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ORIGINAL ARTICLE

Diagnosis and treatment of pancreatoblastoma in children: a retrospective study in a single pediatric center

YiJin Huang[1] · Wei Yang[1] · JiaJian Hu[1] · ZhiYun Zhu[1] · Hong Qin[1] · Wei Han[1] · HuanMin Wang[1]

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Abstract

Background Pancreatoblastoma is a very rare malignant pancreatic tumor in children. Pancreatoblastoma is the most common pancreatic tumor in children less than 10 years of age, accounting for 25% of the pancreatic neoplasm. There were only a few published literatures about the standardized diagnostic and management protocol for PB in the last decade.

Objective To summarize our experience in the management of pancreatoblastoma in children and adolescents with emphasis on the presentation, diagnosis, treatment, and outcomes. A management strategy will also be discussed.

Methods This was a retrospective case-series study of all pancreatoblastoma in patients < 18 years of age who were treated at Beijing children's hospital (BCH) from January 2002–January 2015. The diagnoses of PB were confirmed by histopathology analysis of the resected specimen. The variables being analyzed included patient demographics, age at diagnosis, clinical presentation, tumor size, metastasis if present, tumor markers (AFP), type of surgery, length of follow-up, and outcome. The assessment of the tumor location, size, extent of the tumor, and distant metastasis was made by ultrasound (US), computed tomography (CT), and/or magnetic resonance imaging (MRI).

Result 21 patients with pancreatoblastoma were diagnosed at a median age of 4 years, 7 girls, and 14 boys. The diagnosis of pancreatoblastoma was identified by the histology examination. The most common syndrome was abdominal mass ($n = 11$), followed by abdominal pain ($N = 10$), elevated serum AFP levels were noted in almost all cases (17/18), 17 patients with disease initially unresectable on diagnosis accepted neo-adjuvant chemotherapy consisting of CDV, OPEC, PLADO, IEV, and AVCP. All patients underwent surgery, including pancreaticoduodenectomy (Whipple's procedure), the Pylorus-preserving pancreaticoduodenectomy (traverse-Longmire procedure), Spleen-preserving distal pancreatectomy, and distal pancreatectomy with en bloc splenectomy, Roux-en-Y end-to-end pancreatojejunostomy. In all, 13 children were disease free with a median follow-up of 53 months (range 11–156 months).

Conclusions The pancreatoblastoma in children and adolescents is a malignant tumor. Complete resection combined with chemotherapy is associated with long-term survival. For the unresectable tumor at diagnosis, preoperative chemotherapy was recommended to reduce tumor volume. AFP is critical for diagnosis and monitoring the disease as a tumors marker.

Keywords Pancreatoblastoma · Children tumor · Surgery · Tumor · Pediatric · Rare tumor

✉ HuanMin Wang
bchwanghuanmin@163.com

YiJin Huang
hyj19912006@gmail.com

Wei Yang
yw092011@163.com

JiaJian Hu
mdrhu019@163.com

ZhiYun Zhu
yun_52@163.com

Hong Qin
qinhong999999@163.com

Wei Han
dochanwei@aliyun.com

[1] Division of Pediatric Oncological Surgery, Department of Pediatrics, Beijing Children's Hospital, Capital Medical University, Beijing, China

Introduction

Pancreatoblastoma(PB) is a rare pancreatic malignancy in children [1, 2]. In US, the estimated overall incidence is around 0.004 cases per 100,000 people [3], and PB usually presents during childhood and adolescence, although it is possible to occur in adulthood [4]. Nevertheless, PB is the most common pancreatic tumor in children less than 10 years of age, accounting for 25% of the pancreatic neoplasm [5]. There were only a few published literatures about the standardized diagnostic and management protocol for PB in the last decade [6, 7].

Purpose

The aim of this study was to review our experience in the management of PB in children with emphasis on the presentation, diagnosis, treatment, and outcomes. A management strategy will also be discussed.

