ORIGINAL RESEARCH PAPER

Paediatric Surgery

PANCREATOBLASTOMA: A RARE TUMOUR IN CHILDREN (CASE REPORT)

KEY WORDS:
pancreatoblastoma; pancreatico-duodenectomy; whipples operation.

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ABSTRACT

Pancreatoblastoma is a rarely encountered malignant neoplasm in children. It is a slow growing tumour and attains large size before diagnosis is made. Surgical excision may be challenging because of its large size, local infiltration or distant metastasis. Because of its rarity, there is as yet no standard treatment protocol for pancreatoblastoma. We are presenting a 14 year old male child with pancreatoblastoma, who was treated successfully with pancreatico-duodenectomy followed by a course of radiotherapy and six cycles of chemotherapy. The patient has been on follow up and doing well.

INTRODUCTION:

Pancreatoblastoma is a rare malignant tumour in children and is the most common pediatric pancreatic tumour in children less than 5 years of age [1,2]. The tumour originates from the exocrine gland of the pancreas, is slow growing and because of its large size at presentation, the tumour poses challenge at surgical resection [3]. Complete surgical excision is the treatment of choice combined with adjuvant chemotherapy and/or radiotherapy. We report an adolescent boy, who presented with a large mass arising from the head of the pancreas. The tumour was resected successfully, histopathology revealed it as pancreatoblastoma. The patient received post-operative radiotherapy and completed six cycles of chemotherapy. The patient has been asymptomatic on follow up after 11 months.

Case report:

A 14 yrs, male patient presented with dull aching pain abdomen of 5 days duration following blunt trauma. He had several episodes of non-bilious vomiting. On examination, his vitals were stable but he looked pale. He had deep tenderness in epigastric and umbilical areas with an ill defined lump with smooth surface and there was no movement with respiration appreciable. His serum bilirubin was marginally raised (1.3mg%), and serum alpha fetoprotein level was nomal for his age. Contrast (intravenous Iohexol) enhanced computed tomography showed a pancreatic mass arising from the uncinate process (size-6 cm x 5.2 cm x 7.2cm) with heterogenous enhancement and a dilated major pancreatic duct (MPD). The patient was taken up for surgery. On exploration a large, smooth, globular mass noted arising from the head of the pancreas. Superior mesenteric vein was dilated. Few regional lymph nodes adjacent to pancreatic head were enlarged and sampled. Pylorus preserving pancreatico-duodenectomy and Roux-en-Y choledocho-jejunostomy with Gastrojejunostomy with end to side pancreatico-jejunostomy done. The patient had uneventful recovery. Oral feeding was started from day 7 and gradually stepped up and patient was able to take normal oral diet by day-10. The histopathology report was consistent with pancreatoblastoma with few enlarged regional lymph nodes but with no nodal metastasis observed in sampled lymph nodes. The patient received a course of radiotherapy (3000R) and 6 cycles of chemotherapy with Cisplatin, etoposide and cyclophosphamide. After 11 months of surgery, the patient has been on regular follow up and doing well.

DISCUSSION:

Pancreatoblastoma is a rare pediatric malignant neoplasm, accounts for 0.2% of all pancreatic tumours, which mimics abnormal embryological development [2,4]. Secondary involvement of the pancreas by adjacent tumor, especially neuroblastoma, may be difficult to distinguish from a primary pancreatic tumor and is much more common. Congenital and acquired cystic lesions can occur in children and mimic cystic neoplasms. Pancretoblastoma accounted for 16% of all pancreatic primary malignant neoplasm in children, with an average age at presentation of 5.5 years [5]. The tumour originates from exocrine glands of pancreas and because of its slow growth and patient remain relatively asymptomatic till tumour attains a large size by the time the diagnosis is made [1]. Serum alpha fetoprotein levels are usually raised in most of these tumours and is used to monitor the disease as tumour marker [6,7]. In the present case, however, serum AFP level was within normal limit no correlation was observed with histological grading. The tumour is usually encapsulated, solid, heterogenous and characterized by richly cellular, cytologically uniform stroma and organized in nests and islets with a tendency to form acinar structures [8,9]. Squamoid corpuscles with occasional keratinization, a pathognomonic feature of pancreatoblastoma, helps it differentiate from acinar cell carcinoma [10].

