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• *CASE REPORT* •

Ruptured hepatoblastoma with massive internal bleeding in an adult

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Abstract

Hepatoblastoma is the most common primary hepatic tumor of children. However, only a very few cases have been reported in adults. Most studies support treatment with chemotherapy followed by surgical resection. We present the first reported case of adult hepatoblastoma in Taiwan. A 52-year-old female suffered from sudden onset of abdominal pain and general weakness for days. Internal bleeding with hemorrhagic shock was suspected and two massive lesions in both lobes of the liver with hemoperitoneum were noted from imaging studies. Surgical resection of the larger left lobe tumor and radio-frequency ablation of the right smaller one were performed. The histopathology diagnosis was of a hepatoblastoma.

Key words: Adult hepatoblastoma; Hepatoblastoma rupture; Internal bleeding; Hepatoblastoma; Mixed type hepatoblastoma

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INTRODUCTION

Hepatoblastoma is the most common tumor of the liver for children under the age of 2 years[1]. Reports of adult cases are extremely rare. Most of these tumors arise in the embryo during the development of the liver and contain embryonic features such as numerous mitoses. They sometimes show ribbon-like, rosette-like, or papillary formations[2]. We present the first case of adult hepatoblastoma that was reported in Taiwan.

CASE REPORT

The patient was a 52-year-old female, who suffered from abdominal discomfort and called into a regional hospital for help. Abdominal computed tomography (CT) revealed two large hepatic masses with intra-abdominal fluid accumulation. Hepatic tumor with rupture was suspected, and she was referred to our hospital for further evaluation. She had been relatively healthy in the past. There was no significant past history or family history. Laboratory studies showed the following values: AST 58 U/L; ALT 44 U/L; hemoglobin 74 g/L; platelet count 101 000/UL; hepatitis B antigen negative; anti-hepatitis B antibody positive; anti-hepatitis C antibody positive. CT scans of the abdomen (Figure 1) showed massive accumulation of intra-abdominal fluid and one large heterogeneous mass (c. 11.4 cm×18.8 cm×22 cm) with cystic and solid components in the left lobe of the liver. Another homogeneous well-enhanced lesion about 3 cm wide in segment 8 of the liver was also noted. Catheter angiography (Figure 2) showed a huge hepatic tumor with faint tumor blush in the left lobe and a 2.5 cm large hypervascular tumor in the right lobe. These were presumed to be hepatocellular carcinomas. Further intra-arterial embolization was not performed because of technical problems and lack of cooperation by the patient. We diagnosed a ruptured and bleeding hepatocellular carcinoma. Exploratory laparotomy with a left lateral hepatectomy was performed to resect the tumor. Intraoperative radio-frequency ablation (RFA) was also applied to the smaller tumor in the right lobe of the liver.

The specimen recovered consisted of an encapsulated mass measuring 22 cm×16 cm×15 cm in size with extensive necrosis and hemorrhage. Grossly, it was friable and soft in consistency (Figure 3). Microscopically, histopathology sections showed pictures of immature hepatocytic epithelial elements intermixed with prominent vessels and extramedullary hematopoiesis including all three lineages (Figure 4A). There was extensive tumor necrosis and hemorrhage. Immunohistochemical stains showed the following: vimentin positive (Figure 4B); myeloperoxidase (MPO) positive for the extramedullary hematopoietic myeloid series (Figure 4C); factor 8 positive for extramedullary hematopoietic megakaryocytes (Figure 4D); CD34 positive for vascular endothelium and hematoprogenitor cells; alpha fetoprotein negative; S100 positive. These pathology results were consistent with the diagnosis of a hepatoblastoma. The patient was discharged 10 d after the operation.

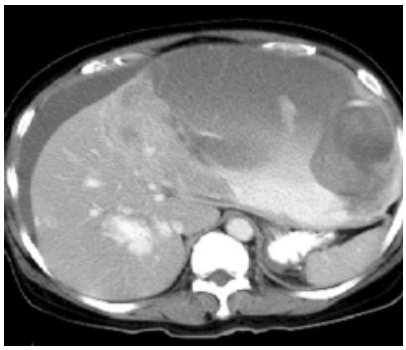

Figure 1 CT scans of the abdomen showed massive accumulation of intra-abdominal fluid and one large heterogeneous mass (c 11.4 cm×18.8 cm×22 cm) with cystic and solid components in the left lobe of the liver.