Methods

This was a retrospective case-series study of all pancreatoblastoma in patients < 18 years of age who were treated at Beijing children's hospital (BCH) from January 2002–January 2015. The diagnoses of PB were confirmed by histopathology analysis of the resected specimen.

The variables being analyzed included patient demographics, age at diagnosis, clinical presentation, tumor size, metastasis if present, tumor markers (AFP), type of surgery, length of follow-up, and outcome.

The assessment of the tumor location, size, extent of the tumor, and distant metastasis was made by ultrasound (US), computed tomography (CT), and/or magnetic resonance imaging (MRI).

Histologic diagnosis was made by light microscopic examination of the specimens and immunohistochemical analyses as well. Surgical excision was performed in all cases.

Results

Patient's characteristics

During the study period, 21 pancreatoblastoma patients aged from 2 months to 10 years were treated at our institution, of which 7 patients (case 2, 7, 10, 11, 12, 13, and 16) were referred from other centres. The median age at presentation was 4 years. There were 7 girls and 14 boys, and the demographic and clinical features of the 21 patients are listed in Table 1. Most of the patients presented with the abdominal mass or abdominal pain, jaundice, fever, abdominal distension, and anorexia were also present. One child (case 11) who was found in a newborn had features of Beckwith–Wiedemann syndrome and also expressed the typical "dysmorphic" faces and a clubbed foot. He also suffered from hypoglycemia before the diagnosis of PB was made.

Diagnosis

Abdominal ultrasound, computed tomography (CT), and/or magnetic resonance imaging (MRI) was performed to define the site, inner ingredients, and relationship with the adjacent structures. The tumors located in different parts of the pancreas with similar frequency; for near half of the patients ($N=9$; 42.85%), the tumor was large and occupied at least two parts of or the entire pancreas. The most common location of the tumor was at the head of the pancreas ($N=7$; 33.3%), the tail ($N=3$; 14.2%), and the body ($N=2$; 9.52%). There were 10 patients who presented with metastatic disease (Table 1).

The serum levels of AFP were measured in 17 out of 21 patients. With an elevated AFP level in almost all patients (16/17) and the median level of 6022.7 μg/L, (2.08–54,000), the AFP seemed to have no correlation with the primary tumor size. However, normalisation of AFP was observed upon the completion of chemotherapy and surgical resection.

The percutaneous needle biopsy was performed in ten patients; the histologic examination reveals the acinar and trabecular areas associated with the nests of squamous epithelium. (Squamoid nests), the immunohistochemical revealed CK (+), AAT (+), confirming the diagnosis of pancreatoblastoma.

Treatment

In 17 of 21 patients, the tumor was unresectable at diagnosis and these patients were treated with neo-adjuvant chemotherapy consisting of CDV, OPEC, PLADO, IEV, and AVCP. The number of administered cycles ranged from 2 to 6 (Table 2).

The different chemotherapy regimens used were:

OPEC: vincristine (1.5 mg/m^2/day on day 1), cyclophosphamide (1200 mg/m^2 on day 1) cisplatin (90 mg/m^2/day on days 2). VP16 (150 mg/m^2/day on day 4).

PLADO: cisplatin (100 mg/m^2 on day 1) and doxorubicin (60 mg/m^2 on day 1).

IE: ifosfamide (1.5 g/m^2/day on day 1–day 5), VP16 (100 mg/m^2/day on day 1–day 3).

