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Clinical case of surgical treatment of undifferentiated pleomorphic liver sarcoma

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Undifferentiated pleomorphic liver sarcoma (UPS), formerly known as malignant fibrous histiocytoma (MFH), represents a very rare primary hepatic tumour. It was first described by O'Brien and Stout in 1964. This type of tumour is the most prevalent malignant soft tissue tumour, which usually occurs in adulthood and affects the extremities, less commonly the retroperitoneum and abdominal organs.

Objective – to present the treatment outcomes of a rare case of UPS of the liver. The article describes a clinical case of surgical treatment of a patient with UPS of the liver. Patient N., 36 years old, complained of pain in the right hypochondrium, fever, and general weakness. According to the results of instrumental examinations, the clinical diagnosis was made: primary liver tumour, tumour rupture, and intra-abdominal bleeding. After preoperative preparation, a right-sided hemihepatectomy, D2 lymphadenectomy, cholecystectomy, and abdominal drainage were performed. The histopathological and immunohistochemical features of the tumour cells were most consistent with undifferentiated pleomorphic liver sarcoma. According to the Federation Nationale des Centres de Lutte Contre le Cancer (FNCLCC) sarcoma grading system, a total score of 7 (G3) was assigned. The diagnosis was undifferentiated pleomorphic liver sarcoma, $pT_{4a}N_0M_0$ G3 stage III, grade 2. Six months later, a CT scan of the abdominal cavity revealed a tumour focus in the right subdiaphragmatic space with invasion of the right diaphragm dome, liver segment IV, and right kidney.

Surgical intervention was performed in volume: viscerolysis, atypical resection of the SgIV liver with right-sided nephrectomy and resection of the right dome of the diaphragm, aortocaval lymphadissection. Postoperative diagnosis: undifferentiated pleomorphic liver sarcoma $rpT_4N_{1(1/2)}M_0$ G3, R0, stage III, grade 2. Currently, the period of recurrence-free observation is 12 months after the second surgery.

Keywords
undifferentiated pleomorphic liver sarcoma, sarcoma, neoplasms.

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Undifferentiated pleomorphic liver sarcoma (UPS), formerly known as malignant fibrous histiocytoma (MFH), represents a very rare primary hepatic tumour. It was first described by O'Brien and Stout in 1964 [6]. This type of tumour is the most prevalent among soft tissue malignancies, typically occurring in adulthood and affecting the extremities, less commonly the retroperitoneum and abdominal organs [8]. The first case of UPS of the liver was described in 1985, but the disease remains poorly understood [1]. The main clinical sign observed in 78.4 % of cases was pain or discomfort in the right hypochondrium, followed by weakness, weight loss, jaundice, and fever. In 15 % of cases, no clinical manifestations were noted [5]. Overall, UPS ranks 4th among the most common soft tissue sarcomas. The incidence is 0.8–1.0 per 100,000 among the European race. In UPS of the liver, a solitary lesion with a size of 3.6–17.8 cm (average 9.7 cm) is observed [4, 7]. Abdominal ultrasound and contrast-enhanced CT combined with

trepan biopsy are the main diagnostic methods for verifying UPS of the liver [3, 9, 10]. A significant proportion of cases involving this pathology are misdiagnosed; specifically, 15 % of patients with UPS are treated as having benign focal liver disease. The local recurrence rate is 19–31 %, the metastasis rate is 31–35 %, and the 5-year survival rate is 65–70 % [2]. In China, a total of 76 cases were reported, with 50 cases involving men, resulting in a male-to-female ratio of 1.9 : 1. The average age of the patients was 51 years, with more than 85 % being over 40 years old. Only 45 clinical cases of UPS of the liver have been described in the English-language literature.

In this paper, we present a clinical case of surgical treatment of a patient with UPS of the liver. Patient N., 36 years old, complained of pain in the right hypochondrium, fever, and general weakness. Ultrasound examination revealed no free fluid in the pleural and abdominal cavities. In the right lobe of the liver, a solid mass with an indistinct contour, measuring 180 × 150 mm, occupied the entire right lobe of the liver.

