Rare disease

Primary liposarcoma of liver: a rare case and literature review

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Summary

Primary liposarcoma of the liver is extremely rare. Because of its rarity, the knowledge of the clinical course, management and prognosis of it are limited. Here we report a case of primary myxoid liposarcoma of the liver in an 83-year-old Iranian woman.

BACKGROUND

Malignant tumours occurring in the liver can be primary or metastatic. Involvement of the liver by metastatic malignancy is far from common than primary hepatic neoplasia. Primary tumours of the liver are divided into epithelial and mesenchymal lesions. Most primary epithelial liver cancers arise from hepatocytes and are termed hepatocellular carcinoma. Among the primary malignant mesenchymal tumours of the liver, angiosarcoma is the most common. Liposarcoma of the liver is a rare malignant tumour^{1–5} and most cases are metastatic.^{4–5} Primary liposarcoma of the liver is extremely rare.^{2–6} Only a few cases of primary liver liposarcoma have been reported. Here we have presented one case of primary liver liposarcoma.

CASE PRESENTATION

An 83-year-old Iranian woman was admitted to a private hospital, 2 years ago, because she had nausea, vomiting and a palpable painful epigastric mass. According to her medical record she had lost 8 kg of body weight within 6 months. Her medical history showed no serious problems. She had no risk factor for liver disease, such as alcohol, drug or viral infection. She had never been exposed to any specific chemical agent. On physical examination, she was not icteric, and no significant signs of chronic liver disease, including oedema, ascites or splenomegaly, were detected. Neither skin abnormalities such as ecchymosis nor lymphadenopathy was identified. On abdominal examination, there was fullness in the epigastric area with extension to the left side that was tender in deep palpation.

INVESTIGATIONS

Routine laboratory test results showed 70 IU/l alanine aminotransferase (normal=5–45 IU/l), 65 IU/l aspartate aminotransferase (normal=5–45 IU/l) but total bilirubin, direct bilirubin, alkaline phosphatase and albumin were normal. Virological examination revealed that she was not infected with hepatitis B or C viruses. Serum levels of alfa-fetoprotein (AFP) and carcinoembryonic antigen (CEA) were measured and both were within normal

limits. Abdominal ultrasonography showed a liver mass. Abdominal CT scan revealed a low-density 21×20×9 cm mass that originated from the left liver lobe and extended to the left side of the abdominal cavity (figure 1). Fine-needle aspiration (FNA) biopsy was performed, but failed to render adequate material. The patient underwent laparotomy. At laparotomy a huge tumour occupied the left lobe of the liver. The surgeon performed a resection of the tumour. The resected specimen weighed 2750 g and gross examination revealed a yellowish tumour measuring 34×26 cm. The cutting section of the tumour appeared yellowish gelatinous with haemorrhagic foci. The diagnosis was myxoid liposarcoma. In her medical record, it was not clear whether surgical margins were involved or not. The patient was discharged 10 days after the first operation.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis of hepatic liposarcoma includes variable fatty hepatic masses such as fatty change,

Figure 1 Abdominal CT scan revealed a low-density mass that originated from the left liver lobe and extended to the left side of the abdominal cavity.

adenoma, focal nodular hyperplasia, lipoma, angiomyolipoma, teratoma, pseudolipoma of the Glisson capsule and fat-containing hepatocellular carcinoma.⁷

TREATMENT

The patient was referred to an oncologist and received six courses of MAID regimen chemotherapy (mesna, adriamycin, ifosfamide, dacarbazine).

OUTCOME AND FOLLOW-UP

She was well for 19 months. After that she complained of abdominal discomfort (abdominal fullness and right upper quadrant pain) and was referred to our hospital. Again CT scan revealed a huge mass in the left liver lobe. The glass slides related to the original resected mass from the outside hospital were reviewed. Microscopic examination revealed oval to round tumour cells without overt pleomorphism in a myxoid background with prominent blood vessels. The blood vessels were delicate, thin walled and arborising, and had a 'chicken-wire' appearance (an arborising and delicate plexiform capillary vascular network that was present throughout these tumours and provided an important clue for distinguishing it from myxomas). A few tumour cells resembled lipoblast with multiple cytoplasmic vacuoles and eccentrically located indented nuclei (figure 2). The diagnosis was myxoid liposarcoma. Reoperation was done and a 14×8×3 cm mass was resected. Histopathological examination revealed myxoid liposarcoma without necrosis or increased round cell percentage and negative surgical margins. Postoperative CT scan did not show any gross tumour residue. After the second surgery, the patient is being followed carefully and now she is in good general condition without any evidence of recurrence.