Table 1 Clinical course and therapeutic management of pancreatoblastoma

	Age	Gender	Site	The presenting symptoms	Extension	AFP (ng/ml)	Preoperative chemotherapy	Type of surgery	Resection margin	Outcome (mouths after diagnosis)
1	1 year	B	Tail	Abdominal mass	Tumor thrombus in splenic vein	66	No	Distal pancreatectomy with en bloc splenectomy	R0	NED (50)
2	4 years	B	Body Tail	Abdominal mass	Splenic vein + mesenteric vein	3616	6 PLADO[a]	Pylorus-preserving pancreaticoduodenectomy	R0	NED (39)
3	7 years	B	Head Body Tail	Abdominal pain + abdominal mass	Tumor thrombus in splenic vein,mesenteric vein, portal vein	209.19	3 CDV[c] 2 OPEC[d] 1CDV + 5-FU 1 IFO + P+VP-16	Distal pancreatectomy with elective splenectomy	R0	Lost (12)
4	7 months	B	Tail	Abdominal mass	No	57.55	NO	Spleen-preserving distal pancreatectomy	R0	NED (52)
5	5 years	G	Body Tail	Abdominal pain + abdominal mass + anorexia	Tumor thrombus in splenic vein	2307.42	2 CDV[c] 2 OPEC[d]	Distal pancreatectomy with elective splenectomy	R0	NED (81)
6	4 years	G	Head	Abdominal pain + jaundice	No	60	1 CDV[c] + 5-FU 1 OPEC[d]	Pylorus-preserving pancreaticoduodenectomy	R0	NED (23)
7	6 years	B	Head Body Tail	Abdominal mass	No	1662	No	Spleen-preserving distal pancreatectomy	R0	NED (11)
8	8 years	B	Body Tail	Abdominal mass	Tumor thrombus in left kidney vein and splenic vein	6069	3 CDV[c] 2 OPEC[d]	Spleen-preserving distal pancreatectomy and kidney vein and splenic vein resection with restruction of kidney vein	R0	Lost (1)
9	3 years	G	Body	Abdominal mass	No	2140	2 VAC 2 VCE	Central pancreatectomy with Roux-en-Y end-to-end pancreaticojejunostomy	R0	NED (50)
10	3 years	B	Body Tail	Abdominal mass	No	6412	No	Central pancreatectomy with Roux-en-Y end-to-end pancreaticojejunostomy	R0	Alive
11	2 months	B	Body	Beckwith–Wiedemann	No	23,748	No	Central pancreatectomy with Roux-en-Y end-to-end pancreaticojejunostomy	R0	NED (40)
12	4 years	B	Head	Abdominal pain	Liver metastasis and mesenteric vein	2.08	3 AVCP 2 IEV 1 IFO + VP + VCR	Abdominal cavity drainage	No	DOD
13	4 years	B	Head	Abdominal pain	No	ND	4 OPEC	Pancreaticoduodenectomy(Whipple operation)	R0	NED (42)

Table 1 (continued)

	Age	Gender	Site	The presenting symptoms	Extension	AFP (ng/ml)	Preoperative chemotherapy	Type of surgery	Resection margin	Outcome (mouths after diagnosis)
14	4 years	B	Tail	Abdominal pain	Liver metastasis tumor thrombus in splenic vein	426.87	5 OPEC	Spleen-preserving distal pancreatectomy resection of liver metastatic lesions	R0	NED (48)
15	5 years	B	Body Tail	Abdominal pain + fever	Tumor thrombus in splenic vein	68.87	2 CDV[c] 2 OPEC[d]	Spleen-preserving distal pancreatectomy + splenic vein resection	R0	NED (48)
16	6 years	B	Head	Abdominal distension	Liver	ND	No	Pancreaticoduodenectomy (Whipple operation)	R0	Lost (12)
17	10 years	B	Head	Abdominal pain + jaundice	No	ND	2 CDDP+ ADR/VP16	Pancreaticoduodenectomy (Whipple operation)	R0	NED (156)
18	4 years	G	Head	Jaundice	Liver	7000	4 CDV[c]	Pancreaticoduodenectomy (Whipple operation)	R0	Lost (6)
19	2 years	G	Head	Abdominal pain + abdominal mass	No	564.85	No	Central pancreatectomy with Roux-en-Y end-to-end pancreaticojejunostomy	R0	NED (74)
20	3 years	G	Head Body Tail	Abdominal pain	Tumor thrombus in splenic vein	ND	2 CDV[c] 2 OPEC[d]	No	No	DOD
21	6 months	G	Head Body Tail	Diarrhea	Lung Liver	54,000	2 CDDP + VP16	No	No	DOD