The patient underwent a CT scan with IV enhancement: a tumour-like mass measuring 188 × 117 mm with areas of haemorrhage was detected in the parenchyma of the right liver lobe (Fig. 1).

This mass invaded the right hepatic vein and the right Glissonean pedicle, passing along the right contour of the middle hepatic vein. There were no signs of contrast extravasation at the time of the study. The patient underwent 3D modelling of the liver (CT volumetry and segmentation) (Fig. 2).

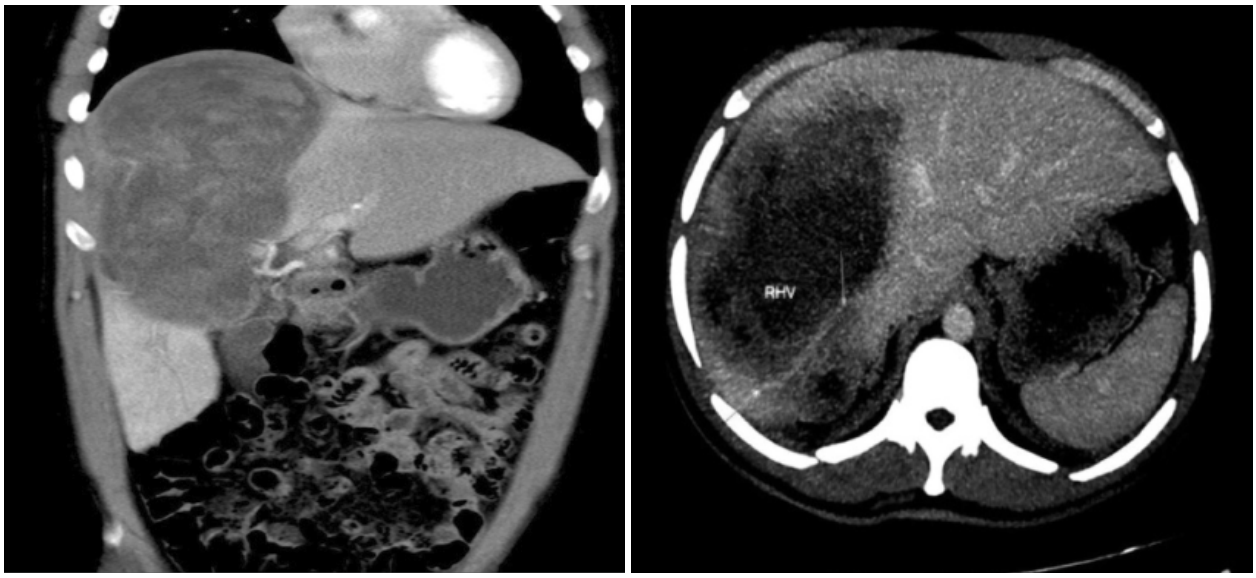

Figure 1. **CT scan of the abdominal cavity**

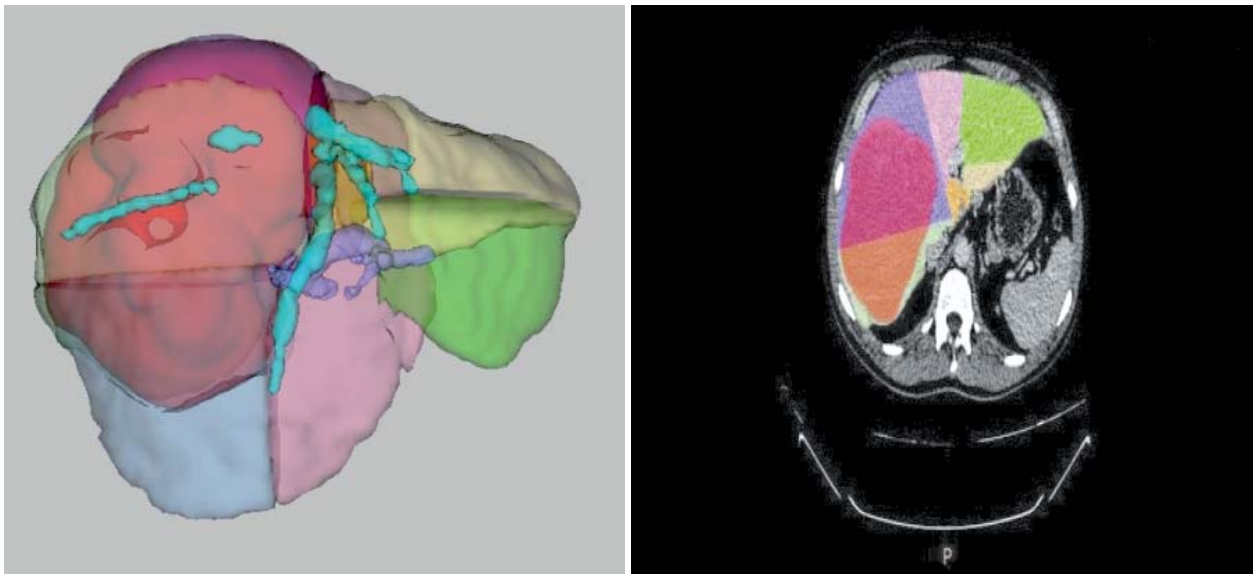

Figure 2. **CT volumetry and liver segmentation**

After the examination, the clinical diagnosis was made: primary liver tumour, tumour rupture, and intra-abdominal bleeding. After preoperative preparation, the patient underwent surgery, including right-sided hemihepatectomy, D2 lymphadenectomy, cholecystectomy, and abdominal drainage. The extent of D2 lymph node dissection included the removal of lymph node groups 12, 8, 13a, and 9 (Fig. 3).

Pathological diagnosis: Solid cystic liver mass morphologically similar to malignant neoplasia of mesenchymal origin (Fig. 4).

