Letters to the Editor

Spontaneous rupture of a hepatic sarcoma in an adult resembling hydatid cyst

Key words: Undifferentiated (embryonic) sarcoma. Hepatectomy. Rupture of liver sarcoma. Hydatid cyst.

Dear Editor,

Undifferentiated (embryonic) sarcoma of the liver (USL) is a rare primary malignant liver tumour. It is most frequently seen in late childhood, but it is quite exceptional in adults (1,2). Only 11 cases have been reported in 2003 in patients over age 40. As clinical presentation in adults it is poor and uncharacteristic USL can reach a large size (10-25 cm). Treatment is usually delayed for months due to vague symptoms and erroneous image findings (3). The prognosis is poor but recent evidence has shown that long-term survival is possible after complete surgical resection with tumour-free margins, with or without postoperative chemotherapy (1).

Case report

A 65-years-old female was referred to our Hospital Emergency with diffuse abdominal pain more intense in the right upper quadrant. The pain had started about 24 hours earlier. She did not present weight loss, fever, anorexia, vomiting or abdominal distension. She had been waiting for elective surgery for one month with hydatid cyst diagnosis. Physical examination revealed a healthy-looking adult woman. The liver edge was palpable near 8 cm below the right costal margin. Laboratory data included hemoglobin, 10.4 g/dl; white cell,

8.800/mm³; c-reactive protein, 5.0 mg/dl; gamma g.t., 54 U/l; AFP, CEA and CA 19.9 yielded normal results. Computed tomography scans showed a large mass (18 x 12 cm) involving the entire right lobe. It had multicystic appearance with solid portions and calcified capsule. There were capsule interruptions around the tumour and peritoneal free fluid. The patient underwent surgery and peroperatively enormous hepatomegaly and hemoperitoneum (800 cc) were observed due to spontaneous rupture in the peritoneal cavity of a tumour involving the entire right lobe of the liver (Fig. 1).

Fig. 1. Peroperative photograph of the capsule rupture situated around the tumour (arrow).

Blood was evacuated and right hepatectomy and cholecystectomy were performed, resulting in complete removal of the tumour with tumour-free resection margins. The patient had a postoperative uneventful recovery. No further adjuvant irradiation

tion or chemotherapy was administered. The specimen from the right hepatectomy presented a ruptured capsule opposite to the resected margin. Most of the parenchyma was replaced by a 15 x 15 x 11 cm multicystic tumour mass with well defined margins but no capsulation. Microscopic examination showed: sarcoma with features reminiscent of a rhabdomyosarcoma. One year after the operation, the patient remains well with no evidence of local recurrence or metastasis.

Discussion

USL it is composed of undifferentiated mesenchymal cells and it is extremely rare in adulthood, mainly in patients over age 60. As in our case, the tumour is a solitary, well-circumscribed solid-to-cystic lesion, most commonly occurring in the right lobe. Distant metastasis at the time of presentation are relatively uncommon. Because of the rarity of this tumour type in adults no prospective treatment studies or larger series reporting survival data and treatment strategies are available (4). Lenze (4) did analyze 68 patients of USL in older children (≥ 15 years) and adults reported in literature, to identify survival rates and optimal treatment strategies. The median survival of all patients was 29 months. Patients who underwent complete tumour resection followed by adjuvant chemotherapy survived over a median follow-up of 28.5 months and had better survival compared with patients who underwent surgery alone. This is an important study but it is not a prospective one, groups are not comparable and it contains only a few number of cases. According to some authors (1,5) tumor resection with chemotherapy is indispensable for the treatment of ruptured USL in children. However, in adult patients over 65 years of age, as in our case, it has not yet been established that adjuvant chemotherapy after complete lesion resection can decrease recurrence or metastasis (6). According to Lenze neoadjuvant therapy may become an option in the strategy for treating adults when diagnosis can be made pre-operatively (4). Our patient was 65 years old, which is older than the peak age group for

USL. The patient had a large tumour with multiple cystic areas resembling hydatid cyst as reported in other cases (2,3). With this diagnosis she was waiting for elective surgery. Spontaneous tumour rupture and hemoperitoneum led us to perform an emergency operation. Tumour involved the entire right hepatic lobe but did not involve diaphragm nor inferior vena cava. Right hepatectomy and cholecystectomy were performed with complete removal of the tumour with free resection margins. No adjuvant irradiation or chemotherapy was administered.

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