PLADO[a] cisplatin + doxorubicin, *DOD* died of disease, *IE* Ifosfamide + VP-16, *ND* not done, *CDV* Cyclophosphamide + Daunorubicin + Vincristine, *NED* no evidence of disease, *OPEC* vincristine + Cyclophosphamide + Cisplatin + VP16

Table 2 Chemotherapy administered and the effect

	Before chemotherapy			AFP (ng/ml)	Chemotherapy	After chemotherapy	
	Site	Size	Extension			Size	AFP
1	Tail	6.9×5.7×8.5 cm	Tumor thrombus in splenic vein	66	No	6×2.5×3.5	2.68
2	Body Tail	10.4×7.2×8.6 cm	Splenic vein + mesenteric vein	3616	6 PLADO[a]	3.5×2.4×3.8 cm	1.42
3	Head Body Tail	12.5×9.5×8.5 cm	Tumor thrombus in splenic vein	209.19	3 CDV[c] 2 OPEC[d] 1CDV + 5-FU 1 IFO + P+VP-16	5.0×6.5×3.6 cm	9.55
4	Tail	6.1×5.9×6.9 cm	No	57.55	NO	6.5×6×5.5 cm	7.55
5	Body Tail	9.1×7.6×9.8 cm	Tumor thrombus in splenic vein	2307.42	2 CDV[c] 2 OPEC[d]	5.5×4.7×6.0 cm	15.2
6	Head	4.5×3.1×3.6 cm	No	60	1 CDV[c] + 5-FU 1 OPEC[d]	4×2.8×2.5 cm	9.4
7	Head Body Tail	15.4×10.8×7.8 cm	No	1662	ND	6.4×4.5×6.6 cm	8.38
8	Body Tail	9.6×9.6×13.2 cm	Tumor thrombus in left kidney vein and splenic vein	6069	3 CDV[c] 2 OPEC[d]	6.1×3.0×5.8 cm	8.22
9	Body	8.6×6.8×7.6 cm	No	2140	2 VAC 2 VCE	4.8×4.4×5.5 cm	41,991
10	Body Tail	8.8×7.7×9.8 cm	No	6412	ND	4.0×2.1×3.8 cm	54.01
11	Body	4.3×4×3.5 cm	No	23,748	No	–	–
12	Head	4.5×3.0×4.8 cm	Liver metastasis and mesenteric vein	–	3 AVCP 2 IEV 1 IFO + VP + VCR	–	–
13	Head	2.6×2.5×4.0	No	–	4 OPEC	–	–
14	Tail	9.1×6.9×6.0 cm	Liver metastasis tumor thrombus in splenic vein	426.87	5 OPEC	3.4×2.9×3.4 cm	2.87
15	Body Tail	10.6×6.3×10.2 cm	Tumor thrombus in splenic vein	68.87	2 CDV[c] 2 OPEC[d]	5.2×2.5×4.3 cm	2.12
16	head	10×5 cm	Liver	–	–	4.1×4.3×4.5 cm	4.64
17	head	ND	No	ND	2 CDDP + ADR/VP16	ND	ND
18	Head	8.2×7.9×9.1 cm	Liver	7000	4 CDV[c]	4.4×2.9×3.0 cm	28
19	Head	7×5×5 cm	No	564.85	NO	–	–
20	Head Body Tail	13.4×15.3×13.7 cm	Tumor thrombus in splenic vein	ND	2 CDV[c] 2 OPEC[d]	11×11×10 cm	ND
21	Head Body Tail	7.7×6.1×5.8 cm	Lung liver	54,000	2 CDDP + VP16	ND	43,060

PLADO Cisplatin + Doxorubicin, *IE* Ifosfamide + VP-16, *CDV* Cyclophosphamide + Doxorubicin + Vincristine, *OPEC* Vincristine + Cyclophosphamide + Cisplatin + VP16

CDV: cyclophosphamide (2.1 g/m^2/day on day 1–day 2) + daunorubicin (25 mg/m^2/day on day 1–day 3) + vincristin (0.67 mg/m^2/day on day 1–day 3).