DISCUSSION

Although liposarcoma is one of the most common soft tissue sarcomas in adults, primary liposarcoma of the liver is very rare. To date, only a few case reports of primary hepatic liposarcoma have been reported in the English literature.^{3 4 6 8–13} To our knowledge, 12 primary liver liposarcoma cases were reported previously; one of them

was an Iranian woman.¹⁴ Among the reported cases there is no gender and location predilection and the ages are between 26 months and 86 years. In previous cases only one case above 80 years (an 86-year-old man)¹⁵ was reported. Our patient was an 83-year-old woman. The clinical characteristics of hepatic liposarcoma are typically a painless slow growing mass.¹⁶ In our patient, a palpable mass was detected on physical examination. Most of the symptoms are caused by displacement of nerve, vessels, biliary tracts and intestinal structures³ and include jaundice, fever, vomiting, nausea, abdominal fullness, right upper quadrant pain and weight loss. In the presented case, nausea, vomiting, abdominal fullness, pain and weight loss existed but jaundice was not detected because the tumour was confined to the left liver lobe without any pressure effect on the common bile duct. Mild increases in the serum levels of liver transaminases, alkaline phosphatase, gamma glutamyl transferase and CA19-9 were reported in previous studies. However in our patient only liver transaminases were mildly increased, but AFP and CEA were in normal ranges. According to radiological features, lipomatous tumours are easily characterised as fat-containing masses, although distinguishing between a malignant tumour and a benign one may not be possible. Therefore histological examination is important for proper diagnosis.¹⁷ Radiologically, myxoid liposarcoma shows a well-defined border with low attenuation cystic appearance or a heterogeneous structure.¹⁸ In our case the first CT scan showed a low-density mass with mild peripheral contrast enhancement. The second CT scan revealed central cystic change. However, in both of them the tumour had well-defined borders. In addition, CT scan is still the best tool to evaluate the extension of the tumour before surgery.¹⁹ Abdominal sonography is a valuable tool for diagnosis and postoperative follow-up.¹⁹ In our patient abdominal sonography examination was performed every 6 months, but when tumour recurrence was detected, it was too large. In another study, Kuo *et al*³ showed that tumours can grow significantly even in a 12 week interval between sonography examination. It seems that regular abdominal sonography must be repeated in shorter intervals. Although classic myxoid liposarcoma has unique cytomorphological features, FNA biopsy renders a definite diagnosis in only 58–82% of cases.²⁰ Ancillary studies may be needed to confirm the diagnosis and to differentiate myxoid liposarcoma (MLS) from other tumours with similar features. In the presented case FNA biopsy was performed but failed to render adequate material. More than half of the hepatic liposarcomas are of myxoid type.¹⁷ The presented case also had similar morphology. According to one study,²¹ old age (>45 years), necrosis and high histological grade are poor prognostic features. In the current case, although the patient was 83 years old, there was no tumour necrosis or increased round cell percentage and she was in good performance status. This may be the reason for the relatively good prognosis despite old age in our patient.

Curative and aggressive surgery with negative surgical margins is still regarded as the best policy in the management of primary liver liposarcoma.³ The role of adjuvant therapy is unclear. Although radiation therapy can be used, the total volume of the liver has a limited radiation dose tolerance and because these tumours are usually large,

Figure 2 Microscopic examination revealed oval to round tumour cells without overt pleomorphism in a myxoid background, ×40.

radiotherapy could be dangerous and harmful. In the present case with regard to the negative surgical margins in the second surgery, we did not prescribe radiation therapy. The 5-year survival rate of patients who have undergone curative resection or radiotherapy is approximately 50%.⁴ Generally, the prognosis is poor without treatment. In conclusion, although primary liposarcoma of the liver is exceedingly rare, this tumour should be considered in the differential diagnosis, especially in those patients who are potential candidates for liver resection or orthotopic liver transplantation. Liposarcoma is an absolute contraindication for liver transplantation.

Learning points

- ▶ Primary liposarcoma of the liver is extremely rare and for this reason the knowledge of the clinical course, management and prognosis of it are limited.
- ▶ It should be considered in the differential diagnosis of hepatic mass, especially in those patients who are potential candidates for liver resection or orthotopic liver transplantation.
- ▶ Histological examination is important for proper diagnosis.
- ▶ En bloc resection of the tumour must be performed whenever possible. Re-resection may be helpful in local recurrences.
- ▶ Because of rapid tumour growth in tumour recurrences, we propose regular abdominal sonography examination in time intervals of less than 12 weeks.

Competing interests None.

Patient consent Obtained.

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