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CASE REPORT

# A case of pancreatoblastoma prenatally diagnosed as intraperitoneal cyst

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**Abstract** Pancreatoblastoma in childhood is a very rare malignant tumor, but is considered to have a relatively good prognosis because of its low metastatic potential. We report 1-day-old female infant who was recently found to have an intraabdominal cyst on prenatal ultrasound examination. The tumor was a unilocular, cystic mass without invasion or metastasis to other organs, allowing total resection. It was diagnosed postnatally with pancreatoblastoma.

**Keywords** Pancreatoblastoma · Prenatal diagnosis · Cystic tumor

## Introduction

Pancreatoblastoma in childhood is a very rare malignant tumor, but is considered to have a relatively good prognosis because of its low metastatic potential. We report an infant who was recently found to have an intraabdominal cyst on prenatal ultrasound examination, and was diagnosed postnatally with pancreatoblastoma after surgical resection, and review the relevant literature (Figs. 1, 2, 3).

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## Case report

The patient was a 1-day-old female infant with an abdominal mass. At 34 weeks of gestation, fetal US had shown an upper abdominal cyst. Her family history was noncontributory. At 40 weeks and 5 days of gestation, the infant was born by spontaneous delivery, with a birth weight of 3,546 g. Postnatal US showed a cyst in the left upper abdomen, and the infant was referred to our department on the following day for further evaluation and treatment. She measured 50 cm in height, and weighed 3,500 g. There were no palpable cervical or body surface lymph nodes. Abdominal palpation revealed a soft mass, 3 cm in diameter, in the left hypochondrium. The laboratory findings at referral, including amylase, were within normal limits except for a slightly increased GOT level and an increased LDH level. Serum tumor markers (NSE, AFP, VHA, and HVA) were normal.

Abdominal ultrasound showed a $40 \times 38$ mm, encapsulated, cystic mass in the region surrounded by the stomach, pancreas, and spleen. The cystic mass was unilocular, and was well demarcated from the surrounding structures.

Fluoroscopic examination of the stomach showed a mass shadow compressing the stomach in the cranial direction. Contrast medium passed readily through the duodenum and small intestine.