The chemotherapy was able to shrink the tumor and has facilitated subsequent resection in all of the 17 children. Depending on the location of the tumor, various procedures including the pancreaticoduodenectomy (Whipples procedure), the pylorus-preserving pancreaticoduodenectomy (traverse-Longmire procedure), splenic-preserving distal pancreatectomy, and distal pancreatectomy with en bloc splenectomy and a Roux-en-Y end-to-end pancreatojejunostomy have been performed. The operations performed are summarized in Table 1.

In case no 3, after total resection of the tumor including the tumor thrombus in the splenic vessels (vein + artery) and superior mesenteric veins, the splenic survived by the bypass vessel; the patient underwent the spleen-preserving distal pancreatectomy.

Pathology

All resected specimen were subjected to histological study. The diagnosis of PB was confirmed with histology showing typical squamous corpuscles and tumor cells that had acinal, glandular, or undifferentiated appearance. In addition, immunohistochemical studies were performed and the results are summarized in Table 3.

Outcome

Post-operatively, all patients were referred to oncologists for further management. During follow-up, serum AFP level and ultrasound scan were performed.

At the time of the analysis, 5-year OS were 94.4%; 14/21 patients were disease free. At a median follow-up of 53 months (range 11–156 months), most of them (13/14) received a complete resection.

There was one patient (case 10) with tumor recurrence. At 10 months from the initial diagnosis in another hospital, the patient had an elevated serum AFP and ultrasound showed tumor recurrence at the body of the pancreas. The patient was transformed into our hospital; after the four cycles of chemotherapy, the boy underwent a complete resection. After 13 months of the second surgery, the boy had a tumor relapse at the head of the pancreas. Their patients decided to receive the traditional therapy (Traditional Chinese medicine). The boy is disease free with follow-up of 25 months.

In case 12, a 4-year-old boy presented with abdominal pain, and physical examination revealed a large mass in the upper abdomen. Abdominal ultrasound and computerized tomography scan showed a large (45.6 × 30.4 × 48.9 cm) mass from the head of the pancreas and metastasis to the liver and mesenteric lymph nodes. The AFP level was subsequently normalised. The diagnosis of pancreatoblastoma was confirmed by the open biopsy. After a combination of chemotherapy regimen consisting of two courses of AVCP, 2 courses of IEV, IFO + VP + VCR, the size of the tumor was found to be 5.4 × 4.1 × 4.0 cm. However, the boy presented with severe ascites and multiple metastases were noted in the liver and mesentery. He was managed with palliative care.

Table 3 Results of the immunohistochemistry

Immunohistochemistry stains	Frequency, % (No. positive cases/total cases)	
CK	84	11/13
AAT	100	13/13
CD56	50	4/8
Vimentin	80	8/10
CD10	72	8/11
Insulin	0	0/8
β-Catenin	88	8/9
SYN	62	5/8
CK7	66	4/6
CEA	80	4/5

Discussion

In 1957, Becker [8] reported pancreatoduodenectomy for carcinoma of the pancreas in an infant, and the histopathology examination showed squamous appearance. In 1977, pancreatoblastoma was described by Horie [9] based on the morphogenesis of the tumors. In 2014, The Papanicolaou Society of Cytopathology named the term "pancreatoblastoma". This tumor is a rare disease with a low incidence. Although extremely rare, pancreatoblastoma is more common in Asians than in the white population [10]. The 5-year overall survival is approximately 66% in pancreatoblastoma [11], pancreatoblastoma usually occurs in children younger than 10 years [3], with a median age of 4–5 years [7, 12], some adults cases have been reported [4], and neonates with pancreatoblastoma have also been reported [13]. There is a slight preponderance in male (males:female ratio of 1.14:1) [6]; in our study, 14 of the 21 cases were boys and the median age was 4 years (2 months–10 years).