Although complete surgical resection is the treatment of choice, because of large size, local infiltration or metastasis, complete surgical resection is not always possible. In such situation, a course of neoadjuvant chemotherapy with Cisplatin and doxorubicin is advisable, although, 5 year survival following chemotherapy is not encouraging [3,6,11]. Radiotherapy may be considered, but its role is limited in young patients [11,12]. Because of its rarity, no standard treatment protocol has yet been available for pancreatoblastoma. Our patient had a large tumour at presentation arising from head of pancreas, but fortunately complete surgical excision of the tumour with a pylorus-preserving pancreatico-duodenectomy was possible. Few enlarged regional nodes were removed, which were negative for metastasis. The patient received a course of radiotherapy following surgery (3000Rads) and chemotherapy comprising of Cisplatin, etoposide and cyclophosphamide were introduced. The Italian TumoriRari in Eta' Pediatrica (the Italian TREP project) has recommended chemotherapy regimen with cisplatin at 80 mg/m^2 as a continuous 24-h intravenous infusion followed by doxorubicin 60 mg/m^2 over 48 h (PLADO) for four cycles for completely resected patients and six cycles for resection with microscopic residual or nodal involvement [14]. The present case has completed six cycles of chemotherapy and has been on follow up since then without any symptom. Literature on management and outcome of pancreatoblastoma is limited, and one study reported that the presence of synchronous or metachronous metastasis, unresectability of the lesion at the time of diagnosis and being over 16 years old worsen the prognosis [13]. On the other hand, multivariate analysis found that complete surgical resection and the development of post-operative metastasis independently influence the long-term survival.

CONCLUSION:

Rare case of pancreatoblastoma in adolescent boy has been presented. The patient responded well to surgical resection followed by a course of radiotherapy and six cycles of chemotherapy. Patient is on followed up.

Images Of The Case:

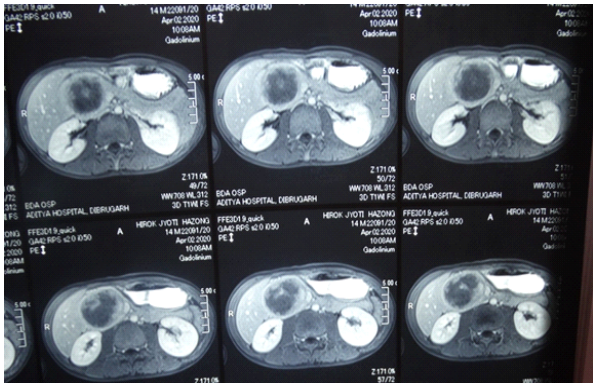

Fig 1: Contrast Ct Shows Large Mass Arising From Head Of The Pancreas Showing Heterogenousenhancement On Post- Contrast Image

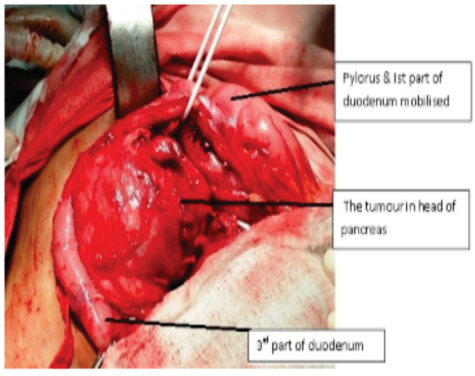

Fig 2: Intraoperative Findings

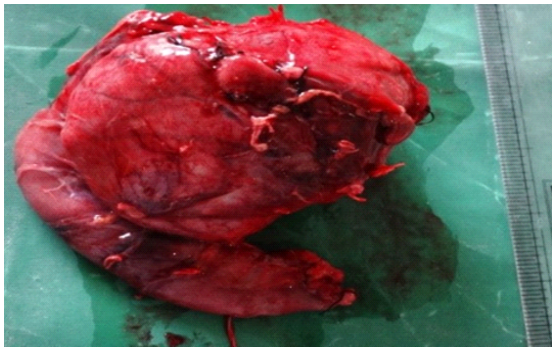

Fig 3: Excised Pancreatic Mass Along With C-loop Of Duodenum

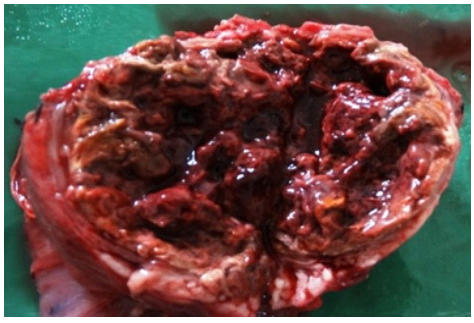

Fig 4: Cut Open Excised Pancreatic Mass Showing Necrotic Debris

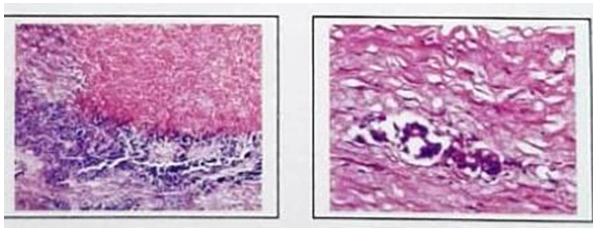

Fig 5: Histopathoogical Examination Showing Extensive Calcification, Necrosis And Acinar Pattern Arrangement Of Malignant Cells

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