypical case has been described by Balasundaram et al where the tumor was small (3.6cm) with extensive metastasis to liver, lungs and breast. Besides this histologically there was absence of acinar component. Our patient was a female, with the tumor involving the root of pancreas, spleen and transverse colon. Diagnosis was possible only by histopathological picture ofsquamoid corpuscles and acinar differentiations.She had no distant metastasis at the time of diagnosis but died 2 years post diagnosis substantiating the high malignant potential of the tumour.

CONCLUSION: Adult pancreatoblastoma is usually a diagnostic challenge and also can have an atypical clinical picture and a small primary with extensive metastasis to unusual sites as well as without metastasis as in our case. Given its rarity, a high index of suspicion is required to correctly diagnose this condition and also well prepared while exploring a retroperitoneal mass in the epigastric region.

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