Immunohistochemical examination was performed. The tumour cells were diffusely positive for histone H3 with a K27M mutation, INI-1, and focally positive for desmin. A focal spot reaction to total cytokeratin and cytokeratin SAM 5.2 was observed. The tumour cells were positive for CD10 and partially positive for CD13, SALL4. The tumour cells were negative for NMV-45, TFE3, inhibin alpha, GFAP, MDM2, MyoD1, myogenin, CD68, CD45, CD34, RAX-8, caldesmon, STAT6, S-100, SOX-10, CDK4, ERG, DOG-1, CD43, EMA, PLAP, synaptophysin, OST3/4, epithelial antigen Ber-EP4. Thus, this morphological picture and immunophenotype of the tumour cells are most consistent with undifferentiated pleomorphic liver sarcoma.

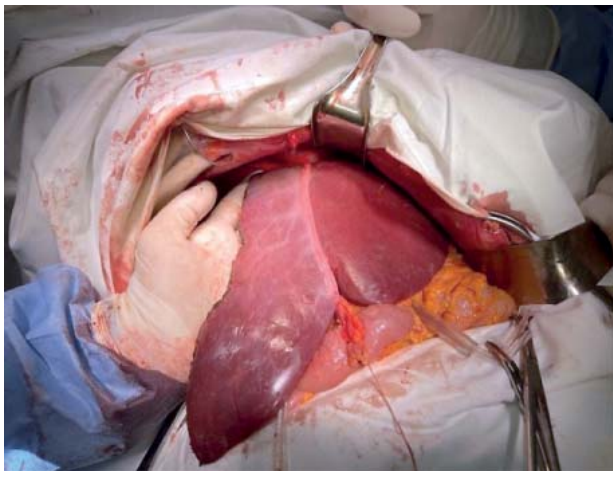

Figure 3. **Intraoperative photo**

According to the Federation Nationale des Centres de Lutte Contre le Cancer (FNCLCC) sarcoma grading system, the tumour scored 3 points for differentiation, 3 points for mitotic activity (22 per 10 high-power fields at magnification × 400), and 1 point for necrosis (10 %), resulting in a total score of 7 (G3). No signs of lymphatic, vascular and perineural invasion were found within the studied specimens. Macroscopically, the tumour was limited to the liver tissue, but had two growth loci (visceral surface and diaphragmatic surface) – multifocal growth and corresponded to an undifferentiated pT4aNoMo G3 pleomorphic liver sarcoma.

