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J Ped Surg Case Reports 3 (2015) 120–122

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

Congenital pancreatoblastoma: a case report

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ARTICLE INFO

Article history:
Received 23 October 2014
Received in revised form
2 December 2014
Accepted 9 December 2014

Key words:
Pancreatoblastoma
Congenital
Surgery

ABSTRACT

The literature describes 15 cases of congenital pancreatoblastoma (PB): 5 had prenatal diagnosis, none had metastases at diagnosis, 7 were associated with Beckwith–Wiedemann syndrome (BWS). In 13 cases resection was radical, while in 2 there were macroscopic residues. Only one patient underwent chemotherapy after distant recurrence. All children are alive except one who died because of problems related to BWS. Our goal is to describe the approach adopted in an infant with congenital PB treated in our center. After a prenatal third semester diagnosis of abdominal anechoic lesion, the radiological investigations (ultrasound, MRI) performed at birth described a cystic lesion of unclear nature. We proceeded to laparoscopic exploration, transformed into open approach after the detection of a lesion located in the body of the pancreas; this lesion was resected, preserving the head and tail of pancreas. The histological diagnosis showed a completely excised PB. After excluding metastatic lesions, we decided to perform only careful follow-up without chemotherapy. The follow-up at 12 months is negative. Although PB is a malignant tumor that requires a multidisciplinary treatment, the congenital cases seem to have a less aggressive biological behavior. The treatment, therefore, in case of complete resection, could be only surgical, followed by a careful follow-up. These forms are often associated with congenital BWS, but in our case the patient did not have the typical characteristics of the syndrome.

Pancreatoblastoma (PB) is a pancreatic malignant tumor. Although rare, with approximately 200 cases reported in literature [1], it is one of the most common pancreatic exocrine tumors in childhood [2].

The congenital form, defined as tumors detected before 3 months of age, is even less common. To date, 15 cases are reported worldwide, summarized in Table 1 [1,3].

aFP: α-fetoprotein; DOD: died of disease; GA: gestational age; N/A: not applicable; NED: no evidence of disease.[legend; see instruction]We report a case of congenital PB with prenatal diagnosis treated in our Center.

1. Case report

Prenatal diagnosis, at last trimester ultrasonography (US), described an anechoic lesion (40 × 40 mm) with intracystic, hyperechoic lesions, 18 × 24 and 6 × 5 mm respectively, developing in front of the left kidney.

The gestation was regular, and at birth the child was asymptomatic. No family factors were reported. After birth the patient underwent the following investigations:

- Abdominal US, performed at 2 days of life, confirmed the presence of a round lesion in the left upper abdominal quadrant, measuring 4.5 × 4.5 × 4 cm, containing approximately 42 ml of fluid; the walls presented two solid hypo-echogenic portions projecting into the lumen, of 20 and 7 mm respectively, the first with internal vascularization. This lesion seemed to be separated from the left kidney, spleen, and stomach, occupying the region of the pancreatic tail, with an otherwise normal pancreas, and it was unclear if this formation had a mesenteric, pancreatic or left adrenal origin.
- Abdominal MRI with contrast, performed at 8 days of life, identified a 5.1 × 4.6 × 5.5 cm round lesion located in the epigastrium, below the stomach and anterior to the body of the pancreas. It was not possible to visualize a plane between the pancreatic parenchyma and the lesion; the head and body

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http://dx.doi.org/10.1016/j.epsc.2014.12.004

Table 1
Congenital PB cases reported in literature.

Age	Presentation	Sex	Tumor location	Tumor size (cm)	aFP	Complete surgical resection	Postsurgical treatment	Outcome	BWS	Reference
20 wk GA	Abnormal US (cyst)	F	Head	11	Increased	Yes	None	NED, 2 y	Yes	[1]
28 wk GA	Abnormal US (cyst)	M	Body-Tail	4	Normal	Yes	None	NED, 26 mo	Yes	[1]
30 wk GA	Abnormal US (cyst)	M	Body	5.5	Normal	Yes	None	NED, 1 y		Current report
30 wk GA	Abnormal US (cyst)	M	Body-Tail	4.2	Normal	Yes	None	NED, 10 mo	No	[1]
30 wk GA	Abnormal US	M	Tail	7	Unknown	Yes	None	NED, 10 mo	Yes	[1]
34 wk GA	Abnormal US (cyst)	F	Body-Tail	4	Normal	Yes	None	NED, 15 mo	No	[1]
Newborn	Incidental finding	F	Head	1.5	Unknown	N/A	None	Stillborn	No	[1]
20 h	Mass	M	Body	10	Unknown	N/A	None	DOD at 12 d	Yes	[1]
3 d	Jaundice	M	Body-Tail	7.5	Increased	Yes	None	NED, 3 y	No	[1]
3 d	Palpable mass	M	Unknown	Unknown	Increased	Yes	None	Unknown	No	[3]
19 d	Mass	M	Body	7.5	Unknown	No	None	NED, 4 y	Yes	[1]
3 wk	Irritability, Jaundice	M	Head	6	Unknown	Yes	None	NED, 10 y	No	[1]
6 wk	Palpable mass	M	Tail	5	Unknown	Yes	None	NED, 29 y	Yes	[1]
2 mo	Hemihypertrophy	M	Head	4.5	Increased	Yes	None	NED, 5 y	Yes	[1]
3 mo	Incidental finding	F	Head	10.5	Unknown	No	Chemotherapy	Recurred at 7 mo; NED at 68 mo	No	[1]
3 mo	Mass	F	Tail	8	Unknown	Yes	None	NED, 39 mo	No	[1]

aFP: α-fetoprotein; DOD: died of disease; GA, gestational age; N/A, not applicable; NED, no evidence of disease.

anatomy were normal. The lesion had relatively thick walls (about 2–3 mm) with at least 4 solid thickenings of 10 and 22 mm. After contrast injection both the wall and the solid thickenings were enhancing (Fig. 1).