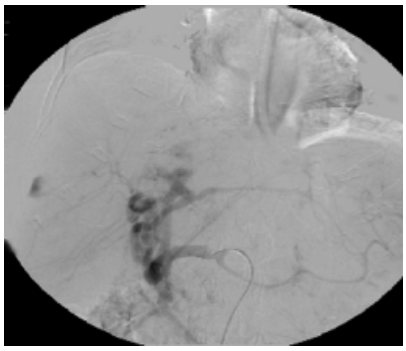

Figure 2 Catheter angiography showed a huge hepatic tumor with faint tumor blush in the left lobe and a 2.5 cm large hypervascular tumor in the right lobe. These were presumed to be hepatocellular carcinomas.

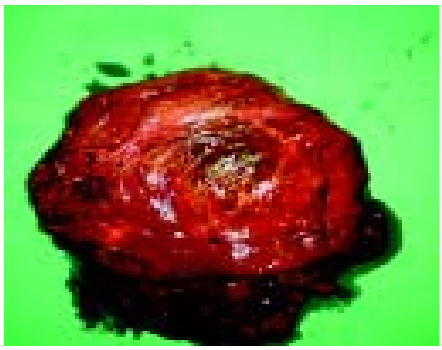

Figure 3 The specimen recovered consisted of an encapsulated mass measuring 22 cm×16 cm×15 cm in size with extensive necrosis and hemorrhage. Grossly, it was friable and soft in consistency.

DISCUSSION

Hepatoblastoma is a highly malignant tumor occurring in infants, and reports of adult cases are extremely rare. A review search in the literature revealed only 23 adult cases of hepatoblastoma including the present one. This case is also the first adult hepatoblastoma that was reported in Taiwan.

Ishank and Glunz classified hepatoblastomas into two groups in 1967: epithelial type and mixed epithelial and mesenchymal type[3]. The tumor type from this patient was distinguished from a hepatocellular carcinoma by the presence of angiodysplasia and also by the peculiar arrangement of bright and dark epithelial cells in sheets or ribbons[3-5]. This patient, therefore, had a tumor of the mixed epithelial and mesenchymal type according to this

Table 1 Summary of reported hepatoblastoma in adults

Case	Reporter	Sex/Age (yr)	Pathology type	Diagnosis	Survival time
1	Barnett[6] (1958)	M/35	Mixed	Embryonic tumor	Died 1 mo after surgery
2	Alexander[7] (1961)	F/68	Mixed	Primary mixed tumor	Postmortem
3	Ojima[8] (1964)	M/48	Mixed	Malignant mixed tumor	Postmortem
4	Kerr[5] (1966)	M/56	Mixed	Hepatic embryonic mixed tumor	Sudden death
5	Blanding[9] (1968)	M/84	Mixed	Malignant tumor	Died after biopsy
6	Cater[10] (1969)	M/78	Mixed	Hepatoblastoma	1 mo after surgery
7	Goldman[11] (1969)	F/65	Mixed	Rhabdomyosarco-hepatoma	Sudden death
8	Meyer[12] (1974)	F/19	Mixed	Hepatoblastoma	Not described
9	Ludwig[13] (1975, case 1)	F/53	Mixed	Mixed malignant tumor	Died after surgery
10	Ludwig[13] (1975, case 2)	M/40	Mixed	Mixed malignant tumor	Postmortem
11	Jameson[14] (1978)	F/51	Mixed	Hepatoblastoma	Postmortem
12	Yoshida[15] (1979)	M/60	Mixed	Hepatoblastoma	Died 2 mo after surgery
13	Honan[16] (1980)	F/27	Mixed	Mixed hepatoblastoma	Postmortem
14	Kishimoto[17] (1984)	M/60	Mixed	Malignant mixed tumor	Died 2 d after surgery
15	Kawarada[18] (1985, case 1)	M/43	Mixed	Nonhepatocytic malignant mixed tumor	Alive for 32 mo after surgery
16	Kawarada[18] (1985, case 2)	M/49	Mixed	Hepatocytic malignant mixed tumor	Postmortem
17	Sugino[2] (1989)	M/22	Epithelial	Hepatoblastoma	Died 9 mo after surgery
18	Altmann[19] (1992, case 1)	M/73	Mixed	Hepatoblastoma	Not described
19	Altmann[19] (1992, case 2)	F/35	Epithelial	Hepatoblastoma	Not described
20	Harada[20] (1995)	M/24	Mixed	Hepatoblastoma	Died 16 mo after surgery
21	Sugino[2] (1995)	F/22	Mixed	Hepatoblastoma	Alive for 38 mo after surgery
22	Ahn[1] (1997)	F/51	Mixed	Hepatoblastoma	Died 2 mo after surgery
23	Present case (2005)	F/52	Mixed	Hepatoblastoma	Alive 6 mo after surgery