The patient was discharged on the 25th postoperative day in satisfactory condition to continue combined therapy under the supervision of a clinical oncologist at his place of residence.

Three months after surgery, oncological screening and a contrast-enhanced CT scan of three anatomical regions were performed. No CT signs of disease progression were detected.

Six months following the prior examination, subsequent oncological screening identified a recurrence of the disease. A CT scan of the abdominal cavity revealed a tumour focus in the right subdiaphragmatic space, with invasion into the right dome of the diaphragm, segment IV of the liver, and the right kidney (Fig. 5).

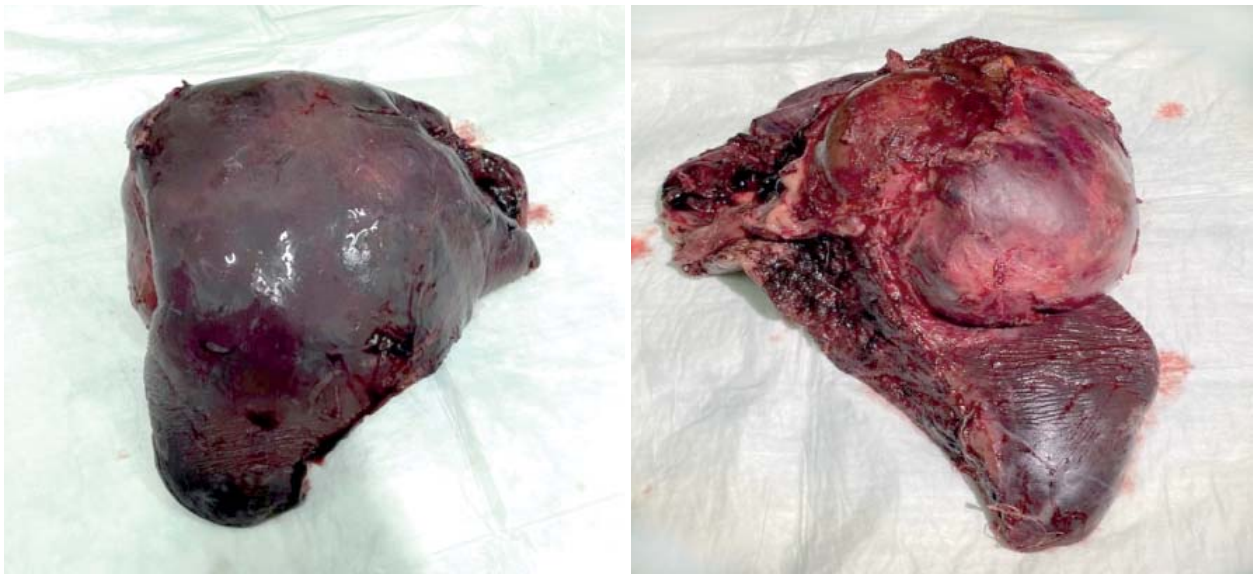

Figure 4. **Macropreparation**

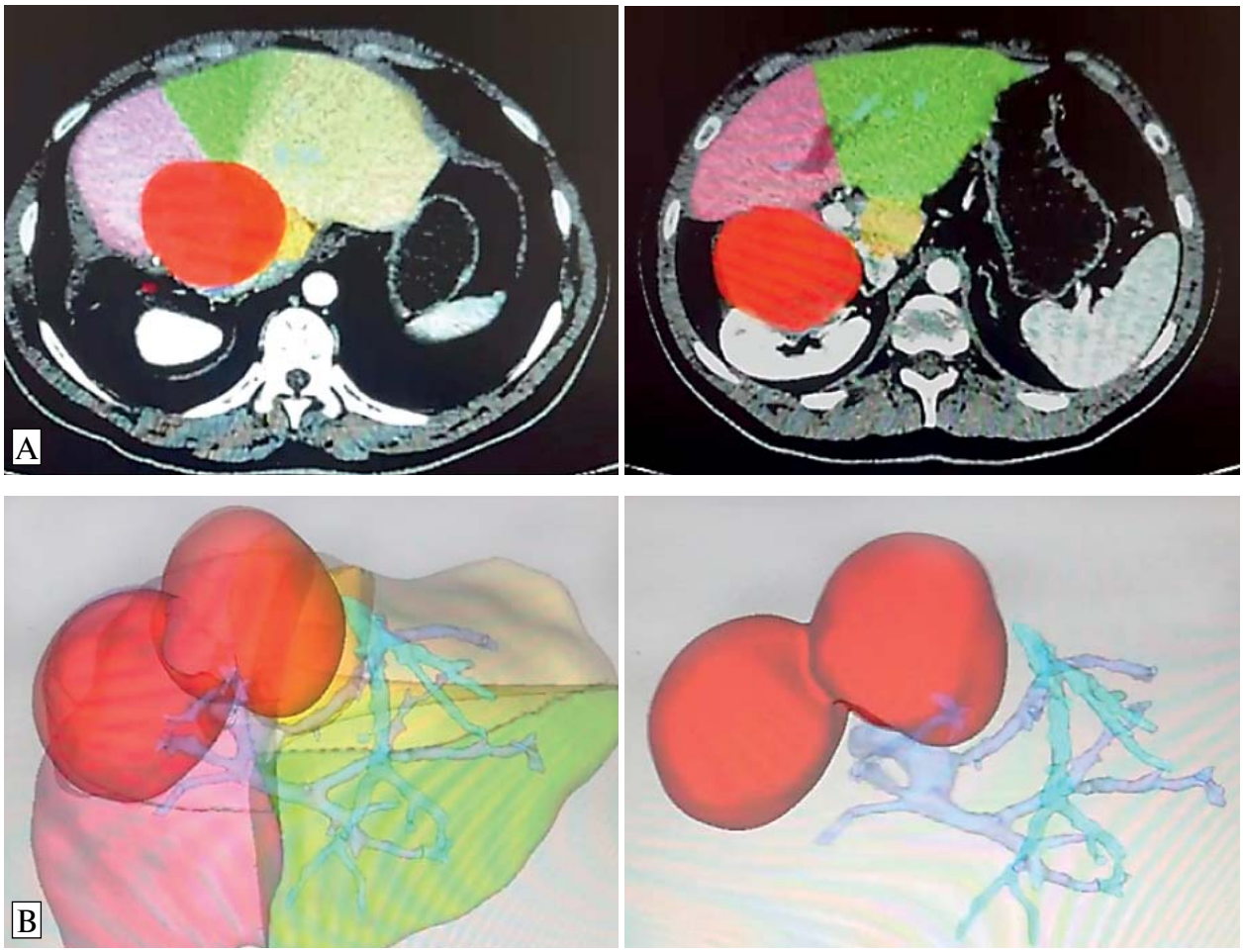

Figure 5. **CT volumetry (A) and liver segmentation (B)**