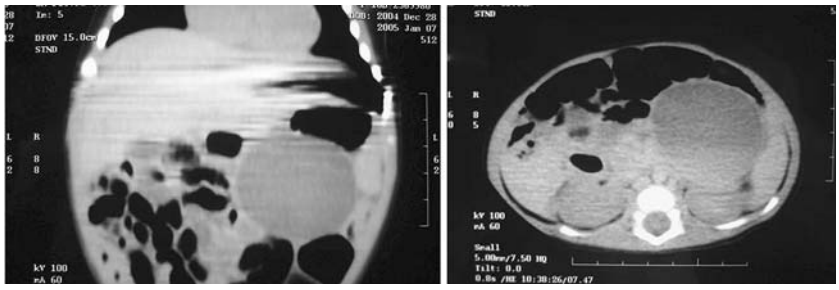

**Fig. 1** Abdominal CT revealed a 5 cm cystic lesion in the left abdominal cavity

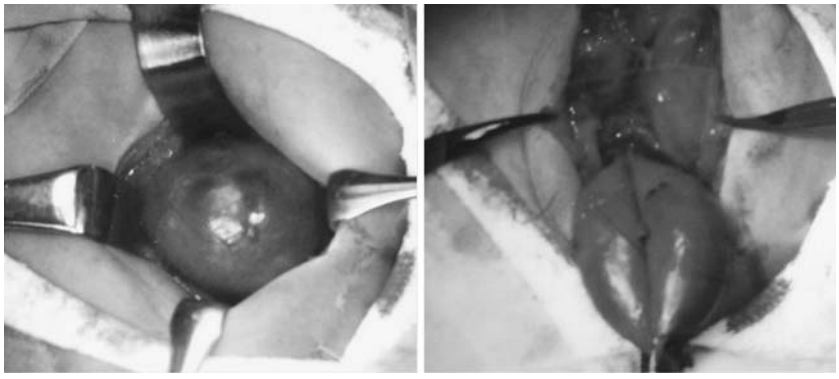

**Fig. 2** The mass originated in the tail of the pancreas and was encapsulated

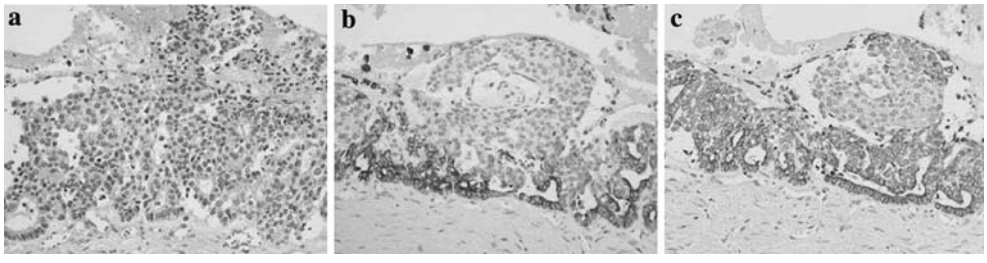


**Fig. 3** Histopathological findings showed a fibrous capsule, inside of which cords and tubules of cell with ovoid nuclei were seen. **a** HE ×200. Immunostaining was positive for CK7 and cytokeratin AE1/AE3. **b** CK7 ×200, **c** AE1/AE3 ×200

Abdominal CT revealed a 5 cm cystic lesion in the left abdominal cavity, which was unilocular and thin-walled, and formed a fluid–fluid level within the cyst.

Since the cystic lesion might shrink with time, we continued to observe the course of the cyst, but it tended to grow in size; therefore, we suspected cystic teratoma, and performed surgery. There was no ascitic fluid. The mass originated in the tail of the pancreas, and was encapsulated. The pancreatic tail mass was resected.

The resected specimen was an encapsulated round tumor, $40 \times 40$ mm in size, and contained 33 ml of reddish-brown fluid, with no solid tissue.

Histopathologically, the lesion appeared to be a cystic tumor arising in the pancreas, had a fibrous capsule, inside of which cords and tubules of cells with ovoid nuclei were seen. No squamoid corpuscles were recognized. Immunostaining was positive for cytokeratin AE1/AE3, CK7, trypsin and amylase. The proliferation (MIB1) was focally moderate. These findings lead to a diagnosis of cystic pancreatoblastoma.

Her postoperative course was uneventful, and she was discharged at one month of age. She received no postoperative chemotherapy. She has had no signs of recurrence during 15 months of follow up.

## Discussion

Pancreatoblastoma was first reported in 1959 by Frantz [1]. It is found more frequently in Orientals, and is considered to be a relatively rare tumor. In Japan, Horie et al. described two cases in 1974, and proposed that the tumor be designated pancreatoblastoma [2]. Compared with neuroblastoma and nephroblastoma, this tumor appears to develop in older children. There are many cases described as neonates and antenatally or in fetal autopsy but antenatal diagnosis and sub-

sequent treatment [3] is rare. Six cases have been reported in the newborn period [4–8]. All of these tumors had cystic morphology, and four of them had a large cyst. Three of the four tumors were associated with Beckwith-Wiedemann syndrome. The present tumor was not associated with Beckwith-Wiedemann syndrome. Defachelles et al. reported seven children with pancreatoblastoma, including a 3-month-old infant with the cystic form of the tumor [9]. Neonatal tumors are far more likely to be cystic than those seen in older children. Neuroblastomas detected in the newborn period are also cystic, suggesting a similar origin, and interestingly, possible spontaneous cure.

Cystic change of pancreatoblastoma is perhaps very rare phenomenan. We also had a definite diagnosis of pancreatoblastoma at surgery. On histological sections, the tumor cells were minimally presented inside the fibrous capsule, which indicated the origin of pancreatic exocrine cells. The moderate proliferative activity seemed be compatible with neoplastic growth of a primitive component of pancreas, neglecting a diagnosis of other cause, like traumatic, inflammatory, mucinous or serous cystic tumors.

Horie et al. subclassified pancreatoblastomas into ventral and dorsal types originating from the pancreas anlage and occurring in the head and the body–tail of the pancreas, respectively [10]. The dorsal type, as described here, is often detected late after metastasis to the liver, resulting in a poor prognosis. However, in the present case, an intraabdominal cystic mass was detected during fetal life. Although it was the dorsal type, its early detection allowed total resection, with a probable good prognosis.

Surgical resection is the treatment of choice. Pre- or postoperative chemotherapy is needed, if tumor invasion of other organs or distant metastasis to sites such as the liver is suspected [11]. Total resection is rarely associated with recurrence [12], and makes chemotherapy unnecessary, leading to a good prognosis.

Prognostic factors in this case remain unclear because of the small number of such cases. However, patients, such as reported here, who developed a cystic mass during fetal life, and in whom its complete resection is possible after birth, are expected to achieve complete cure.

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