The PB is a slow-growing tumor and the clinical presentation can be non-specific. The typical presentation is a large palpable abdominal mass and abdominal pain with or without vomiting. Tumor rupture and hemorrhage have also been reported. Our study demonstrated that the PB tends to be diagnosed at an advanced stage with approximately half of patients (11/21) were diagnosed with the tumor has extended beyond the pancreas or was metastatic. Pancreatoblastoma most often originated from the pancreas, but origination from the mesentery has been reported [14].

Klimstra et al. reported that 35% of patients with pancreatoblastoma presented with metastatic, and the most common metastatic site was the liver, regional lymph nodes, lung [6], or brain [15]. In some case, pancreatoblastoma can be associated with Beckwith–Wiedemann syndrome [16] and/or develop a hepatoblastoma [17]. The familial adenomatosis polyposis syndrome has also been associated with pancreatoblastoma [18]. Abraham et al. demonstrated that allelic loss on chromosome 11p was the most common genetic alteration in pancreatoblastoma, present in 86% (six of seven informative cases) [18]. In a repeated case, a newborn with Beckwith–Wiedemann syndrome presented the simultaneous occurrence of a pancreatoblastoma and an adrenal neuroblastoma [19].

PB should be considered when physical examination shows an asymptomatic mass in the upper abdomen, elevated serum AFP level. Previous studies reported that serum

AFP is always elevated [20]. In our study, with the presence of elevated AFP were found in 17/18 patients at diagnosis. Although AFP seemed to have no correlation with the primary tumor size, and the serum levels of AFP can be normal in the patients with metastatic [7]. However, failure to normalise was observed upon the chemotherapy and complete resection of the tumor. To monitor levels of serum, AFP is useful to follow the course of the pancreatoblastoma. In the series, one patient relapsed with coincident elevations of the AFP levels.

Pancreatoblastoma also releases AFP and other hormones and, therefore, can lead to endocrine syndromes such as Cushing syndrome and antidiuretic hormone [12].

The diagnosis of PB could be established with ultrasound, CT, and MRI which could clearly demonstrate the size, shape, and characteristic of the lesion. However, sometimes, it is difficult to discriminate a PB from the neuroblastoma.

Tissue diagnosis by biopsy is recommended in cases of pancreatoblastoma. Actually, a biopsy is NOT recommended for all pancreatic tumors. For instance, a tumor that appears to be solid pseudopapillary neoplasm in a female teenager does not need a biopsy but a resection. If clinical diagnosis is difficult;we still recommend the biopsy. It may be performed through laparotomy, laparoscopy, or percutaneous core needle.

Grossly, pancreatoblastoma are similar to acrinar cell carcinomas–solitary, well-demarcated [21], and squamoid nests can be useful to establish the diagnosis.

Compared to adults, PB is less aggressive in the children and adolescents. Although no standard treatment for the pancreatoblastoma has been established. The most important prognostic factor is complete surgical excision. The prognosis is good in cases of complete removal.

The resectability depends on the tumor location in the pancreas and the presence of metastasis. In the absence of metastatic disease, surgical resection is most commonly associated with long-term survival [3, 5, 6]. In our series, primary surgical resection was often not possible in most cases due to the extent of the tumor as well as vascular invasion or metastasis. Chemotherapy remains a cornerstone for the management of unresectable tumors [12]. From the previous literatures, pancreatoblastoma is chemosensitive and usually responsive to neo-adjuvant chemotherapy [22–25].