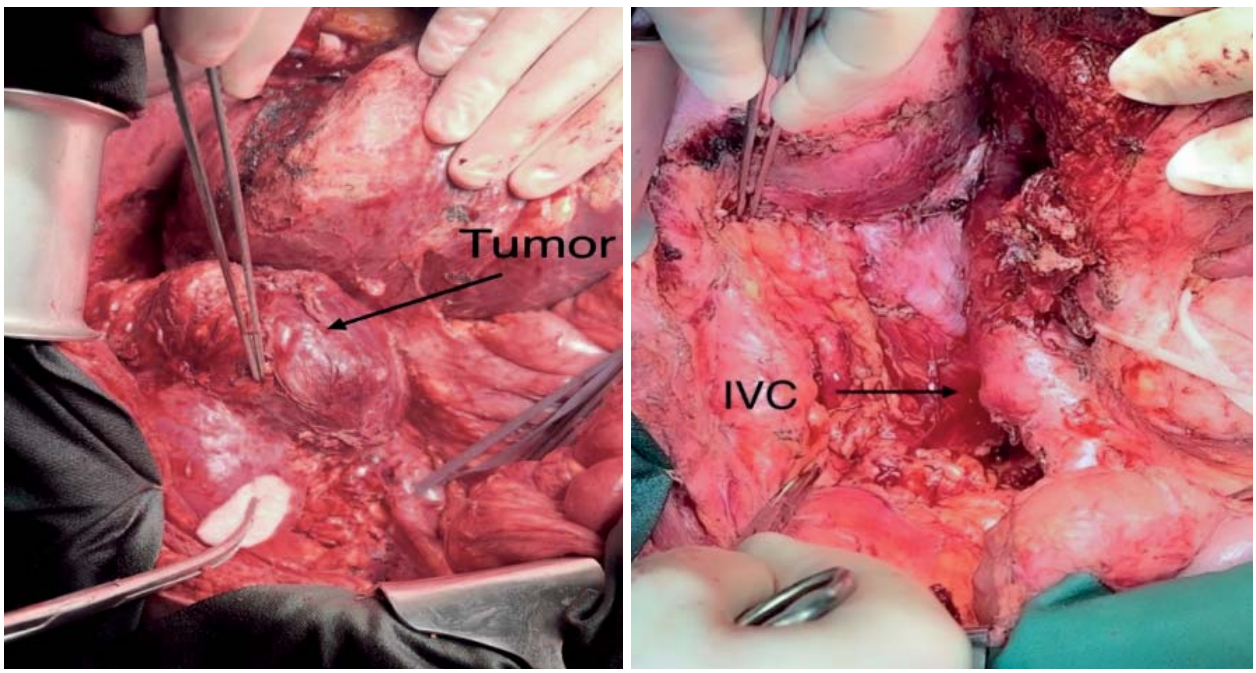

Figure 6. **Intraoperative photo**

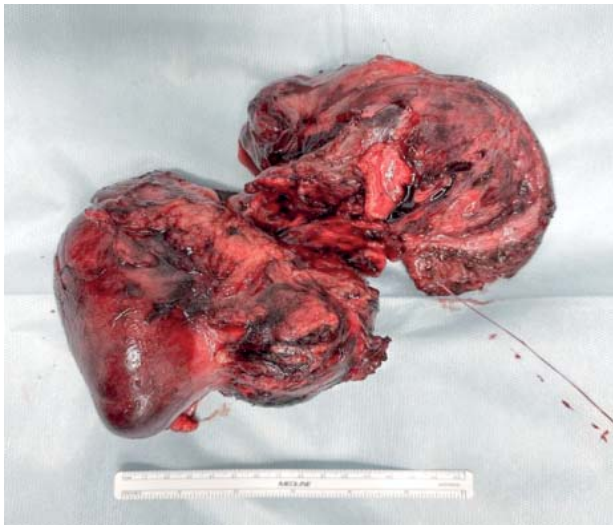

Figure 7. **Macroscopic specimen of the tumour: recurrent tumour with SgIV in the liver and right kidney**

After preoperative preparation, the patient underwent surgery, which included viscerolysis, atypical SgIV liver resection with right-sided nephrectomy and resection of the right dome of the diaphragm, and aortocaval lymphadissection (Fig. 6, Fig. 7). The extent of lymph node dissection included the removal of lymph node groups 16a2, 16b1.

The patient was discharged on the 11th postoperative day in satisfactory condition to continue combined therapy under the supervision of a clinical oncologist.

Pathological examination revealed the following findings:

1. The liver tissue contained a solid cystic mass composed of stellate, spindle-shaped, epithelioid and extremely pleomorphic multinucleated cells, forming solid layers around single duct-like structures lined with cylindrical epithelium. The tumour cells exhibited pronounced polymorphism. The morphological picture was most consistent with recurrent undifferentiated pleomorphic sarcoma. According to the FNCLCC sarcoma grading system, the tumour scored 3 points for differentiation, 3 points for mitotic activity (24 per 10 high-power fields at magnification ×400), and 2 points for necrosis (60%), resulting in a total score of 8 (G3). There were no signs of lymphatic, vascular or perineural invasion within the studied specimens. The largest tumour size measured 15.2 cm. Tumour invasion into the kidney tissue and the adjacent diaphragmatic flap was observed, corresponding to rpT3. The adrenal gland was not involved in the tumour process.