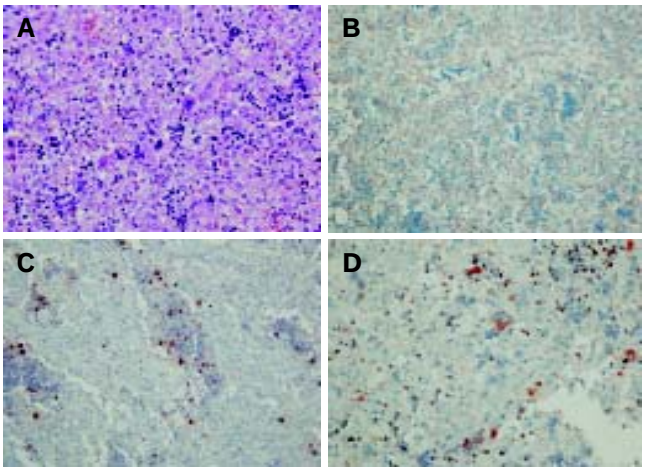

Figure 4 Immature hepatocytic epithelial elements intermixed with prominent vessels and extramedullary hematopoiesis including all three lineages (A). Vimentin positive (B); MPO positive for the extramedullary hematopoietic myeloid series (C); factor 8 positive for extramedullary hematopoietic megakaryocytes (D).

classification. Most cases in the literature are of the mixed type.

The precise mechanism to cause the tumor has been elusive; present investigations of the cytogenetic and molecular genetic aberrations in hepatoblastomas revealed involvement of chromosomal loci on 1q, 2 (or 2q), 4q, 8 (or 8q), and 20[21]. Loss of heterozygosity imprinting at locus 11p 15.5 also suggest a common genetic basis for hepatoblastoma[21]. Shin-ichi *et al*., found that abnormalities in Wnt signaling pathway seen in frequent mutation of the β-catenin gene may play a role in the genesis of hepatoblastoma[22]. They also found that the patients with hepatoblastoma with high expression of polo-like kinase 1 (PLK1) represented significantly poorer outcome[22]. Therefore, PLK1 may become a useful tool to develop new diagnostic as well as therapeutic strategies of hepatoblastoma.

Otherwise, only one of the previously reported cases was a 12-year-old boy who presented with a ruptured tumor with massive bleeding as we saw in this patient[23]. We successfully resected the larger hepatic tumor and treated the smaller one with RFA, although most adult cases of hepatoblastoma have been unresectable[4]. The clinical and pathological findings of previously reported cases and the present one are summarized in Table 1. The mean age of the 23 patients was 48.5 years and there were more males (13) than females (10). Eleven patients received surgery. Twenty one of these tumors were of the mixed type. Survival time ranged from 1 to 38 mo after surgery with a mean survival time of 9.7 mo. The prognosis for patients with this disease is very poor. Current treatments for the tumor include surgery and chemotherapy. Combination chemotherapy with adriamycin and cisplatin has been considered effective for hepatoblastoma[2]. Our patient has just been discharged and will be followed up.

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