MINI REVIEW

Survival Rate of Patients with Pancreatoblastoma (PBL)

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ABSTRACT

Pancreatoblastoma (PBL) is a very rare childhood tumour that develops from pancreatic epithelial exocrine cells. It is the most common malignant pancreatic tumour in children under the age of five, with a mean age of diagnosis of five years. It grows slowly, and its appearance is varied and frequently non-specific. If the tumour is resectable, the prognosis for paediatric cases is usually good, but recurrences do occur. PBL has an aggressive course and a poor prognosis if it is unresectable and has metastasis. Adult cases typically have a shorter survival time, with a median survival time of 15 months. Awareness of this rare pancreatic tumour is critical for early detection and management.

INTRODUCTION

PBL also known as infantile pancreatic carcinoma is an extremely rare pancreatic tumour that accounts for 0.5% of pancreatic non-endocrine tumours in children. Although PBL is most commonly seen in children, it can also occur in adults. When compared to adults, PBL is less aggressive in infants and children. Children with PB typically present later in life with upper abdominal pain and a palpable mass in the epigastrium. Mechanical obstruction of the upper duodenum and gastric outlet by a pancreatic head tumour may result in vomiting, jaundice, and gastrointestinal bleeding. Histologically, PB has distinct acinar and squamoid cell differentiation [1].

The majority of cases of PBL, a rare neuroendocrine tumour, are seen in children. At least sixteen case reports of PBL in patients aged 19 or older have been described to date. The mainstay of curative treatment is surgical resection. Even patients with liver metastasis can live a disease-free life for a long time. A 33-year-old male presented to us for a right hepatic lobectomy to remove the presumed primary tumour, which was later discovered to be a metastasis, followed by a pancreaticoduodenectomy to remove the true primary lesion. This patient is the longest disease-free survivor of metastatic adult PBL five years after resection [2].

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Tumor size, organ of origin, definition and quality of tumour margins, tumour heterogeneity, calcification, enhancement, ascites, biliary and/or pancreatic ductal dilatation, local invasion, adenopathy, vascular invasion, vascular encasement, metastases, and signal intensity on MR images were reviewed in 10 patients with pathologically proven PBL. The results of ten CT, seven US, and three MR imaging exams were reviewed [3].

Adenocarcinoma, neuroendocrine tumours, pancreatic cystic neoplasms with solid component, solid pseudopapillary tumour, PBL, pancreatic lymphoma, and pancreatic metastasis are the most common solid pancreatic lesions. Adenocarcinoma is the most common type of pancreatic lesion. The diagnosis of these lesions can be difficult, but there are currently imaging techniques with high sensitivity and specificity, such as CT scan, EUS, and MRI. The MRI, with its extremely high soft-tissue contrast resolution, aids in the detection and staging of adenocarcinoma. In patients with clinical suspicion of pancreatic lesion, a multimodality approach is usually required. In the case of pancreatic cancer, the local evaluation of the lesion's relationship with vessels, as well as tissue acquisition and the CT scan, MRI is usually required for local and distant staging [4].

PBL is a very uncommon pancreatic neoplasm. The current study sought to examine the clinical, pathological, and immunohistochemical characteristics of PBL and to discuss its management. The tumours were immunohistochemically differentiated as acinar, endocrine, and ductal. The average length of survival was 25.5 months. These findings suggest that the diagnosis of PBL is primarily based on pathological findings, that surgical resection is the most effective method of cure, and that adjuvant chemotherapy may be a good palliative method in some patients [5].

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Childhood pancreatic tumours (PT) are uncommon. There are no standard therapeutic approaches. Our goal was to examine treatment options and outcomes in children with PT. Medical records were reviewed, and patient data was anonymized. There were two cases of tumour relapse and one case of late secondary pancreatic cancer. Chemotherapy, radiotherapy, and surgical intervention concepts varied greatly in P-CA and PBL. Reference pathology and radiology should be involved in all paediatric PT cases. Standardized treatment concepts and prospective data registrations must be established [6].

CONCLUSION

PBL is a heterogeneous tumour with well-defined margins that appears to originate in the pancreas or the liver. It can act aggressively, with localized vascular or bowel invasion, or it can spread widely. Although uncommon, an upper abdominal mass in a child should be considered in the differential diagnosis.

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