The resected margins of the ureter, renal vessels, paranephric adipose tissue, diaphragm, and liver parenchyma showed no evidence of tumour involvement (R0).

2. Of the two examined lymph nodes, one demonstrated tumour metastasis with histological features similar to the primary liver tumour (1/2).

Postoperative diagnosis: undifferentiated pleomorphic liver sarcoma (G3) $rpT_3rpN_{1(1/2)}$ R0.

Currently, the recurrence-free follow-up period is 12 months after the second surgery.

Conclusions

Although UPS of the liver is a rare malignant mesenchymal tumour, it should be considered in the diagnosis of large liver lesions. The clinical manifestations and diagnostic features are variable, which makes preoperative identification challenging. Predictive factors include patient age, tumour size, histological grade, histological subtype, and anatomical location. Surgery remains the primary treatment strategy for undifferentiated pleomorphic liver sarcoma.

Declaration of interests

The authors declare that they have no conflicts of interest.

Ethics approval and written informed consents statements

All procedures were carried out in compliance with the current legislation of Ukraine on ethics, the principles of Good Clinical Practice (ICH 6CP), and the recommendations of the 2013 Helsinki Declaration.

Authors contributions

Y. P. Bakunets: critical review of the manuscript; Y. P. Bakunets, M. S., Zagriychuk I. A., Bryndak R. A. Samokishchuk: work concept, design, and critical review of the manuscript; F. O. Prytkov: work concept and design, data collection and analysis, and writing the manuscript; M. I. Korshunova: data collection, analysis, and writing the manuscript.

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Клінічний випадок хірургічного лікування недиференційованої плеоморфної саркоми печінки

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Недиференційована плеоморфна саркома (НПС) печінки, раніше відома як злоякісна фіброзна гістіоцитома (MFH), дуже рідко локалізується в печінці як первинна пухлина. Вперше описана в 1964 р. O'Brien і Stout. Цей тип пухлини є найпоширенішим серед злоякісних пухлин м'яких тканин, зазвичай виникає в дорослому віці та вражає кінцівки, рідше – заочеревинний простір й органи черевної порожнини.

Мета — представити результат лікування рідкісного випадку недиференційованої плеоморфної саркоми печінки.

Описано клінічний випадок хірургічного лікування пацієнта з НПС печінки. Пацієнт Н., 36 років, звернувся зі скаргами на біль у правому підребер'ї, підвищення температури тіла, загальну слабкість. За результатами інструментальних обстежень установлено клінічний діагноз: первинна пухлина печінки, розрив пухлини, внутрішньочеревна кровотеча. Після проведення доопераційної підготовки виконано правобічну гемігепатектомію, лімфаденектомію D2, холецистектомію, дренування черевної порожнини. Морфологічна картина й імунофенотип клітин пухлини найбільше відповідали НПС печінки. За системою градації сарком Federation Nationale des Centres de Lutte Contrele Cancer (FNCLCC) загальна кількість балів – 7 (G3). Встановлено діагноз: НПС печінки $pT_{4a}N_0M_0$ G3 III ст., 2 клінічна група. Через 6 міс за даними комп'ютерної томографії органів черевної порожнини виявлено пухлинне вогнище в правому піддіафрагмальному просторі з інвазією в правий купол діафрагми, IV сегмент печінки, праву нирку. Виконано оперативне втручання в обсязі: вісцероліз, атипова резекція SgIV печінки з правобічною нефректомією та резекцією правого купола діафрагми, аортокавальна лімфодисекція. Післяопераційний діагноз: НПС печінки $rpT_4N_{1(1/2)}M_0$ G3, R0, III ст., 2 клінічна група. Тривалість безрецидивного спостереження становить 12 міс після повторного оперативного втручання.

Ключові слова: недиференційована плеоморфна саркома печінки, саркома, неоплазія.

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