Giant infantile immature teratoma derived from the hepatoduodenal ligament: Report of a case

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ARTICLE INFO

Article history:
Received 26 July 2013
Received in revised form 8 August 2013
Accepted 9 August 2013

Key words:
Teratoma
Hepatoduodenal ligament
Focal embryoid components

ABSTRACT

We report a 3-month-old female infant with giant immature teratoma derived from the hepatoduodenal ligament. The tumor was very large and the serum alpha-fetoprotein level was high, and it was initially misdiagnosed as hepatoblastoma at another hospital. However, it was confirmed to be an extrahepatic tumor by abdominal CT and ultrasonography at our institute.

We performed laparotomy and the frozen section of biopsied specimen revealed the tumor to be a teratoma. The tumor originated from the hepatoduodenal ligament and was excised. The final pathological diagnosis was immature teratoma with focal embryoid components.

Teratomas are the most common germ cell tumors. They are often seen in the sacrococcygeal region, retroperitoneum, mediastinum, and pineal gland of the brain, and are rarely seen in the neck, stomach and vagina [1]. Here we describe an extremely rare case of teratoma that originated from the hepatoduodenal ligament. The tumor had initially been misdiagnosed as hepatoblastoma because it was so huge that it compressed the left lobe of the liver and the patient showed a high alpha-fetoprotein (AFP) level. The pathological diagnosis was immature teratoma with focal embryoid components, which was also a rare feature of the disease.

1. Case

A 3-month-old female infant presented abdominal distention and was seen by a local doctor. An abdominal mass was noticed and subsequent abdominal CT revealed a huge mass in the upper abdomen. Blood test showed a high level of AFP. From these findings, the doctor diagnosed the mass as hepatoblastoma in the left lobe of the liver and started chemotherapy with platinum. The tumor did not react to the therapy and she was referred to our hospital for possible necessity of living related liver transplantation.

At our hospital, blood test revealed normal bilirubin level and liver functions, but an elevated AFP level (40,923 ng/ml). Abdominal CT showed a 10 × 7 cm mass that was composed of heterogeneous components including a cystic area, calcified components and a solid part (Fig. 1a and b). The mass compressed the left lobe of the liver and displaced the stomach to the ventral side. Abdominal ultrasonography showed an important finding that the mass was clearly separate from the liver and it was assured to be an extrahepatic tumor. There was no ascites nor lymph node enlargement. These preoperative images were compatible with a teratoma and the differential diagnoses were undifferentiated sarcoma or mesenchymal hamartoma. The patient showed prominent distention of the abdomen and she underwent laparotomy. The tumor was confirmed to be of extrahepatic origin and compressed the stomach to the ventral side (Fig. 1c) and the left lobe of the liver to the cranial side. In order to confirm the pathological diagnosis, the tumor was biopsied and the frozen section of the specimen revealed it to be a teratoma. The tumor was strongly adherent to the left side of the...
hepatoduodenal ligament, which we judged was the site of tumor origin. We excised the tumor with no injury to the portal vein, hepatic artery and extrahepatic biliary tree. There was no metastatic lymph node and no disseminated tumor in the peritoneal cavity. The tumor size was $16 \times 13 \times 11$ cm, and its weight was 921 g. Histological examination revealed that the tumor was composed of cysts of various sizes and solid lesions with various tissues such as glial cells, neuroepithelium, cartilage, pancreatic tissue, and choroid. Very focal AFP-positive cell clusters were identified by immunohistochemistry (Fig. 2). The final histological diagnosis was immature teratoma with focal embryoid components. The postoperative course was uneventful and serum AFP rapidly decreased after the operation (Fig. 3). Postoperative chemotherapy was not introduced

Fig. 1. a. Abdominal CT scan shows a cystic and solid huge tumor with local calcification, that expanded to compress the surrounding abdominal organs. Sagittal view. b. The tumor compressed the left lobe of the liver. The margin of the tumor is sharp and clear, which indicated the tumor to be of extrahepatic origin. Transverse view. c. A large tumor was seen in the upper abdomen. The stomach was elongated on the ventral side by the tumor. (The left side is the cranial side of the patient.)

Fig. 2. Microscopic appearance of the tumor shows a focal embryoid component whose cells were AFP positive (original magnification $\times 200$).

Fig. 3. The change of serum AFP level: serum AFP decreased rapidly after the first operation and then gradually decreased to the normal level. An arrow 1 indicated a first operation, and an arrow 2 indicated a second operation.
because the tumor was macroscopically resected and the embryoid primitive components were very restricted. She has been carefully followed at the outpatient clinic. 5 months after the first operation local recurrence of the tumor was detected by abdominal CT although serum AFP did not increase (30.6 ng/ml) and the lesion was resected. The resected specimen was mature teratoma. Our case was immature teratoma with a high AFP level (it was high considering the age correction) and it grew rapidly in a few months, which was a very unusual clinical course compared to the other 10 cases.

2. Discussion

Only 10 cases of teratoma originating from the hepatoduodenal ligament have been reported in the English literature (Table 1) [2–10] and our report is the 11th case. Among them, 3 cases were in adults, and only one case was malignant teratoma with a high AFP level whereas 8 out of 10 cases were mature teratoma. Our case was immature teratoma with a high AFP level (it was high considering the age correction) and it grew rapidly in a few months, which was a very unusual clinical course compared to the other 10 cases.

Teratoma derived from the hepatoduodenal ligament is very rare, but it should be kept in mind if a solid and cystic heterogeneous tumor is seen at the porta hepatis. In our case, the tumor was so huge and AFP was so high that it was initially misdiagnosed as hepatoblastoma in the left lobe of the liver. Careful diagnostic procedures are needed in such situation.

Although simple excision is the first choice of treatment, reconstruction of the biliary tree was often needed among the previously reported cases because of tumor involvement in the extrahepatic bile duct. In case 2, Whipple's procedure was performed because the tumor obstructed the ampulla of Vater and the common bile duct, and rhabdomyosarcoma was suspected. Choledochojejunostomy was performed in cases 1 and 5 [2,6]. Cystojejunostomy and ducto-cystotomy were done in case 3 because of an anomalous common bile duct [4]. Case 9 underwent hepaticoduodenostomy [9]. There was inadvertent injury of a portal vein in cases 6 and 9, which was repaired appropriately [7,9].

Postoperative chemotherapy was performed in cases 1 and 2 [2,3]. Case 1 recurred 1 year after local excision of the teratoma. In case 2, the histological diagnosis was endodermal sinus tumor associated with benign cystic teratoma and finally the patient died due to multiple metastatic recurrences. Our case was immature teratoma with focal embryoid components and it may be controversial whether to add chemotherapy after surgical excision. Macroscopically complete resection and very restricted foci of embryoid cells were our reasons for not introducing chemotherapy, but the local recurrence of mature teratoma and the existence of gliomatosis peritonei became apparent 5 months later. Gliomatosis peritonei complicated with extragonadal teratoma is very rare and few reports were published [11,12], but their prognosis was not poor. Considering these factors, very careful follow-up should be continued in the future in our case.

Conflict of interest statement

All the authors have no conflict of interest.

References