Up to now, varied chemotherapy regimens have been used, such as Cisplatin, doxorubicin, etoposide, cyclophosphamide, vincristine, actinomycin D, gemcitabine, or ifosfamide and carboplatin which have proved to be effective chemotherapeutic agents [7, 22, 26]. Compared to the hepatoblastoma, the pancreatoblastoma are composed of similar genetic alterations which includes the changes in the APC/β-catenin pathway, loss of heterozygosity in chromosome 11p and IGF2 overexpression [18], In some cases, PLADO was used as the chemotherapy regimen [12]. Defachelles et al. recommended preoperative chemotherapy with cisplatin and doxorubicin (PLADO) to reduce the tumor volume of unresectable tumors [25]. Preoperative chemotherapy with irinotecan and vincristine was extremely effective for patients who failed to response to first-line intensive chemotherapy. The patient from whom the CMBS-PB cell line was derived had no response to the multimodal chemotherapy, progressed rapidly, and died [27]. In our hospital, chemotherapy could facilitate subsequent complete resection. However, the most effective preoperative chemotherapy has not yet been established, and our preoperative chemotherapy was a combination regimen (OPEC + CDV) which was for the treatment of advanced-stage neuroblastoma in China. In addition, in our study, the (OPEC + CDV)'s effectiveness has been proved. 9 of our patients with unresectable tumor received preoperative CDV + OPEC that allowed following surgical removal in 8 patients. 5 of them are disease free for a median time of 48.2 months. In addition, the AFP levels can be used as an indicator of response to chemotherapy or other treatment and for evidence of recurrence [5].

Surgical resection remains the most important treatment of pancreatoblastoma. In our study, we identified a total of 18 patients who had complete resection after chemotherapy or initially and 17 are complete remission. According to the location of the tumor, selection of surgical procedures is important.

For lesions at the head or uncinate process, pancreaticoduodenectomy (Whipple procedure) is there commended surgical option. With the advance of the surgical skills, pylorus-preserving pancreaticoduodenectomy (traverse-Longmire) has been performed in two patients. During the follow-up, the two patients had a good prognosis.

For tumors located in the body of the pancreas, the operative approach was central pancreatectomy with a Roux-en-Y end-to-end pancreaticojejunostomy.

Surgical resection of pancreatoblastoma located in the body or tail of the pancreas consists of a distal subtotal pancreatectomy to provide a margin negatively. The splenectomy has generally been performed in the past. However, the spleen plays an important role in the immunologic defense of the host. The spleen should be preserved if it can be safely achieved. In our experience, preserving the spleen during distal pancreatic resection was undertaken in two patients. During the surgical procedures, even when the splenic vein was ligated, the spleen could still survive through the collateral vessels. If the metastasis is solitary and confined to the liver, resection of the metastatic disease is also beneficial.

For some advanced disease, Yonekura completed aggressive surgical treatment including resection and reconstruction of the portal and superior mesenteric veins after preoperative chemotherapy [24], Ohata reported the primary tumor was completely resected with pancreaticoduodenectomy

and Child's pancreatic biliary tract reconstruction. The liver metastases were resected [28].

Radiotherapy was reserved for inoperable cases only [6, 25], and the effectiveness of radiation therapy for local control in pediatric patients remains undetermined.

Conclusion

In summary, PB in children and adolescent is a rare malignant tumor. The standard therapy is yet to be determined; in our experience, the presentation is varied and non-specific. The serum AFP levels play important roles in the diagnosis and follow-up of the PB. Preoperative chemotherapy is recommended in the unresectable or metastasis cases. However, the best chemotherapy regimen is still not clear. PLADO may be a good choice. Complete resection is the upmost important treatment for PB. Various surgical procedures have been developed to achieve complete resection while preserving the organ function. During follow-up, the measurement of AFP levels and the ultrasound should be performed to monitor recurrence.

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