The initial diagnostic impression was that of a mesenteric cyst, and, at the age of 21 days, the patient was admitted by our Center, to be operated on.

The operation was first carried on with a minimally-invasive technique. During exploration of the abdomen, the lesion, with mixed solid and cystic appearance, was clearly originating from the body of the pancreas. The laparoscopic approach was aborted, and the tumor was completely excised through an open laparotomy (Fig. 2). When it was removed, part of the inferior body remained as a bridge between head and tail of pancreas. Since we were not completely confident the duct was sectioned, it was decided not to resect the whole distal part of pancreas.

The histological diagnosis was pancreatoblastoma.

Cancer markers were evaluated: αFP values were always between the age range, and consistently decreased over time (9107 ug/L at d23, 5734 ug/L at d29, 1126 ug/L at d45); HCG and CEA values were negative. After surgery, the patient underwent oncologic staging with a chest and abdominal CT scan. Metastatic disease was excluded and, according to the management described in literature for congenital cases, it was decided to perform only a careful follow-up without adjuvant chemotherapy.

The follow-up at 12 months is negative.

2. Discussion

The initial differential diagnosis of congenital abdominal cystic lesions includes malformations (enteric duplication cyst, mesenteric cyst) and neoplastic lesions (teratoma, neuroblastoma). Even if rare, PB should be considered in the differential diagnosis.

The radiologic diagnosis of PB can be difficult, and even in large primary tumors, it is not always possible to identify the pancreas as the organ of origin. At US, PB presents heterogenous echogenicity, often with hypoechoic cysts and echogenic septa; MRI and CT scan can help in defining localization and the intrinsic characteristics, such as presence of necrosis, septa or calcifications [4].

To date, 15 cases of congenital PB are described in literature. One third (5/15) of them had a prenatal diagnosis, and none had metastases at diagnosis. An important association with BWS was reported, involving almost half of the patient (7/15).

PB is a malignant tumor that requires a multidisciplinary treatment. Chemotherapy including cisplatin and anthracyclines, similar to that used for hepatoblastoma, seems to be the most appropriate treatment. However, the 5-year survival still remains unsatisfactory. Prognosis remains linked to the possibility of a radical resection and the absence of metastases [5].

All the congenital cases described in literature underwent initial surgical treatment: in 13 patients the resection was radical, while macroscopic residues were present in 2 children. The patients with

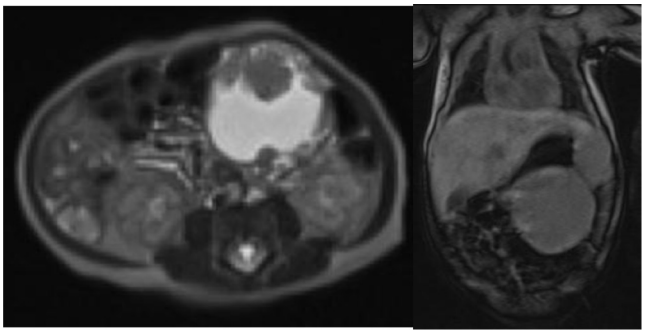

Fig. 1. MRI.

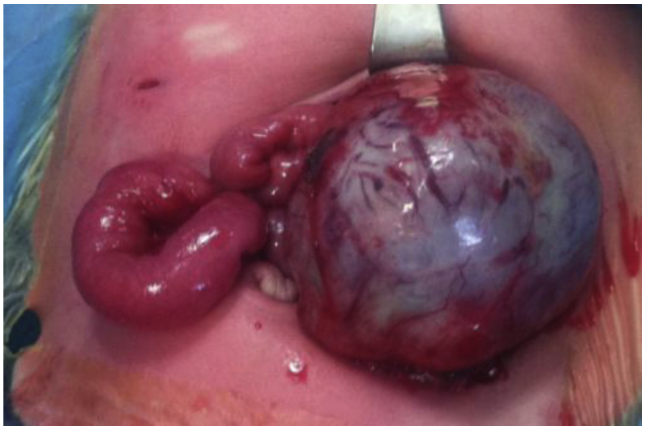

Fig. 2. PB at laparotomy.

complete resection were not further treated with chemotherapy or radiotherapy. In one patient only, who had incomplete initial surgical resection, chemotherapy was utilized because of local recurrence at 7 months from diagnosis [1,6].

3. Conclusion

The congenital cases described in literature present higher association with congenital BWS than pediatric and adult cases; however in half cases congenital PB was isolated.

Despite the aggressive biological behavior in children, the congenital cases seem to have a more benign course. Therefore, in case of complete resection, the treatment should be only surgical, followed by a close follow-up. Cases however are rare, and larger series are probably needed to confirm a conservative approach.

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